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Case Report

Sacroccocygeal Chordoma: A Case Report in a 52 Year Old Female

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ABSTRACT

Introduction: Chordoma is a rare slow- growing neoplasm, thought to arise from the cellular remnants of the notochord.
Presentation of case: A 52 year old female presented with a swelling in the sacrococcygeal region since last 6 months. MRI sacral region showed well defined, multilobulated, solid lesion having smooth regular margin surrounding the sacrum and coccyx. The lesion is iso to hypointense on T1W1 and hyperintense on T2.
Discussion: Sacroccygeal chordoma is a rare primary malignant tumour, and it is the most common primary sacral tumor. They are usually diagnosed late in the course of the diseases and can become quite large.
Management includes surgical resection of the tumour followed by radiotherapy. Despite these therapeutic modalities, recurrence is common.
Conclusion: The aim of presenting this case is its rarity. Surgical excision along with radiotherapy is the treatment of choice. Pre-operative imaging with MRI is essential to plan the most appropriate surgical approach.
Keywords: Sacrococcygeal chordoma, teratoma

INTRODUCTION

Chordoma is a rare malignant bone tumor that arises from ectopic rest cells of notochord. Restricted to the axial skeleton, it has a predilection for the sacrum (50%), sphenoid and occipital bones in the region of the clivus (35%), in the true vertebrae above the sacrum (15%). [1,2] In addition there have been reports of ectopic chordomas arising outside the skeletal axis, within the maxilla, sinuses, larynx and other soft tissues. Although chordomas are tumors of bone, they have also been reported at extra osseus sites, pre-dominantly in the paraphryngeal area. Sacral chordoma occurs almost twice as frequently in men compared with women and is uncommon in individuals younger than 40 years of age. [3]

Chordomas are typically slow growing but locally aggressive. Significant extra compartmental growth is often seen by the time of diagnosis. Most sacral chordomas present with anterior extension into the pelvis. [4,5] The tumor often displaces but does not invade the rectum, because the tough periosteum and presacral

fascia resist the transgression of disease. Metastasis is usually a late event. [6]

The relative rarity and slow growth of these tumors account for why most large clinical studies over many years have been based on cases collected involving variable treatments. Earlier studies [6] reported very high local recurrence rates for sacral chordoma treated with traditional surgical debulking and radiotherapy. These patients were condemned to progressive deterioration and pain from continued tumor growth, deteriorating neurological function, pressure sores, and infection. Local recurrence of sacral chordoma is associated with a 21-fold increased risk of tumor-related death.

Evidence suggestive of its origin is the location of the tumour (along the neuraxis), the similar immunohistochemical staining patterns, and the demonstration that notochord cells are left behind in the saccroccygeal region when the remainder of the notochord regresses during the fetal life.

CASE REPORT

Patient is a 52 year old female presented with a swelling in the sacral region since 6 months. Clinical examination showed a well nourished, vitally stable female. Local examination revealed a solid swelling in the sacral region, extending over the gluteal region, extending over the bilateral gluteal region measuring 10×7 cm. on digital rectal examination mass was palapable. Anal tone was normal. Perianal sensations were normal, no complains of urinary incontinence.

MRI of the lumbosacral spine showed well defined multilobulated, solid lesion, having smooth regular margins and circumferentially surrounding the sacro-coccyx bone. Intra operatively, under GA patient was put in prone position, an inverted U-shaped flap was elevated to expose the sacrococcygeal region, a large, bulky, lobulated, grayish- white, relatively avascular tumor seen. All superficial part excised. Coccyx was dissected down and excised. Partial lower sacrectomy done. Tumour anterior to the sacrum excised appeared to be of different characteristic. It was granular, and highly vascular. Incision was closed on negative suction drain.

Microscopically the tumours are characterized by a distinct, lobular architecture that is formed by the physaliphorous ((soap bubble) cells with ample vacuolated cytoplasm as well as by the presence of signet ring type cells. Broad spectrum antibiotics were given and care was taken to keep the dressing sterile. Careful monitoring of vitals was done. Post operative period was uneventful. Patient was discharged after 20 days and followed up in radiotherapy outpatient department for radiotherapy.

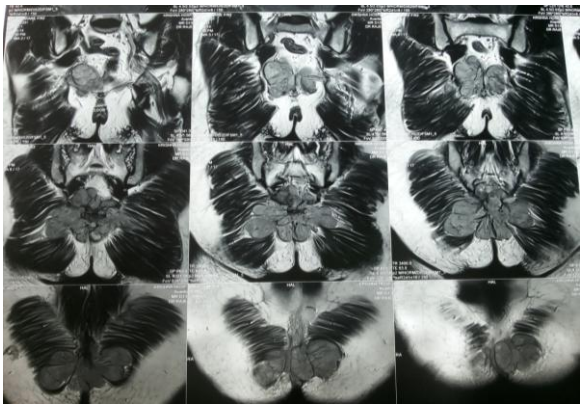


Fig 1 (MRI coronal section sacrococcygeal region)

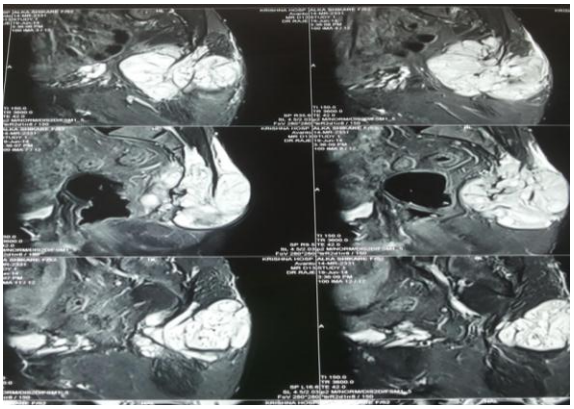


Fig 2 (MRI sagittal section sacrococcygeal region)

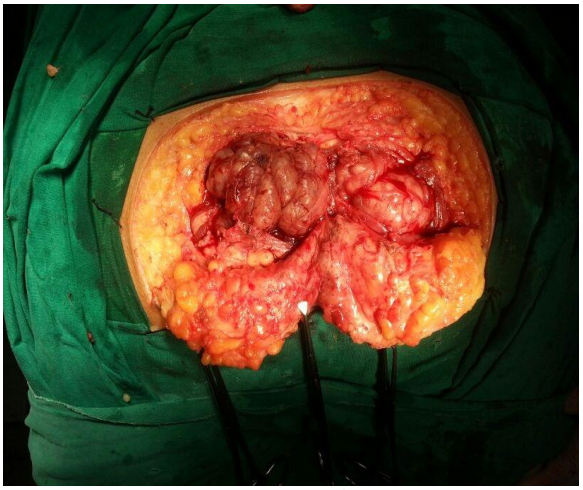


Fig 3 (Intra-operative gross specimen of the tumor)

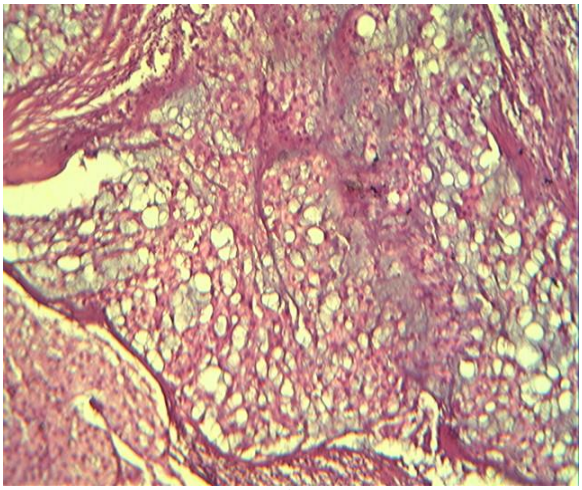
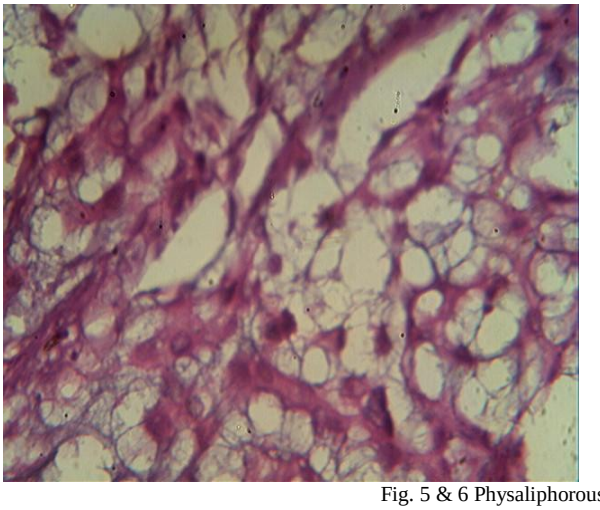


Fig 4 (Microscopic appearance of the excised tumour)



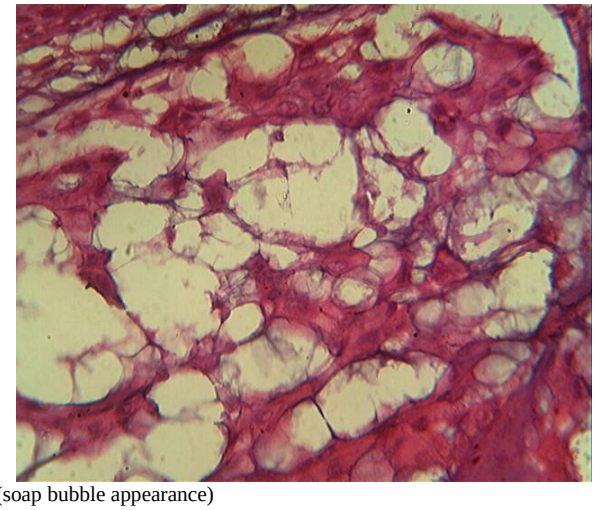


Fig. 5 & 6 Physaliphorous cells (soap bubble appearance)

DISCUSSION

Sacrococcygeal chordomas are malignant low-grade tumours, arising from remnants of the notochord. [3,7]

Because chordomas can develop insidiously, there can be a considerable time-lapse between the onset of symptoms and diagnosis

The conventional radiographic findings in sacrococcygeal chordomas are the central location of the tumour, its destruction of several segments of the sacrum and its appearance as an anterior soft-tissue mass that occasionally contains small calcifications. [8] MRI is the best imaging method for evaluating the extension of the tumour into soft tissue.

The reported rates of metastasis occurrence with chordomas vary considerably in the literature, from 5% to 70%. [3,7-11]

For the surgical treatment of chordomas, methods include the combined abdominal and trans-sacral approach [5] and the posterior approach. [13, 14] For proximal sacral lesions, the combined extended ilioinguinal and posterior approach is also useful. [15] The posterior approach is satisfactory for lesions at the third sacral segment or further caudal. Combined anterior and posterior exposure must be used for lesions that are located more cranially. [6,8,16-19]

The extent of surgical resection plays a major role in determining the length of the disease-free interval. [12,20] In patients who received a wide surgical excision, the recurrence rate was 17%, while in patients who received intralesional or marginal resection the recurrence rate was 81%. [9] It has been emphasized that if gluteal invasion is present, the risk of recurrence is higher, and therefore a wide posterior surgical margin is important. [2]

The risk of postoperative neurological deficits is related to the number of sacrificed nerve roots. [8] The loss of the S_4 and S_5 nerves is not associated with any significant urinary or bowel problems. [9] All of these potential consequences must be discussed with patients before surgery.

Transrectal biopsy is not recommended in chordomas due to the risk of the rectal wall and presacral fascia becoming contaminated with tumour cells. If transrectal biopsy is performed, then the rectum must be resected during chordoma surgery. [7]

For long-term control of sacral chordomas, the superiority of wide resection over subtotal or even marginal intralesional resection is well known. For the patients in our series, wide resection was not possible because the tumours were very big, invading neighbouring tissues, and most patients wanted their sacral nerve roots to be retained; therefore, their disease could not be eradicated. Chordoma is a locally aggressive but slow-growing tumour, so early diagnosis and wide resection increases the chances of long-term control.

CONCLUSION

The aim of presenting this case is its rarity. Complete surgical excision along with radiotherapy and chemotherapy, increases the chances of extended survival in patients of sacrococcygeal chordoma.

Preoperative imaging with MRI is essential to plan the most appropriate surgical approach.

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