

Multidisciplinary Management of Primary Sacral Tumors: A Tertiary Care Center's Experience and Literature Review

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Sacral tumors are rare and can be benign or malignant. Their management is multifactorial and is based on the pathology, extent, and local and distant spread. Managing sacral tumors is challenging due to their proximity to visceral and neural structures. Surgical wide excision has been the standard of care for aggressive benign and malignant tumors. Our purpose was to evaluate the outcomes of a multimodal approach to managing primary sacral tumors in Sakra World Hospital, a tertiary spine care center in Bengaluru, India and perform a literature review to determine a workflow pathway. Our study was a retrospective review of patient records and included 15 patients with primary sacral tumors. Eleven surgically treated patients were evaluated clinically and radiologically and underwent biopsy before surgical excision by an all-posterior approach. A multidisciplinary approach that included intraoperative neural monitoring, plastic reconstruction, adjuvant chemotherapy, and radiotherapy was implemented whenever necessary. Sacral root preservation was attempted whenever feasible. Functional outcomes (based on the Visual Analog Scale [VAS] and Biagini scoring system) were analyzed along with disease control, with a minimum of 2 years of follow-up. The mean follow-up was 29±9.8 months. The mean VAS score significantly improved from 7.8±2.6 to 3.7±3.8 ($p=0.026$). Bowel function showed statistically significant improvement, from a mean score of 0.81±0.47 to 0.63±0.52 ($p=0.026$) at 2 years of follow-up. The mean pretreatment motor and bladder function scores were 0.53±0.31 and 0.74±0.44, respectively, improving to 0.48±0.33 and 0.68±0.56 at follow-up but without statistical significance. There was no significant loss of function, which is expected in radical sacral resections. In conclusion, primary sacral tumors require a multidisciplinary approach and management for optimal outcomes. A stand-alone posterior approach can be employed to treat most sacral lesions. *En-bloc* wide resection is the optimal treatment for primary malignant and aggressive benign tumors. Preservation of at least one functional S2 nerve root is imperative to preserve bowel and bladder function.

Keywords: Sacral tumors; Sacrectomy; Posterior approach; Literature review

Introduction

Primary tumors of the sacrum are very rare entities, con-

stituting approximately 1%–7% of primary spinal tumors, which in turn constitute only a mere 10% of primary bone tumors [1]. Low-grade primary tumors are often diag-

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nosed late, owing to their clinical resemblance to degenerative lumbar spine disorders, while high-grade tumors might have an early and a more dramatic presentation [1,2]. Primary sacral tumors can be benign or malignant and, based on their origin, can be predominantly grouped as primary osseous or primary neurogenic. The most common malignant sacral lesions are chordomas, which are low-grade tumors, whereas the most common benign lesions are giant cell tumors (GCT), which are locally aggressive and potentially recurrent [2].

The management of these tumors is based on numerous factors, such as tumor pathology, the upper limit of the tumor extent, involvement of neural structures, and distant metastasis [3]. Nevertheless, surgery remains the cornerstone for treating nonmetastatic primary sacral tumors [1-4]. The complex sacral anatomy, including the neural structures, adjacent visceral relationships, biomechanical role in load transmission, and late presentation of sacral tumors often poses a surgical challenge, making the surgery a multidisciplinary affair. These operations were once considered long, bloody, and morbid procedures; however, a multidisciplinary approach and recent advances have helped achieve significant breakthroughs in the treatment paradigm of primary sacral tumors. Aggressive sacral resections can cause varying degrees of neurological (bladder, bowel, motor, and sexual) dysfunction based on the level [1,5]. In the end, achieving good disease control and minimizing neurological complications are paramount in reaching positive treatment outcomes.

In our study, we present a group of 15 patients with primary sacral tumors that include bony tumors, neural tumors, and sarcomas, which were treated accordingly. Based on a literature review and our experience, we propose a protocol for managing primary sacral tumors entailing a multidisciplinary approach.

Single Institution Experience of Primary Sacral Tumors

We studied the case records of patients with primary sacral tumors treated between 2011 and 2020 in Sakra World Hospital, Bengaluru, India which is a tertiary spine care center, a tertiary spine care center. We included case records only of patients with a minimum 2-year follow-up. Patients with sacral lesions that were ultimately infections and metastatic lesions were excluded, given these cases were beyond the scope of our study. Ultimately, 15

patients were included, and their medical records, radiological images, and pathology reports were retrospectively reviewed. Given that the study was conducted using only the patients' in-patient and follow-up records, ethical approval for the study was waived.

All of our patients had undergone a uniform management protocol starting with a thorough neurological and clinical (including per rectal) examination, which was followed by X-rays, computed tomography (CT) and magnetic resonance imaging (MRI) to assess soft tissue extension. Positron emission tomography (PET) CT was also performed to search for distant metastasis when necessary. A percutaneous CT-guided biopsy of the lesion was performed by our interventional radiology team with the patient under local anesthesia to obtain a histological diagnosis. After the investigations had been concluded, the management was planned based on (1) the clinical presentation, especially neurological dysfunction; (2) the tumor pathology, as per the histopathological examination; (3) the upper extent of the lesion (MRI); (4) the soft tissue extent and local invasion of adjacent structures (MRI); and (5) the presence of metastasis (PET). All cases were subjected to a tumor board review comprising the treating surgeon, radiologist, pathologist, medical oncologist, plastic surgeon, urologist, and radiation oncologist for the decision-making process regarding treatment. Radiotherapy or chemotherapy was administered for inoperable advanced malignant lesions.

Clear surgical principles were considered for all cases, which included (1) an all-posterior approach; (2) sacrectomy with wide excision to achieve clear margins to minimize recurrence; (3) preservation of S1 and sacroiliac joint integrity to prevent lumbopelvic fixation, if possible (except in one case in which S1 was removed to achieve wide excision); (4) preservation of at least one S2 nerve root to preserve bladder function; and (5) plastic reconstruction with advancement flaps when necessary. Neural tumors were treated by laminectomy and lesion excision. Regardless of the sacrectomy level, neural dissection was started at L5-S1 through a laminectomy and traced distally to preserve at least one functional S2 nerve root (Fig. 1A-D). The functional status of the nerve roots was identified using triggered electromyography of the foot muscles and anal sphincter muscles, a component of intraoperative neural monitoring. Seeding of tumor tissue was avoided by total excision of the tumor and excision of the biopsy tract after its identification by methylene blue

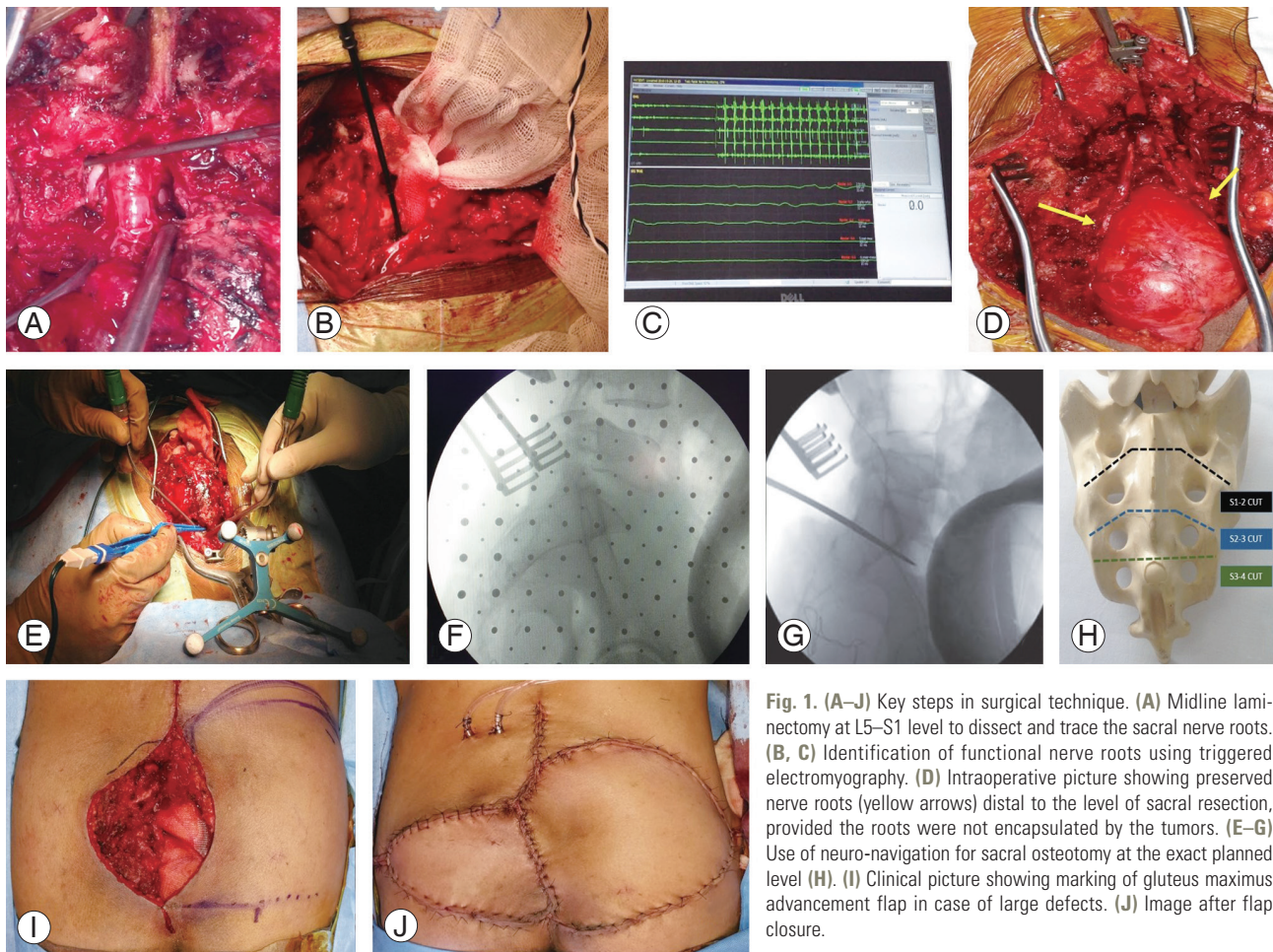


Fig. 1. (A–J) Key steps in surgical technique. (A) Midline laminectomy at L5–S1 level to dissect and trace the sacral nerve roots. (B, C) Identification of functional nerve roots using triggered electromyography. (D) Intraoperative picture showing preserved nerve roots (yellow arrows) distal to the level of sacral resection, provided the roots were not encapsulated by the tumors. (E–G) Use of neuro-navigation for sacral osteotomy at the exact planned level (H). (I) Clinical picture showing marking of gluteus maximus advancement flap in case of large defects. (J) Image after flap closure.

injection. Preoperatively, cytoreduction was attempted in chemosensitive tumors such as Ewing's sarcoma (ES). Digital subtraction angiography (DSA) followed by a preoperative transarterial embolization (TAE) in hypervascular tumors such as GCTs was performed to reduce blood loss.

Prophylactic antibiotic prophylaxis was with 1 g intravenous ceftriaxone administered 30 minutes prior to the surgical procedure and was repeated every 4 hours if necessary. Intraoperative blood loss was reduced by the usage of tranexamic acid infusion in patients who could not tolerate significant blood loss. Neuro-navigation was utilized to perform an accurate sacral resection at the planned level (Fig. 1E–H). The plastic surgery team was involved in cases of large skin or soft tissue defects to plan and perform an appropriate flap cover (Fig. 1I, J). Postoperatively, the same antibiotic was continued intravenously until the day of surgical site drain removal after wound inspection.

Follow-up records of patients at 1 month, 6 months, 1 year, and 2 years were reviewed along with their respective radiological images. Treatment outcomes considered in our study were (1) functional outcome (pain-related disability and neurological function) before and after treatment; and (2) disease control (recurrence and metastasis) after completion of treatment. The Visual Analog Scale was used to assess pain-related disability. The assessment of neurological function, including motor, bladder, and bowel function scoring, was as adapted from Biagini et al. [6] and is shown in Table 1.

The continuous variables are expressed as mean±standard deviation and the categorical variables as frequencies and percentages. For comparison analysis, the Wilcoxon signed rank test was used due to the non-normal data distribution. The data were analyzed using IBM SPSS ver. 21.0 (IBM Corp., Armonk, NY, USA).

The characteristics of all 15 cases included in our study

Table 1. Classification of neurologic function after resection of the sacrum

Function	Score	Description
Bladder	0	Normal
	1	Feels stimulus to micturate and has limited continence at varying times and quantities of urine and/or has increasing postmicturition vesical residual and/or urinary loss in conditions of stress
	2	Does not feel stimulus to micturate and/or is completely incontinent
Bowel	0	Normal
	1	Feels stimulus to defecate and is incontinent when feces are soft or under stress
	2	Does not feel stimulus to defecate and/or is completely incontinent
Motor	0	Normal or mild deficit not requiring the help of external support for motion and common activities
	1	Deficits requiring the help of external support for walking and common activities
	2	Deficits that make walking impossible

Adapted from Biagini et al. *Chir Organi Mov* 1997;82:357-72 [6].

are shown in Table 2. A total of 15 patients with primary sacral tumors were reviewed, of which 10 were men and five were women. Two patients had extracompartmental recurrent chordomas after prior surgery elsewhere. All 15 patients had back pain as a presenting symptom; six had radicular symptoms or signs; five had bladder and bowel dysfunction. The most common diagnosis was chordoma (n=7). The proximal extent of lesions was up to S1 in four patients. However, only two were surgically treated. A total of 11 patients underwent surgery, including eight partial sacrectomies (without any instrumentation), one total sacrectomy (GCT involving S1) with lumbopelvic fixation and two patients with neural tumors treated with laminectomy and tumor excision. The average duration of surgery was 6.5 hours, with a mean blood loss of approximately 900 mL. The patient with a GCT underwent wide excision after TAE considering the aggressive nature and potential recurrence. One patient with ES received neoadjuvant chemotherapy (NAC) and underwent partial sacrectomy after cytoreduction. Four patients were treated with chemotherapy (one non-Hodgkin lymphoma; one adenocarcinoma; chemotherapy with radiotherapy in one case of inoperable ES; and NAC in one operable ES). Two inoperable patients underwent radiotherapy, which included one recurrent chordoma and one ES. One patient with mucinous cystadenoma had to undergo an additional anterior procedure and colostomy.

The mean follow-up was 29±9.8 months. Of eight patients who underwent partial sacrectomy (without additional stabilization), seven patients had no stability-related complications. One patient in whom the sacral cut was taken through the S1 body had an S1 stress fracture in

follow-up and could be managed conservatively. Only one patient (out of five) who had preoperative bladder and bowel dysfunction regained reasonable function post-surgery, whereas the remaining four patients had no recovery and were taught self-intermittent catheterization. None (n=11) of the patients who had intact bowel and bladder function preoperatively worsened post-treatment. Two patients had postoperative surgical site infection, among which one patient needed debridement. One patient with recurrent chordoma had a second recurrence after partial sacrectomy surgery. Two patients (one with adenocarcinoma and one with mucinous cystadenocarcinoma) had a distant spread at the last follow-up and was followed up with chemotherapy. One patient with ES (treated with NAC and partial sacrectomy) developed pneumonia following a compromised immune status and ultimately succumbed to systemic sepsis. The results of our study have been summarized (Tables 3, 4).

Literature Review

Primary sacral tumors are rare, accounting for approximately 1%–7% of primary spinal tumors [1]. The majority of sacral tumors are metastatic lesions from breast, prostate, lung, or colon carcinomas [1], and their therapeutic approach differs completely from primary tumors [2]. Primary tumors can be classified based on their origin as osseous, neural (e.g., schwannoma), notochordal (e.g., chordoma), primitive neuroectodermal (PNET) (e.g., ES), hematopoietic (e.g., lymphoma, myeloma), and tumor-like (e.g., aneurysmal bone cyst) [2]. They can also be grouped into benign and malignant tumors. The most common

Table 2. Characteristics of all cases included in our study along with their treatment and outcome

No.	Age (yr)/sex	Primary/recurrent	Pre-treatment bowel/bladder function	Pathological diagnosis	Extent of lesion	Surgery done (level of sacrectomy)	Radiotherapy/chemotherapy	Follow-up (yr)	Post-treatment bowel/bladder function	Local recurrence	Distant spread
1	19/M	Primary	Intact	Non Hodgkins lymphoma	S2-4 Nil		Chemotherapy	6	Intact	NA	No
2	36/M	Primary	Incontinent	Chordoma	S2-5	Partial sacrectomy (S1-2)	Nil	5	Incontinent	No	No
3	35/M	Primary	Incontinent	Chordoma	S2-5	Partial sacrectomy (S1-2)	Nil	5	Partial regain of control	No	No
4	59/M	Primary	Intact	Adenosarcoma	S1-3 Nil		Chemotherapy	1	Intact	NA	Yes
5	63/M	Recurrent	Intact	Chordoma	S3-5 Nil		Radiotherapy	2	Intact	NA	No
6	42/F	Primary	Incontinent	Chordoma	S2-5	Partial sacrectomy (S1-2)	Nil	1	Incontinent	No	No
7	68/F	Primary	Intact	Chordoma	S4-5	Partial sacrectomy (S3-4)	Nil	7	Intact	No	No
8	24/M	Primary	Intact	Ewing's sarcoma	S1-3 Nil		Chemotherapy + radiotherapy	1	Intact	NA	No
9	53/F	Primary	Intact	Neurofibroma	S2-3	S2 hemi-laminectomy and tumor excision	Nil	3	Intact	No	No
10	42/M	Primary	Intact	Schwannoma	S2-5	S2-4 laminectomy and excision of lesion	Nil	4	Intact	No	No
11	19/F	Primary	Intact	Giant cell tumor	S1-4	Complete sacrectomy and lumbo-pelvic stabilization after TAE	Nil	2	Intact	No	No
12	11/M	Primary	Intact	Ewing's sarcoma	S2-4	Partial sacrectomy	Neoadjuvant + adjuvant chemotherapy	1	Intact	No	No
13	71/F	Primary	Intact	Chordoma	S3-5	Partial sacrectomy (S2-3)	Nil	6	Intact	No	No
14	45/M	Recurrent	Incontinent	Chordoma	S1-2	Partial sacrectomy (S1-2)	Radiotherapy	1	Incontinent	Yes	No
15	65/M	Primary	Incontinent	Mucinous cystadenoma	S2-4	Partial sacrectomy+colostomy	Chemotherapy	1	Incontinent	No	Yes

M, male; F, female; NA, not available; TAE, transarterial embolization.

malignant tumor of the sacrum is chordoma, followed by chondrosarcoma, myeloma, malignant nerve sheath tumors, ES, and lymphoma [2,5]. The common benign tumors include GCTs, schwannomas, and aneurysmal bone cysts. However not all benign tumors are low-grade lesions; e.g., GCTs, which can be locally aggressive. Hence, more usefully, the primary tumors can be grouped as (1) low-grade tumors (chordoma, chondrosarcoma), which are slow-growing with nonspecific clinical symptoms; and (2) high-grade tumors, such as ES, osteosarcoma, and GCT, which can have a more aggressive clinical presentation and recurrence [3]. As reported in the literature, chordoma was the most common primary tumor in our study (seven cases), followed by primary neural tumors and ES (two cases each).

MRI followed by CT is the most informative radiological investigation, given it provides a host of information, such as the possible epicenter of the lesion, presence of soft tissue matrix, probable tumor pathology, and most importantly the extent of tumor, which primarily dictates the level of sacral resection to be planned in cases need-

ing a wide excision [1,7]. PET scan could be an important prognostication tool to opt between a curative treatment or a palliative treatment based on distant metastasis [7]. Given the pathology is the most decisive factor determining the prognosis and treatment of sacral lesions, CT-guided biopsy is a critical component in their diagnostic algorithm. Safaee et al. [8] reported a positive biopsy in up to 95% of sacral lesions, with correlation rates approaching 100%. Excision of the biopsy tract following CT-guided biopsy is less likely to cause tumor contamination of the surgical plane compared with open biopsy. We used methylene blue injection for the identification of the biopsy tract in our patients. Primary sacral tumors are treated principally based on four factors: (1) tumor pathology (based on biopsy); (2) the proximal extent of the tumor; (3) neurological dysfunction; and (4) contiguous spread and distant metastasis. The treatment modalities are chemotherapy, radiotherapy, and surgery, with surgery as the cornerstone of curative treatment in nonmetastatic primary tumors [2-4].

Various surgical approaches have been employed for

Table 3. Functional outcome of patients comparing pain and neurological status before and after treatment

Variable	Pre-treatment	2-Year follow-up	p-value
Visual Analog Scale score	7.8 (5–10)	3.7 (2–7)	0.026*
Motor function (0–2) ^{a)}	0.53±0.31	0.48±0.33	0.125
Bladder function (0–2) ^{a)}	0.74±0.44	0.68±0.56	0.059
Bowel function (0–2) ^{a)}	0.81±0.47	0.63±0.52	0.026*

Values are presented as mean (range) or mean±standard deviation.

*p<0.05 (statistically significant). ^{a)}Classification of neurological function as adapted from Biagini et al. [6].

Table 4. Complications and disease control at 2-year follow-up

Variable	No	Yes
Postoperative sacral stress fracture (n=11)	10 (91)	1 (9)
Surgical site infection (n=11)	9 (82)	2 (18)
Local recurrence (n=15)	13 (87)	2 (13)
Distant metastasis (n=15)	13 (87)	2 (13)
Survival at 2 years (n=15)	1 (7)	14 (93)

Values are presented as number (%).

Table 5. Review of the literature of various studies employing posterior approach for sacrectomy

Study	No. of cases	Indications	Modifications
Gennari et al. [12] (1987)	8	Lesions below S2	Not specified
Waisman et al. [13] (1997)	5	Sacral tumors without local visceral spread	Usage of mersilene mesh to prevent herniation
McLoughlin et al. [15] (2008)	1	Not specified	<i>En-bloc</i> total sacrectomy done by lifting the sacrum from proximal to distal and anterior dissection
Asavamongkolkul et al. [14] (2012)	21	For all sacral lesions as high as L5	Large amount of gauze packing anterior to sacrum for blunt dissection and protection during osteotomy
Clarke et al. [9] (2012)	36	All tumors not extending beyond the lumbosacral junction or invading the bowel	Not specified
Zang et al. [16] (2015)	10	High sacral lesions without local anterior invasion	Use of wire saw for ilium cutting from inside to outside
Our study	11	All sacral lesions (as high as L5–S1) with no local invasion of anterior structures	Division of ligaments, lifting of sacrum <i>en-bloc</i> from distal to proximal, anterior blunt dissection

sacral resections. Conventionally, total and high sacrectomies have been performed via combined anterior-posterior approaches [9-11]. The stand-alone posterior approach was initially advocated for distal sacral resections [12] and was later extended to *en-bloc* total sacrectomy with a few modifications by many authors [9,13-16]. The various literature discussing the applicability of an all-posterior approach has been listed (Table 5). The feasibility of an all-posterior surgical approach for sacrectomies is based on the anterior extent, proximal limit, and accessibility of the tumor [9,16]. An all-posterior approach has been shown to have lesser morbidity and blood loss compared with the combined approach [9,15,16]. The surgeries we performed in all the cases with no anterior visceral involvement employed only a posterior approach. The anterior extent of the tumor was preoperatively decided based on per rectal clinical examination and by MRI. We could remove the tumor *en bloc* even in lesions involved up to S1 employing a stand-alone posterior approach, starting the dissection caudally by division of ligaments and proximal blunt dissection deep to the presacral fascia and mobilization of pelvic structures. One patient in whom the primary lesion was a mucinous cyst adenocarcinoma contiguously involving the colon underwent an additional anterior bowel

resection procedure along with a colostomy.

Wide excision with clear margins is the preferred treatment in aggressive benign and malignant tumors. Based on the level of resection, the surgeries have been termed as low, mid, high, hemi, and total sacral resections with predictable degrees of bowel, bladder, and sexual dysfunction [1,6,17]. Total sacral resections (involving the S1 body) could also result in lumbopelvic instability and warrant lumbopelvic fixation [18]. An optimal outcome could be obtained by minimizing disease recurrence by balancing a wide excision, concurrently preserving at least the S1 body to maintain stability and a few functional sacral roots contributing to the pudendal nerve to preserve bowel and bladder function. The necessary sacral roots to be preserved for good bowel and bladder function has been debated in the literature (Table 6). Various authors have recommended the preservation of the S3 root [19-26], with few among them reporting good function with preservation of one or more S2 nerve roots [12,17,27-29]. In our patients, irrespective of how high the sacrectomy, functional sacral roots (at least one S2 root) were preserved, provided they were not encased in the tumor seven of 11 surgical patients had intact bowel and bladder function before surgery; as a result of pre-

Table 6. Review of the literature discussing the neurological function post sacral resections and surgical recommendations

Study	No. of cases	Results	Recommendation of sacral root preservation
Gunterberg [23] (1976)	10	Intactness of at least one S3 preserves normal bladder and anorectal function	Unilateral S3
Stener et al. [27] (1978)	5	Functional urinary and fecal continence is possible if ≥one S-2 root can be preserved	Unilateral S2
Andreoli et al. [29] (1986)	2	Retaining one S-2 root is sufficient for the maintenance of physiologic continence of bowel & bladder	Unilateral S2
Sung et al. [19] (1987)	54	Excision below S3 does not affect bowel or bladder function	Unilateral S3
Gennari et al. [12] (1987)	8	When both S2 roots were preserved, sphincter problems were mild and reversible	Bilateral S2
Fujimura et al. [28] (1994)	8	Bilateral preservation of S2 nerve roots necessary to maintain bowel, bladder & sexual function	Bilateral S2
Cheng et al. [21] (1999)	23	Preservation of B/L S3 roots helped preserve 100% bowel & bladder function	Bilateral S3
Bergh et al. [24] (2000)	30	S3 nerves appear the most critical determinant of bowel, bladder function	S3 preservation
Todd et al. [22] (2002)	53	Preservation of B/L S3 roots helped preserve 100% bowel function, 69% bladder function	Unilateral S3
Fourney et al. [17] (2005)	78	Middle sacrectomy has lesser bladder dysfunction than high sacrectomy	Bilateral S2
Guo et al. [25] (2005)	50	Bowel incontinence incidence was 37.5% after unilateral S-3 nerve root resection and 75% after bilateral resections	Bilateral S3
Moran et al. [20] (2015)	73	Better bowel, bladder function with low sacrectomy (below S3)	Bilateral S3
Zoccali et al. [26] (2016)	Review	Better bowel and bladder function when S3 nerve root preserved	Unilateral S3
Our study	11+Review	Preservation of one functional (by triggered electromyography testing) nerve root preserves reasonable bowel & bladder function	Unilateral S2

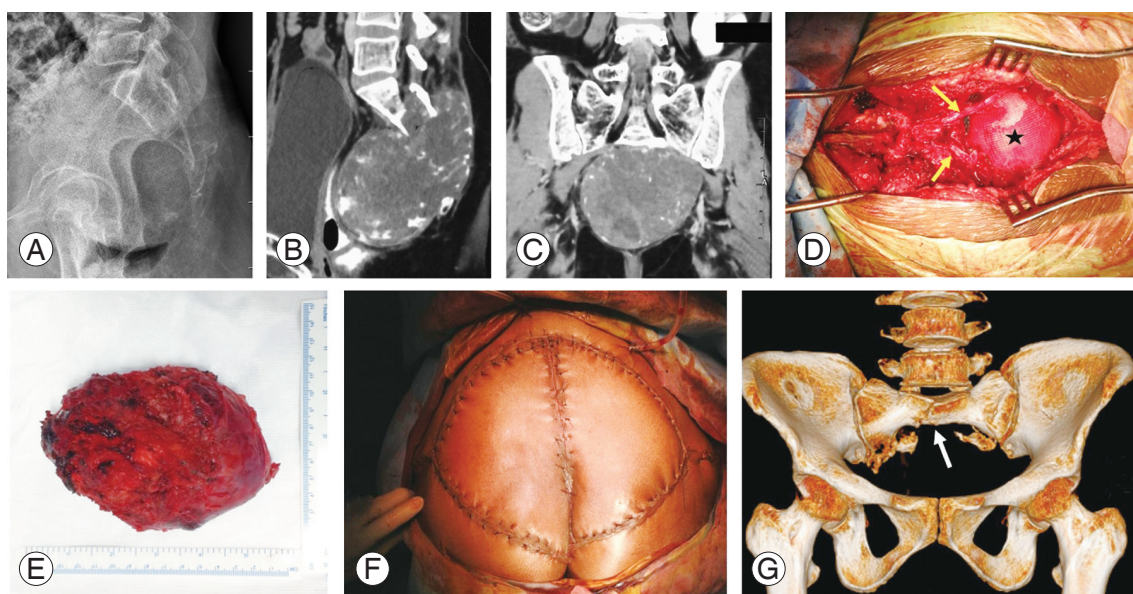


Fig. 2. (A–G) Large sacral chordoma extending up to S1–S2. **(A)** Preoperative X-ray showing destructive lesion involving the sacrum. **(B, C)** Preoperative sagittal and coronal computed tomography (CT) images showing the extent of lesion up to S1–S2 with a large encapsulated pre-sacral component. **(D)** Intraoperative image showing the preserved S2 nerve roots (yellow arrows) and large defect following sacrectomy reconstructed by mesh (black star). **(E)** Clinical image of specimen resected by wide excision. **(F)** Plastic reconstruction and closure of large defect by gluteus maximus advancement flap. **(G)** Follow-up CT scan at 18 months showing a stress fracture involving S1 (white arrow), which was conservatively managed.

serving at least one S2 root, none of them lost bowel or bladder function postoperatively. In our study, of eight patients who underwent partial sacrectomy, one patient (chordoma involved up to S1–S2) in whom the resection was performed through the S1 body had an insufficiency fracture of redundant S1 body (Fig. 2), whereas all seven remaining patients had no stability-related pain. The use of neuro-navigation helped in making precise osteotomy cuts where desired without destabilizing the sacroiliac joints, in addition to reducing the operating time and the amount of radiation exposure to the operating room personnel. One patient who underwent a total sacrectomy had additional lumbopelvic fixation.

Preoperative TAE of the feeding vessel after a DSA is known to reduce intraoperative blood loss and surgical time in all vascular sacral tumors [30]. Its effectiveness in treating chordoma and GCT are well reported in the literature [31,32]. We used preoperative TAE in one patient with a GCT extending up to S1, followed by total sacrectomy and reconstruction, which helped reduce intraoperative blood loss to less than 1,000 mL.

Neural monitoring is a useful adjunct in preserving neurological function by the identification of functional sacral roots. The activity of the anal sphincter can be ex-

trapolated to the activity of the external urethral sphincter because both are innervated by the pudendal nerve, with a root value of S2, S3, and S4. Surgeon-triggered stimulation of these nerve roots can be picked up by sphincter electrodes during surgery [33], indicating the functional status of the nerve roots and warranting their preservation. Large sacral wound defects might require myocutaneous flaps for good healing. Bilateral gluteal musculocutaneous advancement (BGMA) flaps are sufficient in primary tumors treated without radiotherapy and preserved gluteal vessels, whereas trans-pelvic ventral rectus abdominal myocutaneous (VRAM) flaps need to be considered otherwise. Free flaps are considered if VRAM flaps have been scarred by previous abdominal surgeries [34]. Five of our patients had significant dead spaces after sacral resections. Two of them (one GCT treated with total sacrectomy and one patient with a large recurrent chordoma with resection up to S1–S2) were managed by BGMA flaps performed by our plastic surgery team (Fig. 1I, J). Two were managed by a V-Y myocutaneous advancement flap. These four patients had good healing without any wound complications. One patient with a partial sacrectomy in which the dead space was not managed by a plastic procedure developed a deep wound infection and needed

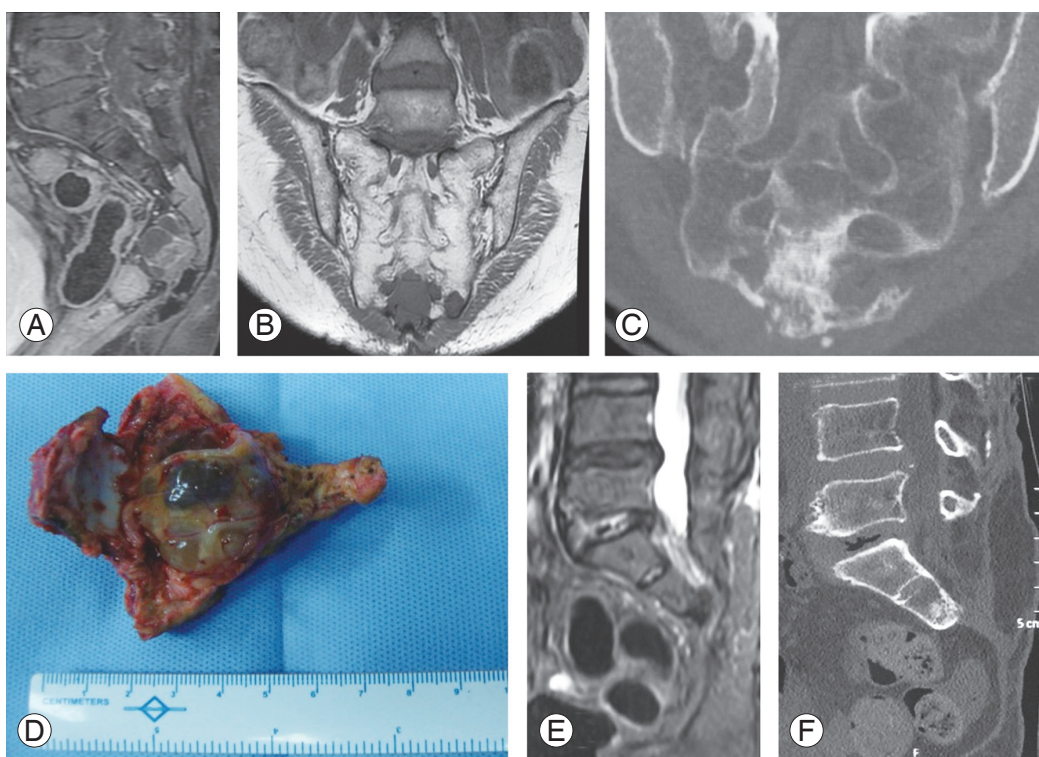


Fig. 3. (A–F) Sacral chordoma. (A, B) Preoperative sagittal and coronal magnetic resonance imaging (MRI) images showing the extent of lesion up to S3. (C) Preoperative computed tomography (CT) scan. (D) Clinical image of specimen resected by wide excision. (E, F) Follow-up MRI and CT scan at 5-year follow-up showing no disease recurrence.

wound debridement and subsequent treatment with intravenous antibiotics.

Chordomas are rare low-grade, slow-growing malignant bone tumors arising from the midline skeleton, most commonly from the sacrum. They are the most common primary sacral tumors and usually present after the fifth decade owing to their slow-growing nature and nonspecific clinical symptoms [35]. The extracompartmental extension at the time of presentation is the main predictor of the patient's life expectancy [36]. Chordomas are known to have poor sensitivity to chemotherapy and radiotherapy, making surgical resection the gold standard treatment [35]. Surgical margins are defined according to the Enneking classification system as intralesional, marginal, wide, and radical. A meta-analysis by Yu et al. [35] concluded that the surgical margin is the most decisive factor in achieving good disease control in sacral chordomas. Recurrence rates were lower in patients with wide surgical margins in contrast to inadequate (intralesional and marginal excision) margins [35,37]. Most of the studies report a median overall survival rate of >5 years, and local recurrence and metastasis contributes to morbidity and mortality in these

patients [35]. DSA followed by a preoperative TAE helps reduce blood loss and the time needed for surgical resection [31]. Seven of 17 patients in our study had chordomas, constituting the majority of our patients. Five cases were primary chordomas and were treated with *en bloc* resection with Enneking appropriate wide margins. An example of a surgically treated primary chordoma is illustrated (Fig. 3). There has been no recurrence to date (follow-up 1.5 to 5 years). Two patients had recurrent chordomas, one of who was treated with partial sacrectomy. The margins were inadequate on histopathological examination, and the patient had a recurrence at 2-year follow-up despite radiotherapy (Fig. 4). The other recurrent case was inoperable, with a large extracompartmental lesion and intrapelvic extension, and was treated with radiotherapy. More recent advents, such as carbon ion radiotherapy, proton beam therapy, and molecular targeted therapy agents (e.g., imatinib, temsirolimus) have been promising in treating recurrent inoperable chordomas [38–40].

Schwannoma and neurofibroma are the two common benign tumors arising from intrasacral neural elements and are sometimes associated with giant presacral com-

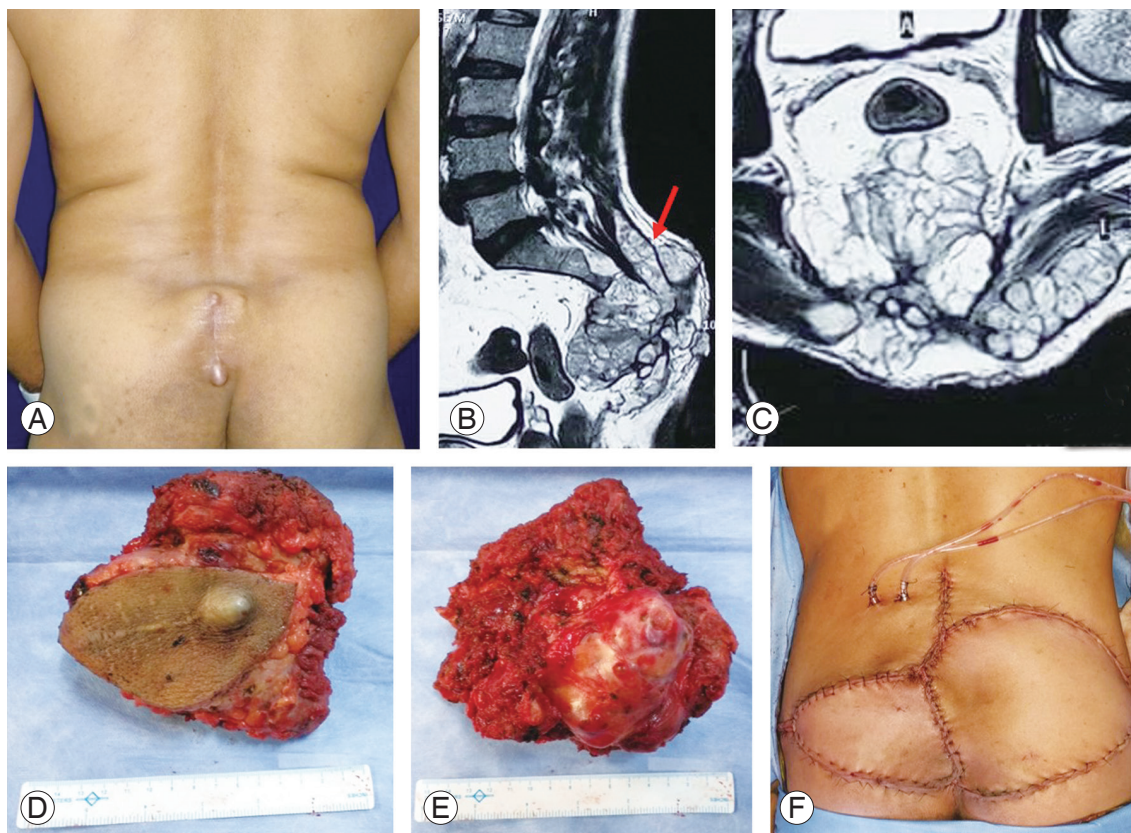


Fig. 4. (A–F) Recurrent sacral chordoma. (A) Preoperative clinical image showing invasion of the skin by local extension of recurrent tumor. (B, C) Preoperative sagittal and coronal magnetic resonance images showing recurrent sacral lesion involving up to S2 with pre-sacral and sub-cutaneous extension (red arrow). (D, E) Clinical image showing removed tumor *en-bloc* with the skin. (F) Intraoperative image showing closure of large skin defect by gluteus maximus advancement flap.

ponents in addition to intrasacral component. Despite the evolution of treatment strategies, surgical resection remains the optimal treatment [41]. A laminectomy is often sufficient to remove intrasacral tumors. However large presacral lesions might require partial sacral amputations: a blunt dissection of the tumor from the rectum before removing them. Owing to their benign nature, these neural tumors can be managed by marginal excision, given they have a distinct pseudo-capsule. We had two such patients with no presacral components who were managed by laminectomy/hemilaminectomy and marginal excision, taking care to preserve the sacral nerve roots using intraoperative neural monitoring. One of them is illustrated (Fig. 5). The local recurrence rate varies between 7% and 21%, and no treatment might be necessary unless symptomatic [41]. The neurological symptoms resolved completely in both of our patients, with no disease recurrence at follow-up.

ES of the sacrum is a malignant tumor belonging to a group of small, round, blue cell neoplasms, differentiated

from the other PNETs by neural and immune histochemical markers. They usually present in the first 2 decades of life and have an aggressive course. They are highly chemosensitive, making multiagent chemotherapy (neoadjuvant and adjuvant) the cornerstone of their treatment. Local disease control can be achieved by wide *en bloc* excision, radiotherapy, or both [42]. The superiority of either surgery or radiotherapy is still debated. Unlike ES of the extremities, surgery for sacral ES is complex because of the anatomy, with potential neurological compromise and inadequate margins. Although many authors advocate wide resection of the tumor as the only way of curing the disease locally [42,43], surgery with inadequate margins is associated with a high recurrence rate and a poorer prognosis. Following ES resection, chemotherapy-induced tumor necrosis is an important prognostic predictive factor to evaluate the efficacy of chemotherapy. With recent advances, radiotherapy provides good local control for ES, justifying its evaluation as an alternative to surgical treat-

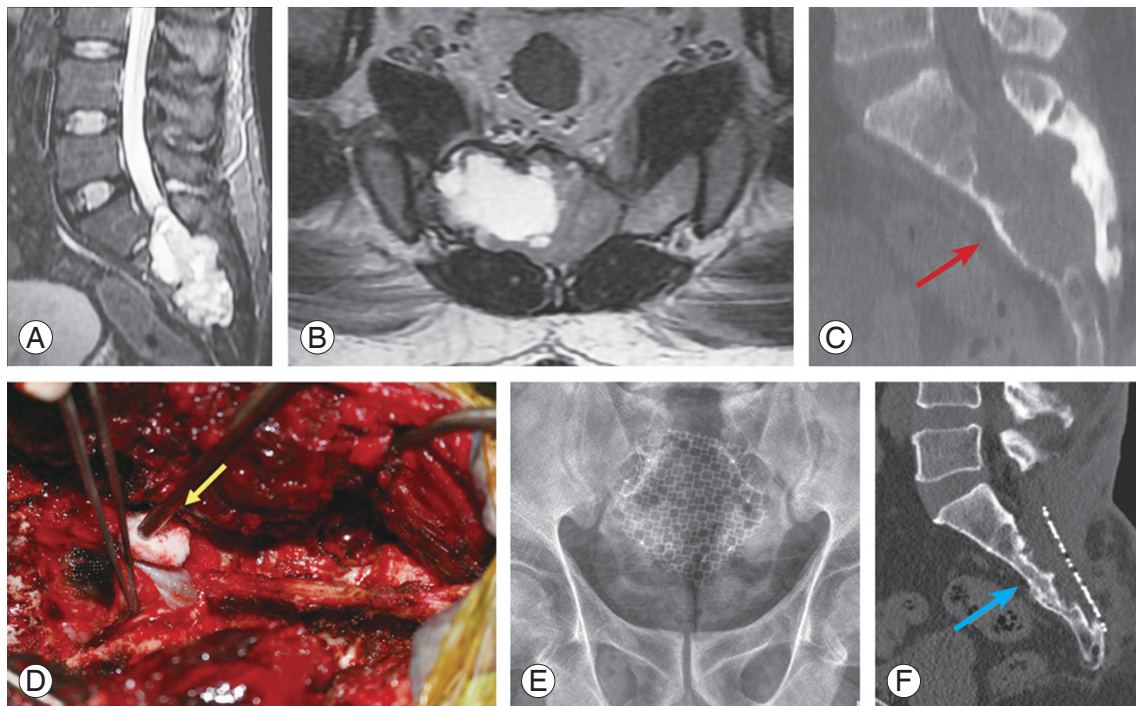


Fig. 5. (A–F) Sacral schwannoma. (A, B) Preoperative magnetic resonance images intra-sacral lesion with no pre-sacral extension. (C) Preoperative computed tomography (CT) image showing the scalloping (red arrow) of the anterior sacral cortex by the lesion. (D) Intraoperative image showing removal of tumor (yellow arrow) arising from right S3 root following laminectomy. (E) Postoperative X-ray showing laminectomy defect reconstructed using a titanium mesh. (F) Follow-up CT scan at 2 years showing re-ossification of the anterior sacral wall (blue arrow), and no recurrence.

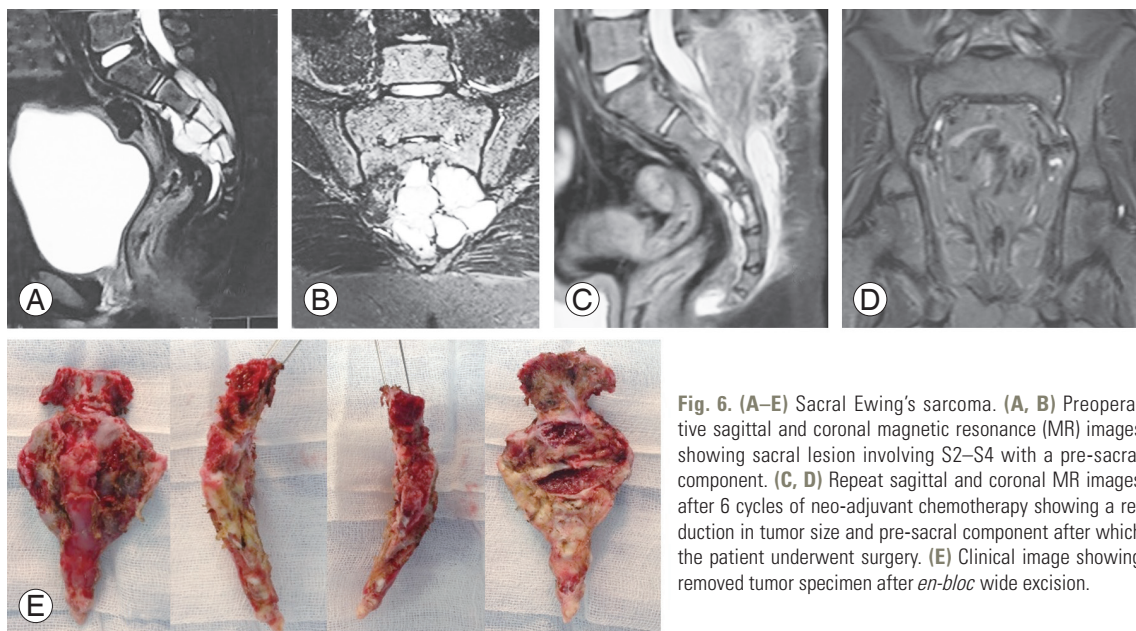


Fig. 6. (A–E) Sacral Ewing's sarcoma. (A, B) Preoperative sagittal and coronal magnetic resonance (MR) images showing sacral lesion involving S2–S4 with a pre-sacral component. (C, D) Repeat sagittal and coronal MR images after 6 cycles of neo-adjuvant chemotherapy showing a reduction in tumor size and pre-sacral component after which the patient underwent surgery. (E) Clinical image showing removed tumor specimen after *en-bloc* wide excision.

ment [42,44,45]. Hence, radiotherapy has a clear advantage over surgery in locally advanced tumors. We had two cases of sacral ES. One patient was an 11-year-old presenting with a large ES extending from S2 to S5, with bowel

and bladder incontinence. He underwent six cycles of NAC (VAC/IE [vincristine+doxorubicin+cyclophosphamide alternating with ifosfamide+etoposide] regimen) over a 12-week period, which showed drastic shrinkage of the

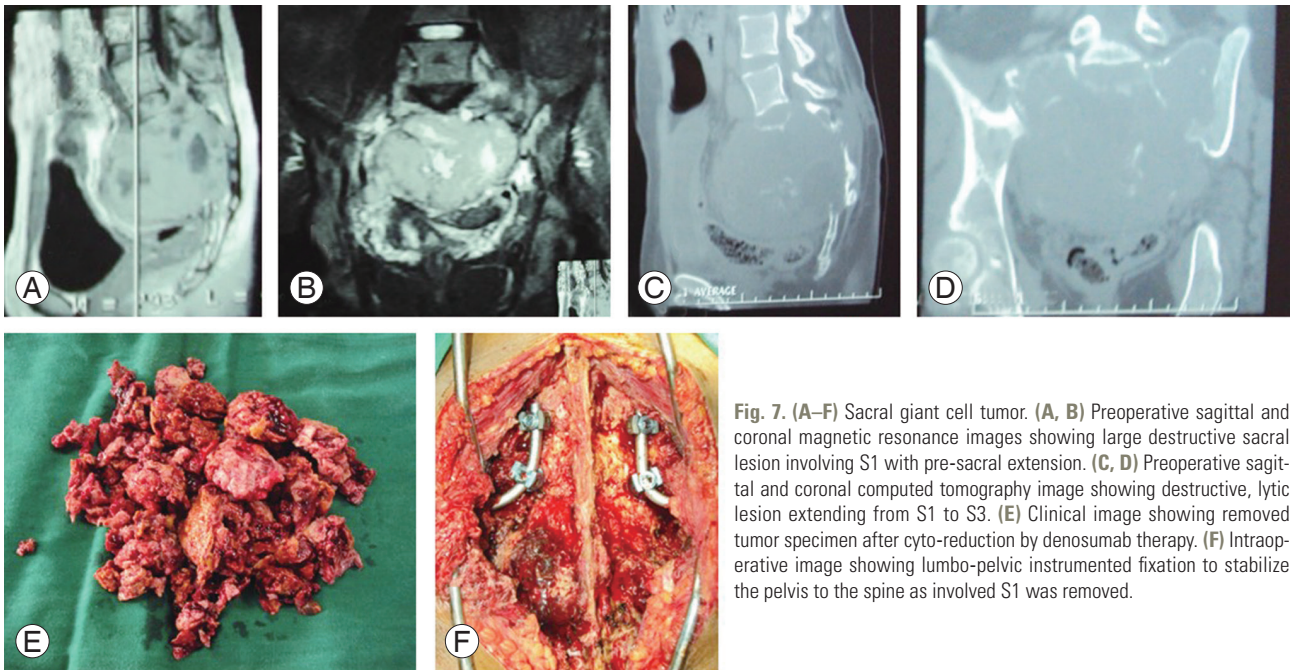


Fig. 7. (A–F) Sacral giant cell tumor. **(A, B)** Preoperative sagittal and coronal magnetic resonance images showing large destructive sacral lesion involving S1 with pre-sacral extension. **(C, D)** Preoperative sagittal and coronal computed tomography image showing destructive, lytic lesion extending from S1 to S3. **(E)** Clinical image showing removed tumor specimen after cyto-reduction by denosumab therapy. **(F)** Intraoperative image showing lumbo-pelvic instrumented fixation to stabilize the pelvis to the spine as involved S1 was removed.

tumor (by 80%). He later underwent partial sacrectomy and adjuvant chemotherapy (Fig. 6). He regained bowel and bladder control and showed no signs of recurrence at the 1-year follow-up. Unfortunately, he succumbed to pulmonary infection and systemic sepsis later. The other patient with ES had a locally advanced tumor involving pelvic organs and was treated with chemotherapy and radiotherapy. He had no signs of local disease progression at 2-year follow-up.

GCT is the most common benign sacral tumor in the literature [2,5,32]. Despite being termed benign, they possess locally aggressive characteristics and can metastasize to the lung [46]. Treatment options include surgery (intralesional curettage; marginal excision; wide excision), TAE, and radiotherapy. Intralesional curettage and marginal excision in high sacral GCTs might help preserve sacroiliac joint stability but are associated with higher recurrence rates and blood loss [32,46]. Adjuvants such as bone cement and liquid nitrogen have been proposed to reduce recurrence with limited results [47]. Wide excision with clear margins offers the lowest recurrence rates and less blood loss at the expense of stability, given the GCTs are often large and their wide resections involve the sacroiliac joint [48]. Preoperative TAE following a DSA helps in reducing blood loss and surgical time by reducing tumor vascularity [30]. Given our patient had a lesion extending up to S1, a total sacrectomy was performed after TAE, and

lumbopelvic mesh reconstruction and stabilization were necessary (Fig. 7). Isolated radiotherapy in inoperable cases has been reported, with a high recurrence rate and risk of developing radiation-induced sarcomas [49]. Serial TAE has also been shown to cause shrinkage of large GCTs, with good pain relief and disease control, and can be useful for inoperable cases [50].

One patient in our study had a mucinous cystadenocarcinoma, which is a rare malignant tumor typically arising from the ovary, colon, or pancreas. They can also arise from the soft tissue surrounding the rectum with contiguous involvement of the sacrum [51]. Such isolated sacral lesions with no distant metastasis can still be surgically resected [52]. Our patient presented with urinary dysfunction and back pain and was diagnosed based on MRI showing a large presacral mass with a destructive lesion in the sacrum, and needle biopsy confirmed the diagnosis. The metastatic workup was negative. The patient underwent partial sacrectomy, with an added anterior procedure for removal of the rectal lesion and a colostomy. Postoperative chemotherapy and radiotherapy were also administered. The patient had no recurrence at 18-month follow-up.

Primary sacral lymphomas are very rare tumors presenting as destructive lesions with or without neural involvement. These are predominantly non-Hodgkin's B-cell lymphomas (NHBL). They are highly chemo- and ra-

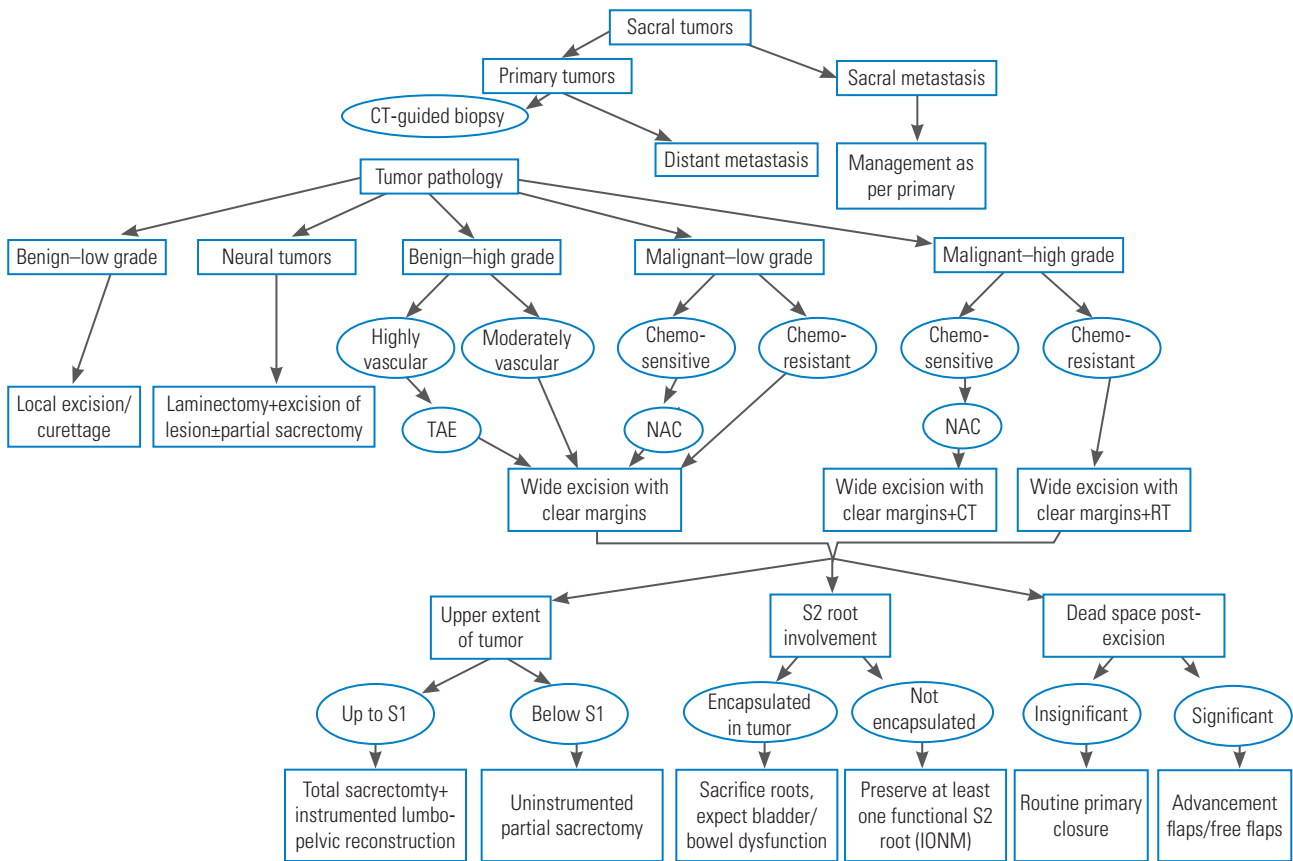


Fig. 8. Stepwise approach to manage primary sacral tumors based on our experience and literature review. CT, computed tomography; TAE, transarterial embolization; NAC, neoadjuvant chemotherapy; IONM, intraoperative neural monitoring.

diosensitive and rarely need surgical removal in instances such as severe neural dysfunction. Chemotherapy employing the chemotherapy regimen has been the standard of care. The recent advent of monoclonal antibodies such as rituximab demonstrates good disease-free survival in up to 85% of patients, especially when combined with field radiotherapy [53]. One of our sacral lesions turned out to be an NHBL and was treated by chemotherapy and radiotherapy with complete resolution of symptoms.

Based on our institutional experience and with input from the literary review, we have formulated a management algorithm to guide the investigation and treatment of primary sacral tumors (Fig. 8).

Limitations

We do recognize the limitations of our study. Being a retrospective study with a relatively small sample size, a prospective study on a larger group of patients is needed to generalize surgical outcomes. However, our recom-

mended approach to any primary sacral tumor was based on our experience and a detailed review of the literature.

Conclusions

Management of primary sacral tumors needs a multidisciplinary approach comprising a team of specialists to optimize patient outcome. A stand-alone posterior approach can be used to treat most of the sacral lesions extending up to S1. An *en bloc* wide resection is the optimal treatment of primary malignant and aggressive benign tumors such as GCTs. Preservation of the S1 body and sacroiliac joint integrity is paramount to avoid lumbopelvic fixation. Preservation of at least one functional S2 nerve root is imperative to preserve bowel and bladder function.

Conflict of Interest

No potential conflict of interest relevant to this article was reported.

Author Contributions

Venugopal Sarath Chander: principal investigator and article writing; Ramachandran Govindasamy; data and literature inputs; Venkata Ramakrishna Tukkapuram: data aggregation and preliminary manuscript ideas; Swaroop Gopal: consultant surgeon, valuable literary inputs; and Satish Rudrappa: clinical decisions, knowledge gap resource, and principal surgeon.

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