Journal of Orthopaedics 15 (2018) 679–684

Contents lists available at ScienceDirect

Journal of Orthopaedics

journal homepage: www.elsevier.com/locate/jor

Review Article

Sacral chordoma : A review of literature

S. Pillai[a,*], S. Govender[b]

[a] King Edward Hospital, Durban University of Kwazulu Natal, South Africa
[b] Consultant Spine Unit, King Dinuzulu Hospital, Durban, South Africa

ARTICLE INFO

Keywords:
Sacral tumor
Chordoma
Pelvic tumor
Bone tumor

ABSTRACT

Chordoma is a rare malignant tumor with predilection for the sacrum. A favorable outcome depends on early diagnosis and surgical excision with tumor free margins.

Aim: To review the incidence and prevalence of sacral chordoma and the appropriate surgical management and formulate a criteria for early diagnosis.

Materials and methods: Thirty eight articles on the subject of sacral chordoma and extrapolated data of clinical and surgical significance.

Results: The commonest presenting feature in patients with sacral chordoma was lower back pain which was worse on sitting and up to a third of these patients had associated urinary tract infections, constipation or symptoms of disc herniation. The hallmark of sacral chordoma is lower back pain with poor response to non-steroidal anti-inflammatory drugs.

Disease free eradication in chordoma is rare and not without neurological deficit. The common sacral levels involved ie. S2,3 have to be sacrificed to achieve adequate margins4. Whilst wide excision via posterior surgery shows a decrease in recurrence, the overall survival still remains low.

1. Introduction

Chordoma is a malignant, slow growing and locally aggressive tumor. It arises from remnants of the notochord and accounts for 1–4% of all primary bone tumors[1,6,7]. Although reported to be rare, chordoma is the most common primary malignant tumor found in the sacrum[39].

The insidious onset and trivial symptoms caused by sacral chordoma may account for the late presentation[31]. These tumors are unfortunately diagnosed at a stage where prognosis is poor. The majority of patients are diagnosed in Eneking stage IB which, due to the close proximity to neurovascular structures in the sacrum and pelvis, present a challenge to the treating surgeon [4,5,12]. Chordomas are slow growing tumors and are not amenable to chemotherapy or radiation[32]. The first line of treatment is usually wide surgical excision.

However, excisional removal of the tumor is not without its complications, due to resection of the proximal sacral nerve roots. Many patients suffer urinary incontinence, sexual dysfunction and weakness of the lower limbs.

The tumor grows within the sacral foramina and obstructs the neural canal proximally. Posteriorly the tumor tends to invade the muscles around the hip and pelvis as well as the sacroiliac joints. Anterior invasion is uncommon due to the tough presacral fascia and thus the rectum is usually spared[10,14,24]. Metastases of sacral chordoma occur late in the disease or even many years after surgical resection. The literature reports secondaries found in lung, bone, liver, lymph nodes, soft tissue and skin.

Recurrence many years after surgery can be explained by satellite lesions seeding via the tumors psuedocapsule which makes it difficult to achieve disease free surgical margins.

We undertook a review of the published literature on sacral chordoma from 1995 to 2014. Level 2,3,4 and 5 studies were included in the study and criteria for exclusion were chordoma present in regions other than the sacrum as well as unpublished literature.

The aim of this article is to highlight the clinical significance of this tumor and its prognosis.

2. Embryology

The spine develops from mesodermal structure known as somites. The development of somites proceeds from a cranial to caudal direction which occurs on either side of the notochord and neural tube. Somites are made up of 3 layers ie. Sclerotome(vertebral bodies and annulus fibrosis), myotome and dermatome [44].

The neural crest progresses to form the peripheral nerves, ganglia and sympathetic trunk, as well as the pia mater.

The neural tube forms by a process called neurulation. The

* Corresponding author.
E-mail address: 202500715@ukzn.ac.za (S. Pillai).

https://doi.org/10.1016/j.jor.2018.04.001
Received 12 December 2017; Received in revised form 20 March 2018; Accepted 29 April 2018
Available online 02 May 2018

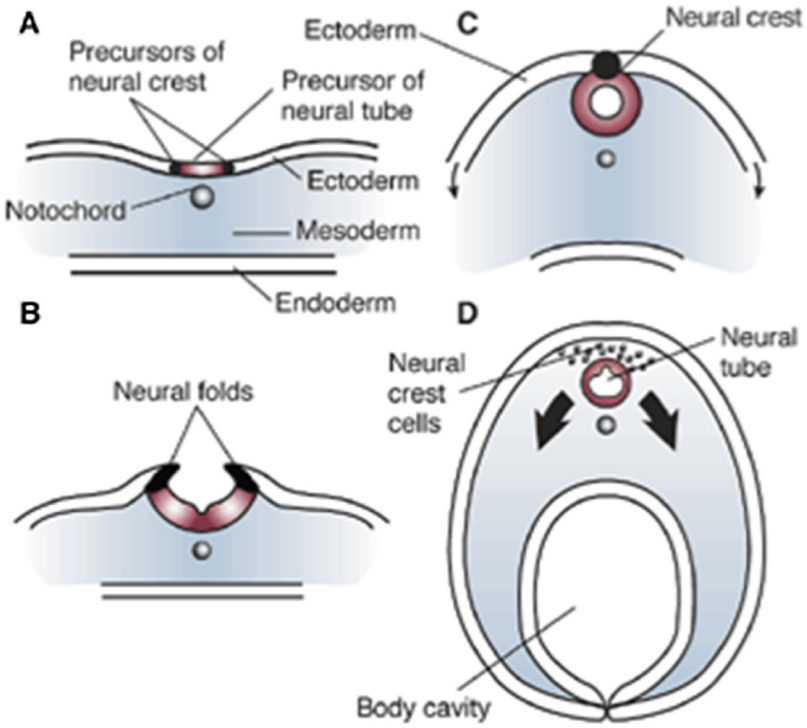
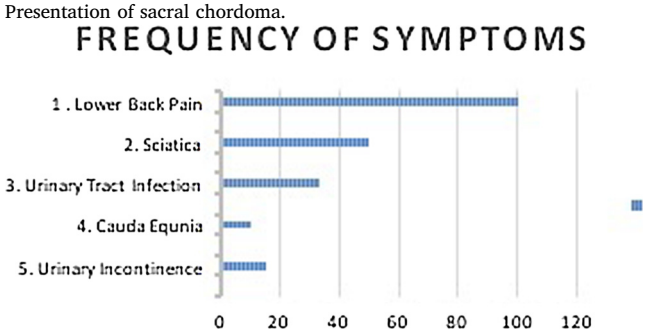


Fig. 1. Embryonic development of the neural tube.

Table 1
Presentation of sacral chordoma.



ectodermal cells elongate to form neural plate cells. The edges of these cells thicken and migrate to the midline. Their fusion leads to formation of a hollow neural tube. This process begins on the 3rd week of fetal life and is complete by the 4th week [37].

The neural tube becomes the spinal cord, and the notochord forms the anterior part of the vertebral body as well as the nucleus pulposus (Fig. 1). It is not uncommon to find remnants of the notochord in the nucleus pulposus of intervertebral discs removed during surgery [38].

2.1. Prevalence

Sacral chordoma is the most common primary malignant tumor of the sacrum. It accounts for 1–4% of all malignant bone tumors[1,6,7].

It is predominantly seen in the caucasian population with males being more affected than females. The male to female ratio ranges from 1.4 : 1 to 2.4 : 1 [2,3,8].

It is typically seen in the 5th and 6th decade with a mean age of 55 years[9]. Less than 25% of patients present in the 3rd and 4th decades, majority of these have chordoma of the mobile spine and cranium[3].

Unfortunately, almost all patients have extra-compartmental invasion at the time of diagnosis [4,5,12]. Symptoms are related to compression and/or destruction of the tumors neighboring structures. Communication between multiple disciplines is essential in decision making and has been shown to minimize neurological symptoms and improve patient outcome [31].

2.2. Presentation

A frequent referral presenting to the orthopaedic surgeon is lower back pain and quite commonly these patients are in the 5th and 6th decade. The vague symptoms produced by sacral chordoma thus poses a challenge for early diagnosis.

All patients with sacral chordoma presented with lower back pain which was typically worse when sitting and pathological in nature[10,11].

Pain and radiculopathy due to compression of the sciatic nerve or iliolumbar trunk was reported in up to one third of these patients[13]. A few cases with tumor presenting in the S1- S2 region had weakness or total loss of L5-S1 function unilaterally[17].

Up to one third of patients presented with symptoms urinary tract infections and 10% had constipation or cauda equina symptoms (Table 1)[1,15,16,19].

Many of these patients had been treated for years for the above symptoms with NSAIDs, antibiotics, laxatives as well as steroid injections by general practitioners before the pathology necessitated further investigations[16,19].

2.3. Investigations

On plain film sacral chordoma appears as a large lytic lesion which crosses the midline > 50% of cases (Fig. 3)[18,19,20]. Lesions may occur anywhere from the first to fifth sacral vertebrae with predilection to the 2nd and 3rd sacral bodies (Fig. 2)[1,6,8].

The destruction of bone extends to the sacro-iliac joints and progresses to the ilium as well as posteriorly which may cause skin changes in the lower back. Destruction of the anterior ligamentous structures as well as the L5-S1 junction leads to listhesis and pelvic instability[39].

A full skeletal survey must be performed to include the hips, lumbar spine as well as the skull. Technecium[99] scan may assist in localizing the pathology for a more focused review.

CT scan assists in delineating the bony extent of the lesion and is of

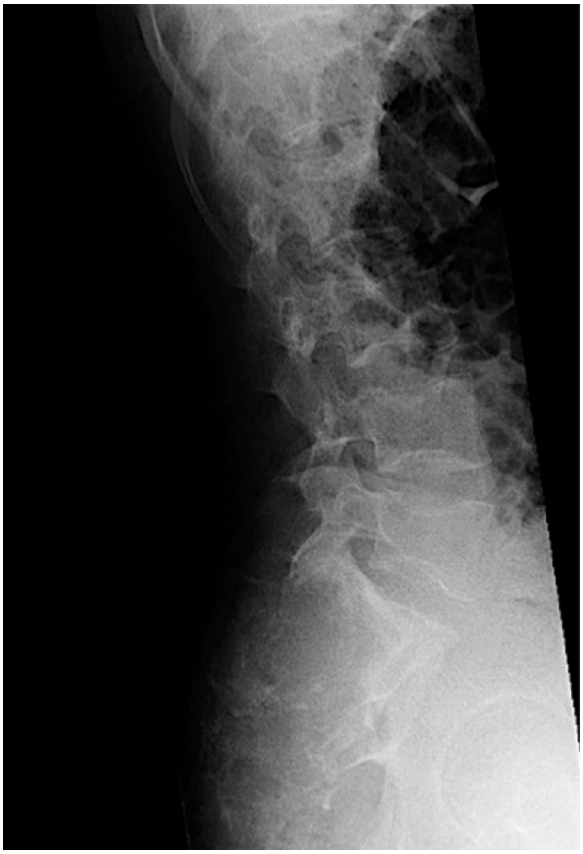


Fig. 2. Chordoma involving almost the entire body of the sacrum with minimal presacral swelling due to the tough anterior fascia. (KGV Hospital 2013).

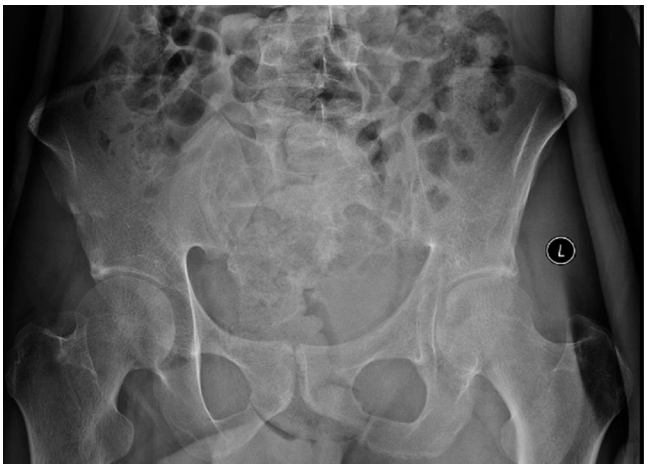


Fig. 3. Pelvis x-ray showing tumor mass extending across midline and involving (R) sacroiliac joint.

great value in patients with chordoma to plan any surgical intervention (Fig. 4)[14,19].

the significance of the abdominal MRI and has reported metastasis in the anterior abdominal wall[23].

At surgery chordoma is commonly found attached to the piriformis and gluteal muscles which is of significance in determining the surgical approach (Figs. 5 and 6).

Several authors have correlated the volume of the tumor as a predictor of the outcome[1,10,25,26]. They have found that at the time of diagnosis sacral chordomas larger than 10 cm or a volumetric dimension of > 200cm3 invariably carried an unfavorable prognosis.

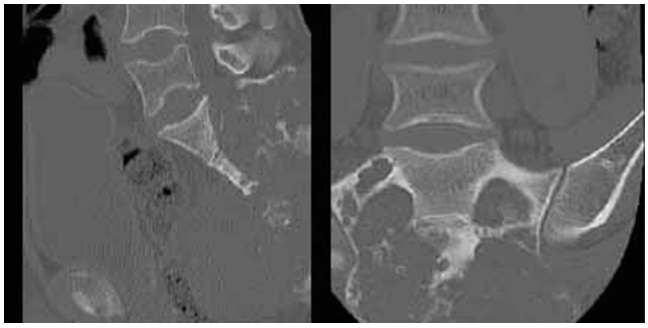


Fig. 4. CT scan delineates extent of lesion and shows sacroiliac joint involvement. MRI (T2) findings show a large hyper-intense inhomogeneous mass with a pseudocapsule (Fig.5). The mass is usually lobulated or may be multilobulated [12,14,19–22]. MRI should include the entire pelvis as well as the abdomen. Santos-Rancano et.al highlighted.

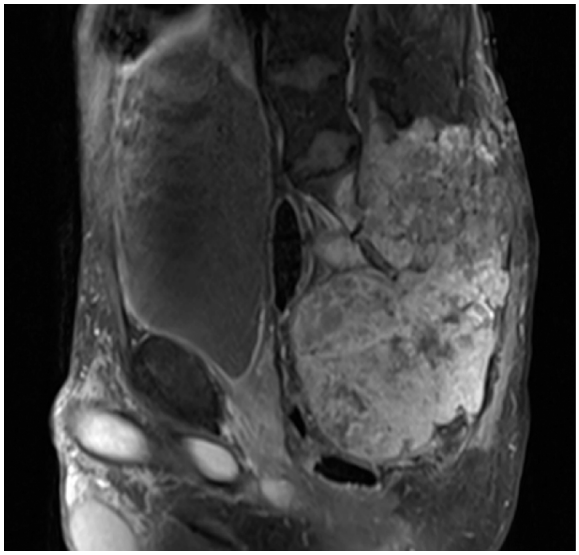


Fig. 5. T2 weighted saggital MRI show large lobulated chordoma invading soft tissues. Anterior extension of the tumor is seen in less than 25% and is probably due to the tough sacral fascia present on the anterior part of the sacrum10,14,24.

Histologically the classic chordoma mass consists of chords of tumor cells and lobules with a mucoid matrix and extensive fibrous tissue inbetween (Fig. 7).

Physaliferous cells are pathognomic of chordoma and are characterized by their large size with a vesicular nucleus and vacuolated cytoplasm. Mitotic figures as well as bone and cartilage may also be seen.

Toru and Majunder have attributed poor prognosis to histological patterns which contain areas of high grade sarcoma and have been termed dedifferentiated chordoma[11,27].

Sakwe and Cuenca et al. reported a cartilaginous counterpart and is termed chondroid chordoma. These tumors show chondroid differentiation and carry the most favorable prognosis. However, it is often found in the spheno-occiptial region and rarely seen in chordomas involving the sacrum[28,29].

2.4. Differential diagnosis

Several conditions may mimic sacral chordoma and should be considered in the differential diagnosis. Chondrosarcoma closely mimics sacral chordoma both radiologically and histologically and should be high on the list of differentials. Immuno-histochemical studies may

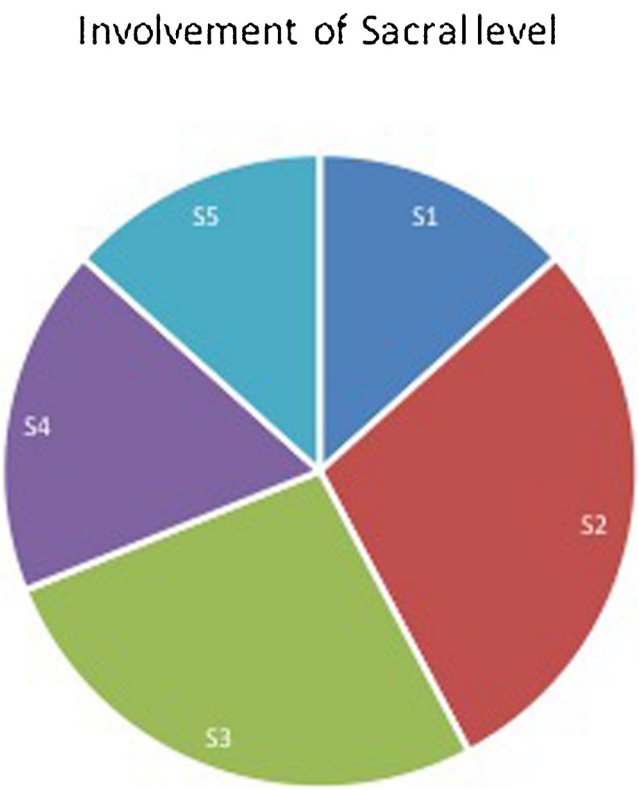


Fig. 6. 45–60% of sacral chordomas occur at S2-3, S1 being the least likely location for tumor.

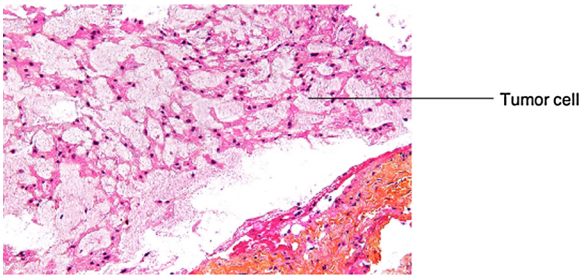


Fig. 7. showing physaliferous arrangement of tumor cells Image courtesy of http://www.wikimedia.org.

be required to differentiate these tumors histologically. Chordoma shows positive for epithelial membrane antigen (EMA), pan-cytokeratin (panCK)[40].

Giant cell tumor and Plasmacytoma are also found in the sacrum.

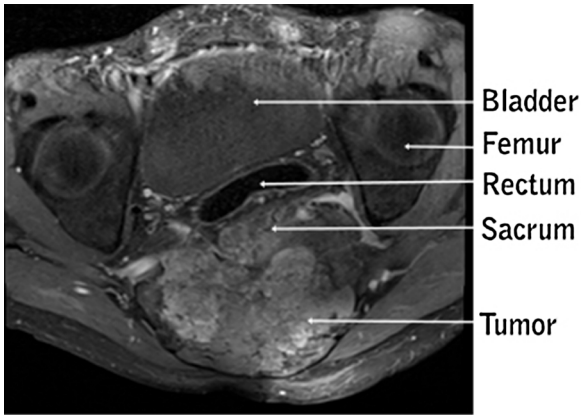


Fig. 9. Large lobulated chordoma displacing the rectum but not invading the pre-rectal fascia. Note involvement of the femoral heads bilaterally.

The radiographs appear as large lytic lesions with poorly defined borders. These tumors are aggressive and expansile, and may cross the sacroiliac joint posing a radiological challenge to differentiate from chordoma.

Metastatic disease is especially seen in the sacrum in the elderly with primaries from the prostate, breast, thyroid, lung and colon. A thorough history pertaining to symptoms and previous ailments may help in guiding towards investigations (Fig. 8).

Chronic infections caused by tuberculosis and fungus should always be considered.

3. Management

3.1. Surgical

The goal of surgery in the management of sacral chordoma is eradication while retaining as much function as possible.

Meticulous planning the level of resection is important and requires careful assessment as tumor may extend via the sacrospinal canal into adjacent segments[17,18,19] It is therefore not sufficient to determine the level of sacrectomy by assessing bony involvement alone. MRI is essential in the preoperative assessment and staging of the tumor.

Mavrogenis used target artery embolization to reduce pain as well as intraoperative blood loss and suggested that this may be the only option in patients not amenable to more invasive surgical intervention[41].When possible at least one nerve root at each level of sacral resection should be preserved. Care should be taken not to penetrate the capsule around the tumor at the time of surgery as this has been correlated with a high incidence of recurrence[4,21].Sacral amputations below the level of S1 show great variance in neurological outcome. Motor deficit is rare but saddle anaesthesia and loss of bowel and

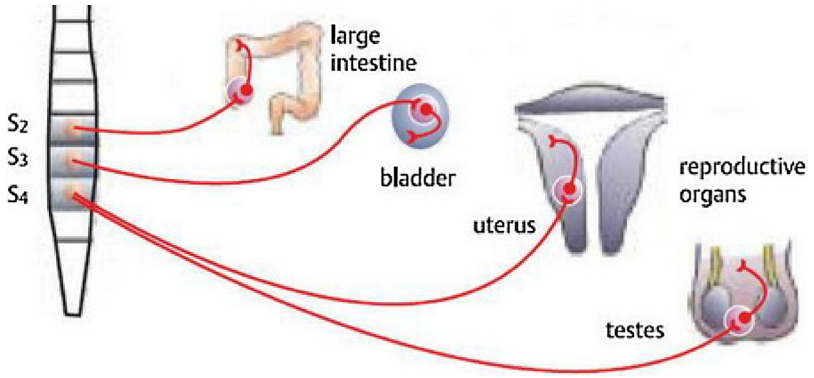


Fig. 8. Diagram showing innervation of bowel and bladder.Image courtesy of http://www.ccptr.org.

Table 2
Posterior only approach has provided favorable outcomes on long term follow up. Anterior surgery requires an access surgeon as abdominal contents may show gross distortion due to tumor size or adhesions (Fig. 9). It provides direct access to the prerectal fascia and allows for bowel resection if needed. In cases where a layer of fat is still present anterior to the sacrum, the rectum can be left intact and a colostomy may not be required [41].

Author	No. of patients	Site	Surgical procedure	Recurrence	Metastases	Survival	Urinary / fecal incontinence
G. Housari	13	S2-5	Posterior	30% at 2 years	45% at 5 years	27% at 10 years	90%
M. Waisman	5	S2-3	Posterior	20% at 5 years	0% at 5 years	100% at 5 years	20%
A. Asavamongkolkul	21	S1-5	Posterior	0% at 2 years	9% at 7 years	5% at 7 years	100%
M. Clarke	30	S1-4	Posterior	20% at 5 years	6% at 3 years	96% at 4 years	67%

bladder sphincter control is commonly seen[5].

In resections involving the S1 nerve root, motor deficits and sexual dysfunction is almost always seen. Generally, patients with sacrectomy below the level of S3 have sphincter control with some perineal anaesthesia. In all cases, the lumbosacral trunks and Sciatic nerves should be protected where possible.

Gunterberg et al reported that one side of the sacroiliac joint may be removed before weight bearing was affected. Total sacrectomy however results in gross instability of the spino- pelvic junction and stabilizing procedures must be done before any mobilization[42].

Ishida reported that spinopelvic reconstruction following sacral tumor resection requires three components ie. Anterior column support, posterior pelvic ring stablisation and spino- pelvic fixation. S2 alar-iliac screws has shown to have the greatest mechanical strength at

the lowest risk of visceral damage and blood loss. Fusion across the sacro-iliac joint however is rare with this construct which may lead to early osteoarthritis and require removal of the screws[43].

There has been much debate around the most appropriate approach for excising the tumor mass. While some authors advocate a posterior only approach[35], others believe that a combined anterior and posterior surgery is beneficial in reducing reccurrences[36].

The posterior only approach has been successfully carried out with good results at a number of institutions[35]. The rate of recurrences has been shown to be marginally lower than that of the combined anterior approach (Table 2)[36]. This approach however may not be possible with tumors involving bowel and other viscera[35].

The combined anterior and posterior approach were previously reserved for tumors more than 10 cm and always included a colostomy[5]. The achievement of a wide resection came at the expense of increased morbidity and mortality. In a study done by Ozger et al. in 2010 an average 9 units of blood were required during surgery and neurological deficit post operatively was not uncommon.

3.2. Radiation therapy

It has long been believed that sacral chordoma was not amenable to radiation. However, the work published by Park et al.[32] showed excellent results with localized radiation post en- bloc resection. He reported a 5yr and 10yr survival or 93% and 91% respectively.

For patients who are not candidates for surgical intervention due to poor host factors(renal/cardiac etc), radiation therapy alone may be warranted. Chen et al.[33] reported a 5yr survival rate of 78% with high dose radiation in his study of patients who where not fit for surgery

3.3. Chemotherapy

Due to poor sensitivity, chemotherapy is not instituted in patients with sacral chordoma[31] in most centres and thus surgical management remains the mainstay of treatment. An ongoing study by Stacchiotti et al documents the use of Imatinib 800 mg/day for the non-surgical management of chordoma. He has reported a benefit rate of 64% with a 9 month progression free period of the tumor[34].

4. Discussion

Early diagnosis of sacral chordoma still remains an issue. The time of onset of pain to time of diagnosis is almost always more than 18 months. With the advent of CT/MRI as well as the availability of bone scan and X-Ray, these tumors may very well be diagnosed at an earlier stage offering the patient a more favorable diagnosis.

Criteria should be devised which should include a close follow up plan.From this review the following risk factors for sacral chordoma are highlighted :

Patients in the 5th and 6th decade presenting with a history of 1 year of dull aching but progressive lower back pain require further investigations. Pain has been typically described as worse on sitting and commonly disrupts sleep.

A history as above together with symptoms of a urinary tract infecrtion, saddle anaesthesia and/or constipation warrants an earlier follow up.

A plain film radiograph and inflammatory markers will help categorise patients into high or low risk.

Unlike most bone and spinal tumors, sacral chordoma does not have its own classification. The Weinstein-Boriani classification of spinal tumors does not hold much significance in management of sacral chordoma as the sacrum and surrounding structures require a different surgical approach.

The enneking classification serves as a more useful tool in staging and preoperative planning. Prognosis can be more accurately determined after biopsy and CT/MRI results.

With regards to management, a multidisciplinary approach must be adopted as resources permit. Although the frequency of recurrences is high, radical resection provides a longer disease free interval which may be further prolonged with radiation after surgery.

The posterior only approach has provided favourable results[35] when compared to the anterior approach. The combined approach has shown a greater margin of resection but at the expense of patient mortality with no increase in long term survival[5].

All patients should be counselled on the high likelihood of loss of bowel and bladder function post operatively. Males in particular must be made aware of the loss of sexual function.

The posterior only approach has yielded favorable results and carries a lower risk of haemorrhage but may not always be possible where visceral adhesions are present.

Funding

No funding was received for the above study.

References

1. Baratti D. Chordoma: natural history and results in 28 patients treated at a single institution. *Ann Surg Oncol.* 2003;10(3):291–296.
2. Stacchiotti Silvia. Chordoma of the mobile spine and sacrum: a retrospective analysis of a series of patients surgically treated at two referral centers. *Ann Surg Oncol.* 2010;17:211–219.
3. McMaster Mary L. Chordoma: incidence and survival patterns in the United States, 1973 ± 1995. *Cancer Causes Control.* 2001;12:1–11.
4. Asavamongkolkul Apichat. Wide resection of sacral chordoma via a posterior

approach. *Int Orthopaedics (SICOT).* 2012;36:607–612.
5. Ozger Harzem. Surgical management of sacral chordoma. *Acta Orthop. Belg.* 2010;76:243–253.
6. Varga Peter Paul. Chordoma of the sacrum: “En bloc” high partial sacrectomy. *Eur Spine J.* 2010;19:1037–1038.
7. Hanna SA. Sacral chordoma can local recurrence after sacrectomy be predicted? *Clin Orthop Relat Res.* 2008;466:2217–2223.
8. Ferraresi, et al. Chordoma: clinical characteristics, management and prognosis of acase series of 25 patients. *BMC Cancer.* 2010;10:22.
9. Chen Kang-Wu. Expression of vascular endothelial growth factor and matrix metalloproteinase-9 in sacral chordoma. *J Neurooncol.* 2011;101:357–363.
10. Atalar H. Management of sacrococcygeal chordomas. *Int Orthopaedics (SICOT).* 2006;30:514–518.
11. Akiyama Toru. Juxtacortical chordoma of the sacrum. *J Orthop Sci.* 2008;13:476–480.
12. Nagra Inderjeet. A 45-year-old woman with a pre-sacral mass lesion: diagnosis and discussion. *Skeletal Radiol.* 2010;39:199–200.
13. Fourney Daryl. Current management of sacral chordoma. *Neurosurg Focus.* 2003;15(2) Article 9.
14. Sung Mi Sook. Sacrococcygeal chordoma: MR imaging in 30 patients. *SkeletalRadiol.* 2005;34:87–94.
15. Jeys Lee. Sacral chordoma: a diagnosis not to be sat on? *Int Orthopaedics (SICOT).* 2008;32:269–272.
16. Osaka Shunzo. Clinical significance of a wide excision policy for sacrococcygeal chordoma. *J Cancer Res Clin Oncol.* 2006;132:213–218.
17. Bergh Peter. Prognostic factors in chordoma of the sacrum and mobile spine. *Cancer.* 2000;88(9):2122–2134 1 May.
18. Kishimoto Riwa. Imaging characteristics of metastatic chordoma. *Jpn J Radiol.* 2012;30:509–516.
19. Gerber S. Imaging of sacral tumours. *Skeletal Radiol.* 2008;37:277–289.
20. Maheshwari Aditya V. Low Backache in a 70-year-old woman. *Clin Orthop Relat Res.* 2009;467:596–602.
21. Akiyama Toru. Juxtacortical chordoma of the sacrum. *J Orthop Sci.* 2008;13:476–480.
22. Carey Kathleen. Diffuse skeletal muscle metastases from sacral chordoma. *Skeletal Radiol.* 2014;43:985–989.
23. Santos-Rancaño, et al. First case of metastasis of a Chordoma in the abdominal Wall. *J Cancer Sci Ther.* 2013(11):5.
24. Rao BSS. Sacral chordoma – a report of two cases. *Indian J Surg.* 2005;67:207–209.
25. Kayani B. Prognostic factors in the operative management of dedifferentiated sacral chordomas. *Neurosurgery.* 2014;75(September (3)):269–275.
26. Ishil Ken. Local recurrence after S2-3 sacrectomy in sacral chordoma. Report of four cases. *J Neurosurg (Impact Factor: 3.23).* 2002;97(August):98–101. http://dx.doi.org/10.3171/spi.2002.97.1.0098.
27. Majumder N. Sacral chordoma - a rare case report with brief review of literature. *JKIMSU.* 2013;2(July–December (2)).
28. Sawke GK. Sacrococcygeal chondroid chordoma: a case report with brief literature review. *Clin Cancer Investig J.* 2014;3:116–118.
29. Cuenca G, et al. Sacrococcygeal Chordoma: Combined Surgical Approach In An 86-Year-Old Male. *Internet J Surg.* 2002;4(2).
31. González Mariana, Calero Purificación, Beni Rafael, Lobo Eduardo. Sacral chordoma: management of a rare disease in a tertiary hospital Gada Housari. *Clin Transl Oncol.* 2013;15:327–330.
32. Park, et al. Sacral chordomas: Impact of high-dose proton/photon-beam radiation therapy combined with or without surgery for primary versus recurrent tumor. *Radiat Oncol.* 2006;65(August (5)):1514–1521.
33. Chen, et al. Definitive High-Dose Photon/Proton Radiotherapy for Unresected Mobile Spine and Sacral Chordomas. *J Spine.* 2013;38(July (15)):E930–E936.
34. Stachiotti, et al. Phase II study of imatinib in advanced chordoma. *J Clin Oncol.* 2012;30(March (9)):914–920.
35. Clarke, et al. v: Posterior-Only Approach for En Bloc Sacrectomy: Clinical Outcomes in 36 Consecutive Patients. *J Neurosurg.* 2012;71(August (2)):357–364.
36. Ozger Harzem, Eralp Levent, Sungur Mustafa, Atalar Ata Can. Surgical management of sacral chordoma. *Acta Orthop Belg.* 2010;76(2):243–253.
37. Williams S, Beckerman B, Fonti MEP. A 69-Year-Old Presenting With Musculoskeletal Low Back Pain: A Case of Lumbosacral Chordoma. *J Chiropr Med.* 2014;13(June (2)):144–148.
38. Wang WL, Abramsom JH, Ganguly A, Rosenberg AE. The surgical pathology of notochordal remnants in adult intervertebral disks: a report of 3 cases. *Am J Surg Pathol.* 2008;32(August (8)):1123–1129.
39. Farsad Khashayar, et al. Sacral Chordoma. *RadioGraphics.* 2009;29(5):1525–1530.
40. Choy HY. Immunohistochemical comparison of chordoma with chondrosarcoma, myxopapillary ependymoma, and chordoid meningioma. *Appl Immunohistochem Mol Morphol.* 2009;17(March (2)):131–138.
41. Mavrogenis AF. Embolization of Bone Tumors. *Orthopedics.* 2011;34(April (4)):303–310.
42. Fourney DR. Spinal instability and deformity due to neoplastic conditions. *Neurosurg Focus.* 2003;14(1) Article 8.
43. Ishida W. *Spinopelvic reconstruction following lumbosacral tumor resection.* Department of Neurosurgery, e Johns Hopkins University, School of Medicine WScJ 1; 2016:25–30.
44. Kaplan KM, Spivak JM, Bendo JA. Embryology of the spine and associated congenital abnormalities. *Spine J.* 2005;5(September–October (5)):564–576.