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Case series:

Sacrococcygeal sarcoma – a case series

Harish Rao, Ashwin Pai, Muthukumaran, Rajesh Kumar Reddy, HS Shankar Ram

KMC Mangalore, Manipal University

Introduction

Chordomas are rare tumours. They are slow growing but locally aggressive neoplasms derived from primitive notochordal elements. Approximately 50 % originate in the sacral region, 35%in the clivus (Base of skull) and 15% elsewhere along the axial skeleton. These tumours rarely metastasize but local recurrence is common due to largely intralesional excision of the tumour. Therefore local irradiation becomes essential.

Here we present 3 patients with sacrococcygeal teratoma for whom wide local excision was done and are now on regular follow up.

Case 1:

We present a 54 yr old male patient, a manual labourer by occupation who presented with a swelling in the lower gluteal cleft since 3 months which was insidious in onset and progressive in nature. He consulted a local practitioner who performed an Incision and drainage 1 month back following which there was discharge from the wound site for 1 month.

No history of bleeding Per Rectum or pain. No sleep disturbance or loss of appetite or weight. His bowel, bladder habits were normal. There was no weakness or numbness of lower limbs .No other co morbidities .

On examination his vitals were stable. On Local examination there was a swelling about -7x5 cms in the gluteal cleft overlying the coccyx. It was firm in consistency, non-mobile. There was no local rise of temperature, no tenderness, no fixity to overlying skin but fixed to the underlying bone .Granulation tissue was present over Incision and drainage wound. Per Rectal examination was normal. Proctoscopy was normal. There were no Focal Neurological deficits.

Patient was evaluated with FNAC of the swelling which showed the following

Pleomorphic cells in dyscohesive clusters and singly scattered .Large round cells having abundant vacuolated cytoplasm (physaliferous cells) and round vesicular nuclei, prominent 2-3 nucleoli with intranuclear inclusions.

Many binucleate, trinucleate and multinucleate cells were noted The cells are interspersed in a myxoid background and an Impression that features are suggestive of Sacrococcygeal Chordoma was given.An MRI scan was done to confirm the same and to know the plane of the swelling. Patient underwent surgery through posterior sacral approach. The tumour was found attached to the coccyx. The specimen was then sent for histopathological examination which showed the following. Well encapsulated, attached fibro fatty tissue, soft to firm in consistency. Lobules of large cells with eosinophilic to multivacoulated cytoplasm- Physaliphorous cells Occasional atypical mitoses seen . Tumor infiltrating the capsule. Focal myxoid change with areas of haemorrhage and necrosis.

The surgical approach to this case was through the posterior sacral approach. The main concerns during the surgery were injury to the rectum and bleed from the presacral plexus. Incomplete excision was one other concern which had to be dealt with.

Radiation was considered to counter this risk of incomplete excision and recurrence.

Case 2:

A 46 year old female presented with low back ache which was not subsiding on medications .It was persistent for the last 4 months. She also complained of constipation. On examination a hard mass was felt in the posterior wall of the rectum with the rectal mucosa freely moving over the mass. CT scan confirmed the lesion to involve from S4 to the coccyx. She underwent wide local excision followed by an uneventful recovery. She is currently on regular follow up and is being considered for radiotherapy.

Case 3:

A 50 year old male presented with difficulty in defecation and pain in the lower back near the coccyx .He was treated for coccydynia and constipation. As the symptoms did not subside he was referred to our hospital. On examination a mass was felt in the posterior wall of the recto anal region. The mucosa was not involved. An MRI scan was done which suggested a sacrococcygeal mass lesion. Surgery was performed and the mass was removed in toto with a piece of the sacrum and coccyx. The mass was found abutting the rectal wall but not infiltrating through it which made excision

difficult. Therefore radiation is being considered for this patient.

Discussion

The notochord develops at the 4th week of intrauterine life and extends from the occipital bone to the coccyx [17]. It regresses in the 7thweek, following the formation of vertebrae. During the grouping of vertebrae, the notochord tissue may remain and it is from this residual tissue that the chordoma arises [3, 8]. Chordomas are divided into three groups depending on their origin from the axial skeleton: 1, cranial; 2, vertebral; and 3, Sacrococcygeal chordomas. Fifty per cent of chordomas are sacrococcygeal, 35% cranial, and the remaining 10%-13% vertebral [14].

Chordoma is the most frequent primary malignant tumour of the sacrococcygeal region [7]. These tumours can be seen in any age group, but cranial chordomas occur more frequently at earlier ages (average 30--40 years). The average age of onset is 40-50 years for vertebral chordomas, and 50 to 60 years for sacrococcygeal chordomas [5, 9]. These kinds of tumour are reported 2 to 3 times more frequently in men than in women. The mean age being 40-50 years and male predominance were reported by Dahlin and McCarty [5].

The earliest and the most frequent symptom of the Sacrococcygeal Chordoma is sacral pain which extends to both legs. Initially, the pain is discontinuous and not severe. It usually starts 6 months to 1 year before the tumour is seen. If the tumour invades the sacrum and the roots of the nerves, there is intractable pain localized at the anal region. Therefore, the complaints may be misdiagnosed as haemorrhoids, pilonidal sinus, and intervertebral disc disease. However, rectal and urinary complaints may occur at a later stage. The most important complaints encountered are low back pain, constipation, and mass. Urine or faecal incontinence, intestinal obstruction, bleeding from the rectum and bladder are seen at the last stage of the disease [3]. The tumour may cause gluteal ulcers by way of invading the sensitive nerves. In addition, trophic deformation of the feet may develop.

On physical examination, the tumour is felt between the rectum and the sacrum. Characteristically, it is quite soft, flat, and sometimes lobular. In our experience, mucosa can easily move on the tumour. The tumour may cause displacement of the bladder and the pelvic organs. If the tumour lies in front of the sacrum, it pushes the rectum forward. Depending on its size and the nerve roots infiltrated, there may be urinary complications such as pollakiuria, dysuria, and hydronephrosis. It is said that most of the time the tumour in the pelvis has a tendency to grow unilaterally [5].On proctoscopy, the rectal mucosa is always found to be normal [4]. In general, sacrococcygeal chordomas cause metastasis on regional lymphoid glands, but visceral organ metastases are seen less frequently [21].Bone destruction is the most prominent finding in the chordomas [10]. They show reduced uptake or a cold lesion on bone scintigraphy [19]. In advanced cases, the sacrum can be completely destroyed. In wide lytic areas, there may be amorphous calcifications [5]. On the other hand, osteoblastic chordomas are reported less frequently [17]. Within the pelvis, the contour of the tumour can be detected on radiological examination. Primary bone tumours cause extensive destructive changes and palpable masses. Because of their radiological appearance, the chordomas may be confused with chondroma, chondrosarcoma, and giant cell tumour. These primary bone tumours lead to advanced bone erosion and palpable masses.

All of the presacral tumours are included in the differential diagnosis: metastatic bone tumours, anterior meningocele [10], tuberculosis, pyogenic infections, hydatid cyst [22], sacrococcygeal teratoma, and tumors of female pelvic organs.

CT provides adequate demonstration of the bulk of the tumour as well as the extent of sacral destruction [11].MRI offers an advantage as it demonstrates the soft-tissue component of the tumour [16].It seems likely that in the near future, progress in MRI technology will make it the method of choice in evaluation of these lesions.

Moreover, rectal cancers and sarcomas of soft tissue should be considered in the differential diagnosis. As a rule, chordomas should be diagnosed histologically rather than radiologically. In the cross-section of chordomas with a solid structure, cystic, haemorrhagic, and necrotic areas are seen together. In the microscopic structure of a well-differentiated chordoma, cuboid epithelial tissues invading the regular cavities are found. Typically, tumour cells are large, stretched, and vacuolar. However, a specific diagnosis can be made when these mucinous intercellular matrix and tumour cells are present together [17]. Recently, cytogenetically aberrant chordomas have been reported by Martens et al. [13].

The first thing to be considered for the therapy of the chordomas is the possibility of surgical removal of the tumour. As reported by Gennari et al. [7], both the margins of surgical removal in the sacrum and the preservation of sacral roots present major surgical problems. Most of the surgeons recommend removal of the tumour, even if the

margin is inadequate, with preservation of both S2 roots [1, 2, 7, 12, and 18]. Even an inadequate operation will reduce the amount of pain and remove some of the other symptoms.

Besides, the only therapy available for patients with a smaller tumour volume and for those who are inoperable is radiotherapy with super voltage or cobalt-60 [15]. Most of the time, radiotherapy is applied in the form of a sandwich, from front and behind or, depending on the localization, from the sides, as a supplementary measure under super voltage conditions [14].

Conclusion

Chordomas though rare need to be considered as a differential diagnosis in mid line swellings in the sacrococcygeal region, the skull base and axial skeleton. Wide local excision followed by radiotherapy is adequate for this locally aggressive tumour. Risk of recurrence is high and needs to be explained to the patient.

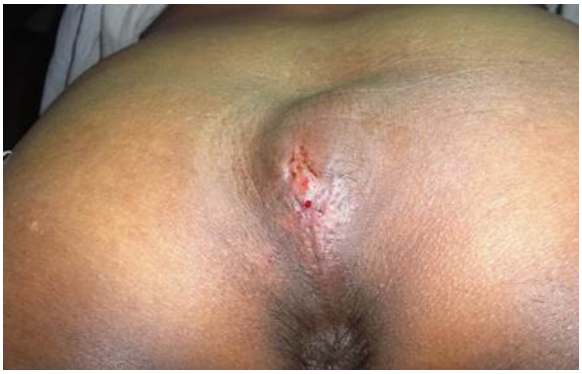

Fig 1 – Swelling in the coccygeal region

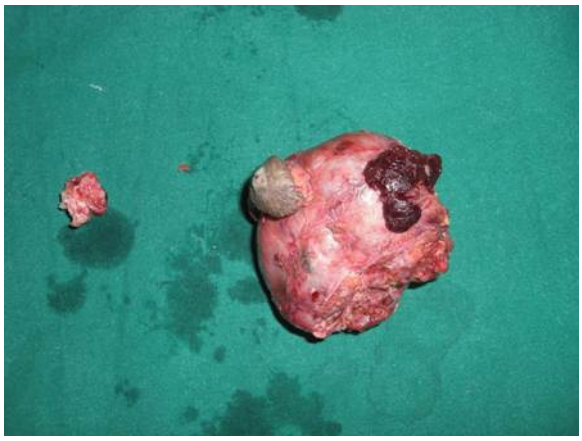

Fig 2 – Gross specimen after excision

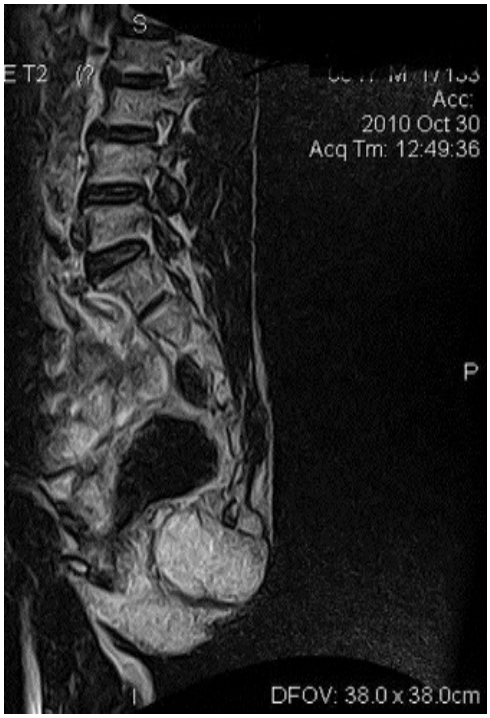

Fig 3 – MRI showing a swelling in the sacrococcygeal region abutting the rectum

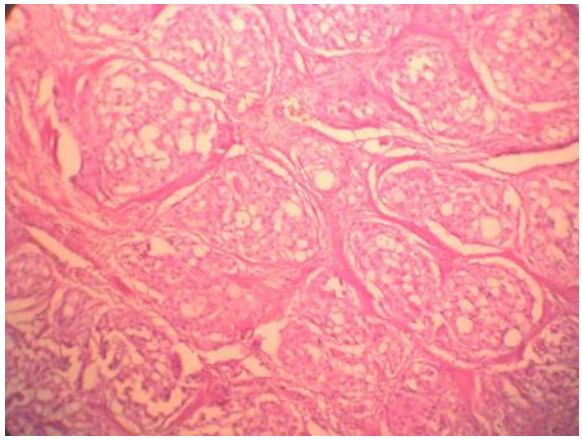

Fig 3 – Slide picture showing large cells with eosinophilic to multivacoulated cytoplasm- Physaliphorous cells

1.Anson KM, Byrne PO, Robertson ID, Gullan RW, Montgomery AC.Radical excision of sacrococcygeal tumours. Br J Surg 1994; 81:460
2. Azzarelli A, Qnagliuolo V, Cerasoli S, Zucali R, Bignami P, Mazzaferro V, Dossena G, Gennari L.Chordoma: natural History and treatment results in 33 cases. J Surg Oncol 1988;37 :I85
3. Beagie JM, Mann CV, Butler CB. Sacrococcygeal chordoma.Br J Surg 1969;56 : 586
4. B ethke KP, Neifeld JP, Lawrence W Jr. Diagnosis and management of sacrococcygeal chordoma. J Surg Oncol 1991;48:232
5. Dahlin D, Maccarty CS. Chordoma: a study of fifty nine cases. Cancer 1974;5 : 1170
6. Fleming GF, Heimann PS, Stephens JK, Simon MA, Ferguson MK, Benjamin RS, Samuels BL. Dedifferentiated chordoma. Response to aggressive chemotherapy in two cases. Cancer 1993;72:714
7. Gennari L, Azzarelli A, Quagliuolo V. A posterior approach for the excision of sacral chordoma. J Bone Joint Surg[Br] 1987;69 : 565
8. Heffelfinger MJ, Dahlin GG, Maccarty C, Beatbuth JW. Chordoma and cartilaginous tumors at the skull base. Cancer 1967;32:410
9. Higinbotham NL, Phillius RF, Farr HW, Hutso HO (1967) Chordoma-thirty five year study at Memorial Hospital. Cancer 20:1841
10. Karmin RF, Potanos JN, Pool JL. An evaluation of chordoma.J Neurol Neurosurg Psychiatry 1964; 27 : 157
11. Krol G, Sundaresan N, Deck M (1983) Computed tomography of axial chordomas. J Comput Assist Tomogr 7 : 286
12.Maccarty CS, Wauhg JM, Coventry MB, O'Sullivan DC. Sacrococcygeal chordomas. Surg Gynecol Obstet.1961; 113:551
13.Mertens F, Kreicbergs A, Rydholm A, Willen H, Carlen B,Mitelman F, Mandahl N. Clonal chromosome aberrations in three sacral chordomas. Cancer Genet Cytogenet.1994; 73 : 147
14. Mindell ER. Chordoma. J Bone Joint Surg [Am] 1981;63:501
15. Pearlman AW, Friedmann M. Radical radiation therapy of chordoma. Am J Roentgenol Radium Ther Nucl Med1970; 108 :333
16. Rosenthal DI, Scott JA, Mankin HJ, Wismer GL, Brady TJ. Sacrococcygeal chordoma: magnetic resonance imaging and computed tomography. AJR 1985;145 : 143
17.Russel DS, Rubinstein LJ (Ed) Pathology of tumours of the nervous system. London, Edward Arnold, 1989 p 820
18. Samson IR, Springfield DS, Suit HD, Mankin HJ Operative treatment of sacrococcygeal chordoma. J Bone Joint Surg[Am] 1993;75 : 1476
19. Suga K, Tanaka N, Nakanishi T, Utsumi H, Yamada N. Bone and gallium scintigraphy in sacral chordoma. Report of four cases. Clin Nucl Med 1992;17 : 206
20.Tewfik HH, Cinnis MC, Nordstrom DG, Latourette HB.Chordoma: evaluation of cranial behaviour and treatment modalities. Int J Radiat Oncol Biol Phys 1997;2 : 959
21. Yarom R, Horn Y.Sacrococcygeal chordoma with unusual metastases. Cancer 1970;25 : 259
22. Yegen C, Ozer AF, Aktan AO, Yahn R Sacrococcygeal hydatid cyst: another entity in the differential diagnosis of sacrococcygeal chordoma, case report. Paraplegia 1993; 31:479

Corresponding Author
ASHWIN PAI MS(Sur) MRCS (Edin) FAGE
Asst Prof of Surgery, KMC Mangalore , Manipal University.
#13-10-1336, "Gopal Bhavan",
Near Mahamaya Temple ,
Field Street,
Mangalore-575001, Karnataka, India
Phone: +91-9886661017
Email: drashwinpai@gmail.com