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Review

Surgical Management of Spinal Chordomas: A Narrative Review

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Abstract: Background: Chordomas are rare malignant tumors derived from notochordal remnants, accounting for 1–4% of skeletal malignancies. These slow-growing tumors most commonly affect the sacrum, skull base, and spine, posing significant challenges due to their anatomical location near critical neural structures. Despite their low metastatic potential, chordomas have a high local recurrence rate, necessitating aggressive surgical intervention. **Methods:** A narrative review of the literature was conducted, analyzing studies on the surgical management and adjuvant radiotherapy of spinal chordomas. Electronic databases, including MEDLINE, Scopus, ProQuest, and Google Scholar, were searched using relevant keywords. The retrieved studies were reviewed to assess surgical approaches, complications, outcomes, and the role of radiation therapy in treatment. **Results:** En-bloc resection with negative margins remains the primary treatment approach for spinal chordomas, though achieving complete excision is challenging due to anatomical constraints. Advances in radiotherapy, including proton beam and carbon ion therapy, have shown promise as adjunct treatments, especially when gross total resection is not possible. However, the benefit of adjuvant radiation remains debated. Postoperative complications, such as infections, neurological deficits, and mechanical instability, highlight the need for a multidisciplinary approach in patient management. **Conclusion:** The management of spinal chordomas requires an individualized treatment approach, balancing surgical feasibility, functional preservation, and disease control. While en-bloc resection remains the gold standard, newer radiotherapy techniques may improve outcomes in select cases. A multidisciplinary team is essential to optimize prognosis and quality of life for affected patients.

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INTRODUCTION

Virchow first characterized chordoma in 1846; it is an uncommon primary tumor with malignancy that makes up 1-4% of skeletal malignancies and develops from the embryonic remains of the notochord [22,54]. Chordomas are uncommon primary cancerous sarcomas of the bone having an average lifespan of 7.7 years [47]. Given their placement along the midline of the axial skeleton and their close connection to the brainstem, cranial nerves, and spinal cord, chordomas have a poor natural course that is significantly accompanied by morbidity and death [40]. Recently, it has been discovered that chordomas can also develop from intraosseous benign notochordal cellular tumors that develop into cancer [56]. Chordoma makes up 4% of cancerous bone tumors, with an incidence of around 1 per one million individuals [22,37]. Chordomas are slow-growing tumors that have a low initial tendency for metastasis but a high probability of local relapse [22]. Around 50–60% of cases include the sacrum, 25–35% the skull base, 10% the cervical spine, and 5% the thoracolumbar spine [17]. The most common sacral malignancy is chordoma [17]. Furthermore, 3:2 is the male-to-female

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ratio [17]. Adults with chordomas often develop them between the ages of 40 to 70 [37]. Females and younger individuals are more likely to develop cranial chordomas [10].

In radiographic examinations, chordomas commonly show up as a degenerative lesion that damages the axial skeleton and is connected to a sizable soft tissue mass at the tumor's core within the vertebral body [36]. In contrast to osteosarcoma and chondrosarcoma of the vertebral body, which is more frequently detected in the appendicular bone, chordomas are malignancies of the soft tissue [36]. Typically, chordomas migrate to nearby vertebrae after initially spreading locally into the intervertebral disc space [28]. Chordomas are frequently present on computed tomography (CT) scans as an osteolytic, mixed osteolytic, and osteosclerotic bone lesion that typically contains a myxoid component [36]. Intravenous infusion of contrast mediates the visualization of chordomas [36]. Due to the more widespread use of MRI (Magnetic Resonance Imaging), chordomas are now more frequently found in patients in their 30s than in the past [17]. The symptoms are vague and include gluteal distress, perineal heaviness, irritation, or worsened pain while seated, and incomplete sciatica typically lasting over 18 months (about 1 and a half years) after the symptoms first appear [17]. Chordomas exhibit hyperintense on MRI but can vary from isointense to hypointense on T1-weighted images [36]. Gadolinium contrast may be utilized as well to see improved images of the chordoma [36].

The only treatment that offers a chance of success and extended survival is broad en-bloc resection, which includes all invading tissues and has free surgical margins [16,29,42,48,54]. The feasibility of en bloc, wide margin, and radical resection is constrained by several issues relating to spinal anatomy [55]. The cauda equina or neural foraminal level involvement of functional nerve roots, structural restrictions on removing involved osseous elements surrounding the spinal cord and thecal sac, potential adhesion of tumor to thecal sac dura, and the longitudinal musculature encompassing the entire spinal column that can become involved with tumor invasion making en bloc resection challenging are just a few general limitations that must be taken into account [1,55]. Planning is necessary for broad resection since it involves sacrificing roots, bone, muscle, skin, and/or organs [17].

By itself, primary radiation has a poor therapeutic effect on chordomas [41]. Although some studies have shown an increase in survival whenever adjuvant radiation therapy is utilized in marginal and intralesional resections, the application of adjuvant high-dose radiotherapy in spinal chordomas remains debatable [4,59,61]. A recent database analysis has shown not much benefit in cases where gross total resection (GTR) has been achieved [58]. Promising developments have been made in more accurate radiotherapy techniques, such as stereotactic radiotherapy, carbon ion treatment, and proton beam therapy [5].

This article aimed to review the literature regarding principles of surgical management, approaches, and complications of spinal chordoma surgery. Additionally, we also overviewed studies concerning the adjuvant radiation therapy given in chordoma patients.

METHODS

To find articles about patients with spinal chordomas who underwent surgical intervention as well as radiotherapy, the authors searched electronic literature. MEDLINE, Scopus, ProQuest, and Google Scholar were among the databases used. Keywords used were, “chordoma” [OR] “chordomas” [AND] “spine” [OR] “sacrum” [OR] “lumbosacral” [OR] “lumbar” [OR] “thoracic” [OR] “cervical” [AND] “surgery” [OR] “radiosurgery” [OR] “radiotherapy” [OR] “proton”, were utilized as search phrases to conduct a literature search. Additionally, the retrieved papers' references were checked for other articles.

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MANAGEMENT

Principles of Surgical Management

To comprehend why invasive surgery is frequently used to treat chordomas, it is helpful to concentrate on the sacrum, where chordomas most frequently develop [7]. Early reports on chordoma outcomes were mostly negative [7]. Without computed tomography or magnetic resonance imaging, controlling a chordoma would have been extremely challenging and impossible [7]. It is significant to highlight that for the majority of the 20th century, surgical intensive care units, or ICUs, and donations of blood were uncommon, which further complicated operative management [7]. Due to this, intralesional subtotal resections accompanied by palliative radiation dosages were frequently used to treat the majority of chordomas [26]. Over 90% of tumors were observed to recur, which is not surprising given that most tumors were only partially excised [7]. Recurrent chordomas in the sacrum can unfortunately be a troublesome issue, as their gradual growth can cause loss of bowel and bladder control, and if left untreated, they frequently erode through the skin, causing discomfort and wounds that smell bad [7]. Bertil Stener, a Swedish physician from Goteborg who advocated for the en bloc removal of the sacrum, started taking a harsher stance because patients frequently suffered for extended periods with these intractable disorders [26]. Loss of bowel, bladder, and sexual function would result from a moderate to high sacrectomy [7].

To propose such a drastic strategy, Stener must have been humbled by what he had seen in patients following intralesional resection [49]. En-bloc resection was accepted, but it soon became apparent that mortality and morbidity linked to high sacrectomy posed a grave issue [7]. However, the evidence was strong in the near term in favor of attaining negative margins, with better local control observed in patients who underwent en-bloc resection and had negative margins [30]. However, the latest information has shown that local control cannot always be assured by extensive resection alone [7]. The results of 99 patients of primary chordoma in the sacrum were discussed in a recent study from two sizable referral institutions [7]. Two problems came up. First off, only around 50% of cases were able to have extensive resection, and more crucially, even after wide resection, only 50% of patients were still disease-free at 10 years [44]. With these facts, questions inevitably arise, and one must ponder why a tumor might come back even after a large excision [7]. The solution might be found in a Japanese paper that shows the presence of tumor cells, or so-called skip metastases, discovered outside the primary tumor mass [2]. Up to 2 cm (about 0.79 in) away from the primary mass, these skip metastases were discovered in 43% of patients. In that series, skip metastases were linked to ineffective local control [7].

Goals of Surgery

The Enneking classification was first used in 1980 to categorize people who had primary tumors that were malignant of the appendicular bone [25]. These patients' local control was improved, and their survival rate went up, thanks to this theory [27]. As a result, primary malignancies of the axial skeleton are now treated according to the Enneking classification [7]. The main objective of surgery was to obtain negative margins verified by the musculoskeletal pathology department because numerous studies have indicated that local recurrence rates dropped, and rates of survival improved following spinal tumor excision with negative margins [8,12,15,19,27,43,46,50]. For the best local management, a surgical margin has been suggested based on the tumor grade, tumor area, and the presence or absence of metastases [18].

When applying the Enneking classification standards, these tumors should all receive a large resection due to the aggressive nature of chordoma [18,25,27]. The spine, where marginal resection is most frequent, may make the wide-margin notion less useful [7]. Enneking appropriate (EA) refers to resections that produce marginal or wide margins; Enneking inappropriate (EI) refers to resections that produce intralesional or infected margins [18,27,43]. But as was already indicated, recently published material demonstrates that micro skip metastases, which are invisible by modern imaging techniques, can happen as far as 20 mm (about 0.79 in) from the primary tumor [2]. It is important to

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note that just 1 study including the sacrum has looked at skip metastases [7]. If the movable spine falls under this definition, it is not clear [7]. The best current evidence, however, favors a more robust surgical approach for treating these tumors despite the idea of skip metastases and attaining a negative margin is associated with longer survival [12,27,30]. Therefore, until improved long-term outcomes from other types of therapy are available, evidence-based oncologic guidelines should be followed [7]. Achieving these margins, however, comes at higher morbidity and greater cost to neurologic function [7]. To effectively treat these individuals, patient interest should always be a primary consideration [7].

Surgical Approaches

Resection typically entails two anterior steps followed by a posterior step; however, this is less common [17].

ANTERIOR APPROACH

The anterior technique often entails sub-umbilical medial laparotomy so that the tumor is dislodged anteriorly at the retro rectal region and facilitates peritumoral devascularization [23]. The procedure is paired with the anterior rectus flap in the release plane to fill the posterior wall defect [17]. Surgeons have been performing this laparoscopy procedure to lessen hemorrhage and morbidity [24]. In recent years, clinicians abandoned using epiploon flaps because the Da Vinci robot requires reinstalling for release to be more shorter and distal [14]. On the other hand, surgeons stopped using the epiploon flap because the robot needs to be reinstalled [17]. The anterior step might be connected to a definitive colostomy or a colostomy with a rod [17]. In cases of sacrectomy where the roots from S2 are sacrificed, a definitive colostomy is advised [17]. To lower the risk of postoperative infection, a temporary colostomy with a rod may be suggested after sacrectomy sacrificing the S3 root [17].

POSTERIOR APPROACH

The posterior approach is carried out when the patient is in the prone position, either during the previous step of surgery or, ideally, within 24-48 hours (about 2 days) following the anterior step [17]. The biopsy zone is always included in the incision, which may be arched transverse or medial longitudinal [17]. The medial technique permits accessibility high on the spine although necessitates more relaxation to access laterally [17]. The disadvantage is closeness to the anus, which increases the danger of postoperative contamination by bacteria, particularly due to sphincter problems [17]. The transverse technique restricts reaching above the first sacral segment and based on the biopsy route, it is occasionally non-practical [17]. In contrast, this was found helpful in lateral tumor cases progressing into the gluteals and remaining away from the anus [17]. Gluteus maximus can be split in the lateral direction on the posterior approach to expose the sacroiliac foot, occasionally up to its insertion on the trochanter and away from the tumoral invasion [23]. With cystic forms that have an intramuscular invasion, intraoperative ultrasonography can help secure the sectioning [17]. Piriform muscles are divided laterally after ligation of the inferior gluteal pedicle [17]. If the tumor has already reached the ligaments, osteotomy of the sciatic spine or incising of sacrospinous and sacrotuberous ligaments is performed [17]. Forward of the sciatic nerve, the anterior release plane is discovered [17]. To ligate the dural sac upstream of the invasion, a sacral laminectomy is necessary in the healthy zone [17]. The foramen liberates the final set of sacral roots that are still conservable [17]. Sacral osteotomy is carried out from posterior to anterior in the anterior release zone by the use of a finger [17]. Navigation and patient-specific cutting guides can also aid with osteotomies [17]. After raising the specimen, the anterior dissection of the sacrum, the rectum, and the sacrorectogenitovesicopubic laminae is finished [17]. The specimen is extracted in one piece, and parietal reconstruction is carried out, as well as any essential bone reconstruction if needed [23]. For distal malignancies that do not extend over S3, an isolated posterior approach is an option [17]. However, two-step surgery is

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preferred for osteotomies performed through the S3 holes in cases of significant anterior extension [17]. If the lesion is central and does not include the sacroiliac region or if the anterior tumor is not progressed, Angelini advises against trans-S2 sacrectomy [3]. He reported 2,961 ml of average loss of blood [17]. The mere viable option at that point is a gluteal flap, albeit flaps are not usually required for distal sacrectomy [17].

A costotransversectomy and lateral extra-cavitary approach can be used to achieve posterior access to chordomas of the thoracic region [40]. The capacity to distinguish clearly between the normal dura and the tumor, the ability to see the ventral dura, and the ability to do reconstructions of both the anterior and the posterior column from the same technique are all benefits of the posterior approach to the chordoma of the thoracic spine [40]. The amount of the tumor's ventral extension from the vertebral body, its lateral extension, and the major arteries' proximity are typically taken into consideration when deciding whether to do a thoracotomy [40]. To see the parietal pleura, a blunt plane is created between the parietal pleura and the vertebral body for chondromas susceptible to a posterior approach [40]. Ribs must be removed to perform this process. Segmental vessels are cut down as part of the dissection process [40]. Following the development of this plane and the ventral mobilization of the major vessels, thread wire saws might be inserted above and below the tumor's level inside this plane [52,53]. During this time of the laminectomy, the epidural plane is formed anteriorly to the spinal cord [40]. The removal of posterior rudiments is then carried out after this [40]. Following that, the thread wire saws are transferred ventrally toward thecal sac and dorsally to vertebral bodies [40]. The affected nerve roots need ligation before osteotomies can be done safely [40]. Compared to the cervical and the higher thoracic spine, the functional impact of sacrificing thoracic nerve roots in lower and middle thoracic levels is typically negligible [40]. Blood supply to the spinal cord may be jeopardized, though, if the bilateral nerve roots are sacrificed on more than the two successive points [40]. In this case, intraoperative monitoring and temporary clip placements across the thoracic nerve root before sacrificing the nerve root can give information about vascular supply to the spinal cord at that level [40]. Thread wire saws can finish osteotomy after the thread wires have been safely passed, the epidural plane has developed around the thecal sac, and the nerve roots at the afflicted area have been sacrificed [40]. The soft tissue attachments are removed, and tumor is moved away from spinal cord once it is free of its osseous attachments [40]. It is at this time that anterior/posterior reconstruction is started after the specimen has been examined by pathology for margin analysis [40].

Bone Reconstruction

If the adjacent sacroiliac joints and the first sacral vertebra are preserved, no bone replacement is required [17]. In contrast, sacrifice of more than half of the sacroiliac joint or total sacrectomy necessitates the restoration of spinopelvic continuity through the lumboiliac fusion using "Eiffel tower" assembly which consists of stability triangle formed by the rods connected to the pedicular screws and screws in iliac bone, as well as the rods connecting the two iliac bones transversely, along with the anterior reconstruction by allograft or cage [6]. Reconstruction lengthens operation time and raises the risk of infection, regardless of whether a bone graft is made of the posterior iliac crest, tibial strut, or non-vascularized or vascularized fibula [6]. Nearly 25% of patients experience non-union, and 80–90% can move around, with technological aid needed in 50% of instances [6,23,51]. Without the reconstruction, the spine methodically declines into the pelvis, occasionally with an acceptable function and occasionally with spontaneous fusion [17]. In the older series, there was a frequent lack of reconstruction, due to a dearth of appropriate instrumentation [17].

Radiation Therapy

Radiation therapy is frequently thought to have a negligible effect on chordomas [7]. However, the majority of historical data indicate that in the salvage context, relatively low radiation doses (less than 50 Gy) were employed [7]. The application of radiation appeared to have been kept for situations where it was impossible to treat a recurring chordoma under ideal conditions [7]. Additionally, it is

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acknowledged that chordoma cannot be treated with lower radiation doses [7]. The radiation dose, however, has been increased thanks to improvements in radiation administration. Some, such as Massachusetts General Hospital (MGH), have called for doses of more than 70 Gy [7]. With time came experience at MGH employing elevated levels of radiation as an adjunct to surgery, and the results compared well to earlier studies [20]. At MGH, radiation is delivered using protons, which have advantageous distribution patterns that include a high concentration of dose at the tumor and no exit dose [7]. These elements lessen radiation's effect off-target, enabling one to raise the dose.

Carbon ion radiation devises more popular recently for the treatment of chordoma [7]. The use of carbon ions exploits its characteristics to increase the radiation dose [31]. It has also been suggested to use hypo-fractionated dosing schedules to boost the biologically effective dose as a result of improvements in photon radiation delivery [33,41]. Within the last ten years, local control rates using these several radiation treatments, both as a stand-alone therapy and as assistance to surgery, have been more encouraging [7]. In the beginning, radiation's inherent morbidity is greater than what can be anticipated from surgery [7]. Although the enduring effects of radiation on nerve function are unknown, most individuals who had short- to mid-term follow-ups did not have any issues [7].

Similarly, to this, control rates using just radiation are encouraging with the short- to mid-term follow [7]. Chordoma patients must be closely monitored over an extended time because recurrence of chordomas is considered later in comparison with the majority of bone tumors that are malignant [7]. The management variability between facilities across the globe is a reflection of the ambiguities regarding the best course of action for chordoma treatment, whether radiation is used or not [11].

Complications

INFECTIOUS PROBLEMS

With rates ranging from 20% to 60% [23,45], cutaneous and infectious complications are the most common. An early scar disunion following surgery necessitates surgical revision and ongoing antimicrobial therapy [17]. Additionally, it possible is a late infection [17]. The risk of infection increases with closer resection [38].

MECHANICAL SIDE EFFECTS

Following radiation therapy, a pathologic condition known as fatigue fracture affects almost half of the patients [9,39], varying in duration from 1 to 62 months (about 5 years) in the case of Bostel et al. [9]. In a group of 24 chordoma patients who had solely radiation therapy for their chordomas, including 19 sacral chordomas, 8 pathologic fractures were found, and all the patients were inpatients using morphine derivatives at the time of the last follow-up [13]. Contrary to the findings of Bostel and Osleret found that fractures occurred more frequently in surgery combined with radiation therapy than only in radiation therapy [9,39]. Even without radiation therapy, sacrectomy above the S2 only carries a risk of pathological fracture, hence prophylactic internal fixation should be used [17].

HEMORRHAGIC COMPLICATIONS

When compared to laparotomy, the anterior technique requires less blood loss [24,57]. Nevertheless, the primary source of bleeding is the posterior approach with the sacral osteotomy [17].

ADDITIONAL PROBLEMS

Following a sacrectomy for chordoma, many additional problems have been documented, including sacroiliac instability [35], non-union [23], digestive fistula [17], osteomyelitis [17], ureter wound [17], CSF leaking [17], and pseudo-meningocele [23]. To improve their quality of life, they need specialist interdisciplinary management [17]. Five patients (20.8%) displayed an exacerbation or emergence of the neurologic impairment (saddle anesthesia, saddle anesthesia, worsened

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incontinence, and erectile dysfunction) [13]. Of sacral chordoma patients treated with carbon ion radiation therapy, 211 (96%) of which were treated without surgery, Demizuet al. [21] reported grade 3 complications in 5.9% of patients at a mean of 56 months with 2.7% skin complications, 1.4% myositis, 0.9% pathologic fracture, 1.4% chronic pain.

According to Imai et al. (69), during a mean follow-up of 62 months, 5% of patients receiving carbon ions experienced grade 3 problems, such as skin issues or neuropathic pain. Imai et al. [32] found no vesical or sphincteral problems in sacral chordoma patients (primary or recurrent after surgery) that had carbon ion radiation and were thought to be incurable. However, 15.8% of the patients had significant sciatic neuropathy that required medical attention [32].

Outcomes of Chordoma Surgery

METASTASIS, RECURRENCE, AND SURVIVAL

After broad excision, there is a 40% to 20% local recurrence rate of sacral chordoma and a 20% to 40% metastatic resurgence rate [34]. Difficulty in performing successful wide resection or gross total resection (GTR) of the spinal chordomas is demonstrated by high rates of recurrence and metastasis, which emphasizes the significance of cautious surgical management of these patients [5]. Because infection in the resection margins can increase the risk of recurrence and mortality, as shown in a recently published meta-analysis, it is crucial to achieve a wide excision [60].

By itself, primary radiation has a poor therapeutic effect on chordomas [41]. Although some studies have shown an increase in survival whenever adjuvant radiation therapy is utilized in marginal and intralesional resections, the application of adjuvant high-dose radiotherapy in spinal chordomas remains debatable [4,59,61]. A recent database analysis has shown not much benefit in cases where GTR has been achieved [58]. Promising developments have been made in more accurate radiotherapy techniques, such as stereotactic radiotherapy, carbon ion treatment, and proton beam therapy [5].

CONCLUSIONS

The patient should be carefully informed about the surgical process, expected advantages, and any problems. It is difficult to standardize this operation, and the surgeon must know to modify the surgical process to meet the needs of each patient. As a result, the surgical technique and fixing method must be uniquely tailored to the individual patient. The patient should be psychologically and physically prepared, as well as informed about the difficulties and anticipated aftereffects that may reduce the quality of life. If a colostomy is anticipated, learning how to self-catheterize may be necessary before surgery, along with visiting a stoma therapist. Thus, surgical management requires multidisciplinary skills from the fields of surgery, anesthesia, radiology, etc. Surgery carried out outside of a reference facility is a separate risk factor for fatal results.

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