



Sacrococcygeal chordoma presenting as a retro rectal tumour

Pragnya Chigurupati*, Vishnukumar Venkatesan, Manuneethimaran Thiyagarajan, A. Vikram, Kaundinya Kiran

Sri Ramachandra University, Chennai, Tamil Nadu, India

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ABSTRACT

INTRODUCTION: Chordomas are rare, slow growing, locally destructive bone tumours arising from the notochord.

PRESENTATION OF CASE: Presenting a case of a 65 year old man, who presented with complaints of swelling on the right lower back for 1 year associated with pain.

On physical examination, a swelling measuring 5 cm × 4 cm was noted in the lower back with posterior wall indentation on per rectal examination.

MRI revealed a mass lesion involving the sacrum (s3–s4) and coccyx. FNAC showed features of a chroma. At surgery, we excised a mass from the retrorectal space and biopsy proved it to be a chondroid chordoma, a variant of chordoma.

DISCUSSION: Chordomas are solid malignant tumours that arise from vestiges of the foetal notochord. Common locations are the clivus and the sacrococcygeus region.

Annual incidence of these tumours is 1 in one million. MRI is the imaging modality of choice. Prognosis improves based on the age, resected margins and postoperative treatment.

CONCLUSION: Here, we shall discuss the literature, variants, treatment and prognosis of this rare tumour.

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1. Introduction/background

Chordoma is a rare, low-grade malignant bone tumour arising from primitive notochord remnants of the axial skeleton.¹ Their incidence rate is 0.1/100,000/year. Chordomas involve the sacrococcygeal region in 50–60% of the cases and the clivus or the sphenoid occipital region in 30–35% cases.² They account for over 40% of all sacral tumours. Local invasiveness and destructiveness are characteristic features of the disease. Complete surgical excision is the main therapeutic modality able to effect a cure. The importance of radiation has gradually increased overtime.³ Imaging techniques in particular MRI, play a crucial role in surgical planning.

2. Case presentation

Our patient was a 66-year-old male with complaints of swelling over his lower back for well over a year. The swelling was progressively increasing in size and was associated with pain for 5 months. On examination, a 6 cm × 8 cm firm, immobile swelling was noted

over the sacrum. Per rectal examination revealed an indentation on the posterior rectal wall.

3. Investigations

Blood investigations were done, which were within normal limits.

An MRI of the lumbosacral region was done which showed features suggestive of a Chordoma attached to the sacrum at S4 and S5. The tumour was seen extending both posterior to the sacrum which was clinically evident and anterior to it, in the retro rectal space Fig. 1.

F.N.A.C. of the swelling was done which showed features suggestive of CHORDOMA.

4. Treatment

We decided to approach the tumour using a posterior approach. The patient was in prone position with hips raised. Using a longitudinal incision over the tumour, flaps were raised to visualise it. The tumour was dissected free from surrounding structures and its attachment to the sacrum was visualised. A partial sacrectomy was done, removing s3, s4 and s5. After the sacral attachment was released, the tumour was excised in toto, by using the retrorectal pad of fat as a dissection plane (Figs. 2 and 3).

* Corresponding author at: Sri Ramachandra Medical College and Research Institute, Chennai, Tamil Nadu, India. Tel.: +91 9500002141.

E-mail address: pragnyac89@gmail.com (P. Chigurupati).

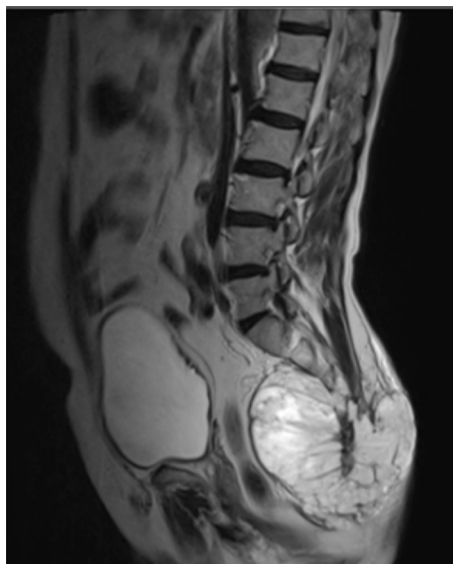


Fig. 1. A 12 cm × 8 cm heterogenous mass lesion arising in the retrorectal space involving the sacrum (s3–s5), coccyx with pre-, paravertebral, intraspinal and posterior spinal components. Fat planes with the rectum well preserved.

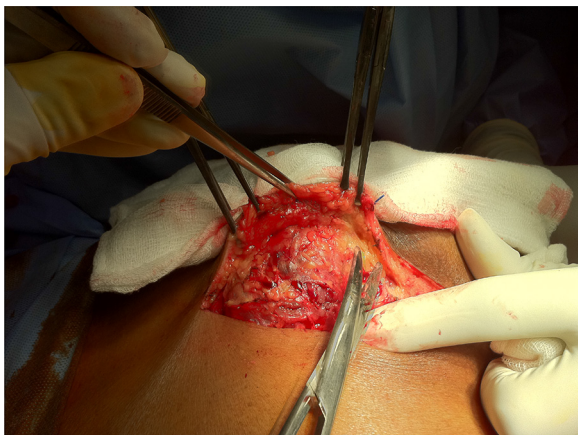


Fig. 2. A plane being created between the swelling and the skin.

5. Pathology

An irregular grey brown to grey black specimen with soft tissue and bony bits measuring 12 cm × 8.5 cm × 5 cm. Features suggestive of a *Chondroid chordoma*.

IHC – cytokeratin, s100 and EMA positive [Figs. 4 and 5](#).

6. Discussion

Tumours that occur in the retrorectal space comprise an uncommon and mixed group. They are estimated at one in every 40,000 hospital admissions. Retrorectal tumours may be classified as congenital, neurogenic, osseous or miscellaneous.⁴ Chordomas are solid malignant tumours that arise from vestiges of the foetal notochord, usually from within the vertebral bodies. As the spine develops, notochordal remnants are relegated to the intervertebral regions where they evolve into the nucleus pulposus. This explains why chordomas are in the midline or the paramedian location.⁵ The estimated yearly chordoma incidence is 0.5 cases per million inhabitants.

They account for 2–4% of all primary malignant bone tumours and are the most common primary malignant sacral tumour, with



Fig. 3. The excised specimen.

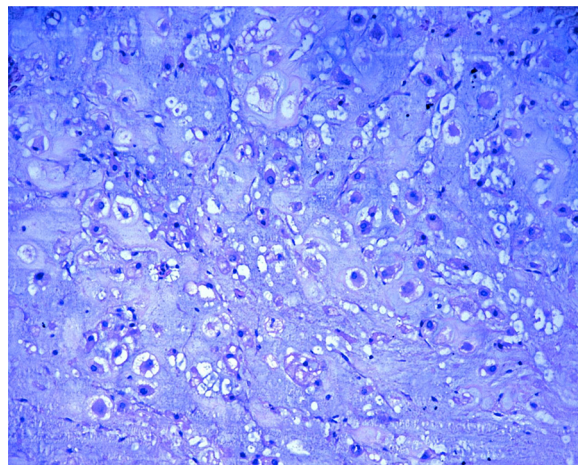


Fig. 4. 100×–200×–large cells with abundant vacuolated cytoplasm, a partially vesicular nuclei and few showing prominent nucleoli resembling physalliferous cells.

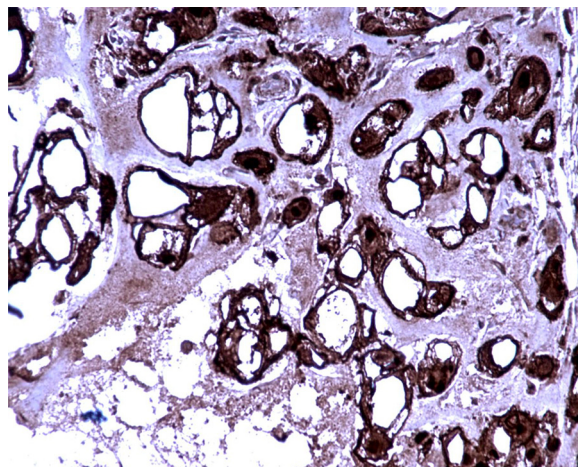


Fig. 5. Tumour cells positive for s100.

the exception of lymphoproliferative disease. They occur between 4th and 7th decade, peak in the 5th decade with a male to female ratio of 2–3:1.⁵

Clinical signs and symptoms may vary depending on the location, the size of the tumour and the extent of neural invasion. Symptoms of sacral chordomas are indolent and include pain, numbness, constipation, weakness and incontinence. Common

locations are the sacrococcygeus and the spheno occipital regions, anywhere along the vertebral column, not in the extremities.

Macroscopically, sacrococcygeal chordomas are usually well demarcated by a pseudocapsule. Most of them involve the bone and the surrounding soft tissue and skeletal muscle almost always. The cut surface of the tumour is characteristically soft, gelatinous, mucoid, and haemorrhagic.⁹ Histological features include a mixture of epithelioid and physaliferous cells. Chordomas are classified into classical or conventional, chondroid and de-differentiated types. Most common is chondroid type and de-differentiated type has the worst prognosis amongst them.

The appearance of localised growth could be misleading and soft tissue invasion is often missed in a CT for chordoma. Contrast-enhanced MRI is the gold standard in imaging. The combination of high T2 signal intensity and a lobulated sacral mass that contains areas of haemorrhage and calcification is strongly suggestive of a chordoma.⁵

The curative treatment of chordomas is en bloc surgical resection with negative margins. It is associated with long-term disease control and potential cure. But most often, due to the location, size and the extent of the tumour, margin free resection is not possible. The expected local failure rate in case of a marginal resection is 70%.⁶

Local recurrences, however, have often been observed even after total en bloc resection. Although the anterior approach is strongly recommended for sacral chordoma tumours, the posterior approach is adequate for total resection using retrorectal fat tissue as a cleavage line between tumour and rectum.

Despite surgical treatment, there has been a need to resort to radiotherapy in combination with surgery for positive or close margins and even as the definitive treatment modality for unresectable lesions. There are multiple modalities of RT such as proton beam radiation therapy, carbon ion radiation therapy, IMRT. At doses up to 40–60 cGy, however, local control at 5 years with conventional photobeam radiation therapy does seem to be in 10–40% range.^{7,8}

Chemotherapy has never played a role in the disease. Studies show that most patients responded to 800 mg Imatinib daily. The

possibility that an antiangiogenic effect maybe useful clinically is being explored.⁶

Prognosis depends on the resection of the tumour in surgery and postoperative treatment. Although metastases also may occur, most patients who succumb to the disease do so because of local recurrences. 5 year survival rate is 51% and 10 year survival rate is 35%.⁹

7. Conclusion

Chordoma is a slow growing, malignant tumour of the bone arising from the notochordal remnants. They occur more in men than in women. Surgical resection with negative margins is the treatment of choice. Radiotherapy is useful in cases with negative margins or when the disease is unamenable to surgical treatment. Chemotherapy is used as a supportive care. Prognosis depends on the surgical resection of the tumour with postoperative radiotherapy with or without chemotherapy.

Conflict of interest

Not applicable.

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Ethical approval

Not applicable.

Author contributions

Dr. Pragnya Chigurupati, assistant surgeon, primary author. Dr. Vishnukumar Venkatesan, co author. Dr. Vikram, primary surgeon. Dr. Kiran, Dr. Manuneethimaran, assistant surgeons.

Key learning points

- Chordomas are rare aggressive tumours from notochordal remnants.
- Enbloc excision with negative margins is the treatment of choice.
- Chemotherapy and radiotherapy do not have a definitive line of management in these tumours.

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