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CASE REPORT

Chordoma Of Lumbar Spine

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Abstract: Chordoma, lesion derived from the notochord, represents about 4% of the primary malignant bone tumours. Males are affected more commonly than females and it is very rare in children;the peak incidence is in the sixth decade of life.The sacrococcygeal region accounts for 50% of cases and the spheno-occipital region or the base of the skull for 37% of cases.The remainder of the cases reported occur in the descending order of frequency in the cervical,thoracic and the lumbar spine.It is found to be rarely involving the lumbar spine(about 2%).We present a case of chordoma involving L_4 and L_5 vertebral body and disc.The role of epithelial cell marker study has proved a well adjunct to the histopathological diagnosis of chordoma.

Introduction

Chordomas are rare representing 4% [1] of all malignant tumours of the bone. It can occur at any age but usually develops in adults. Most of them are seen at the base of the skull (clivus) and sacrococcygeal region. However,it is found to be rare involving the lumbar spine (about 2%) [2].We present a case of chordoma involving L_4 and L_5 vertebral body and disc mimicking metastasis clinically and aneurysmal bone cyst radiologically.The lesion occuring in the lumbar spine is rare.

Case History

A 62 year old male presented with progressive backache and left sciatica since 1 year. Routine haematological and biochemical investigations were normal. Magnetic Resonance Imaging (MRI) of the lumbo-sacral spine revealed $L_{4/5}$ disc with a diffuse bulge and posterocentral protrusion indenting the thecal sac and causing bilateral neural foraminal narrowing with signal changes in the posterial aspects of L_5 vertebral body associated with anterior epidural soft tissue significantly indenting the cauda equina - ? Atypical haemangioma, ? Metastasis with Lumbar spondylosis and diffuse bulge of $L_{4/5}$ disc with posterocentral protrusion.

Decompression with biopsy from lesion (L4 vertebral level) was done.

Histopathological study: Macroscopically, the specimen consisted of multiple, gelatinous, greybrown soft tissue and bony fragments. Microscopically, the sections showed tumor tissue composed of cords and islands of pleomorphic cells arranged in lobular pattern. Sheets of cohesive epithelioid cells having small hyperchromatic nuclei and abundant cytoplasm were seen. Numerous polygonal cells with abundant intracytoplasmic mucin (physaliferous cells) were also seen. The background showed pale mucous material (Figs. 1 & 2).The sections showed positivity for PAS stain (Fig.3).The features were those of chordoma.

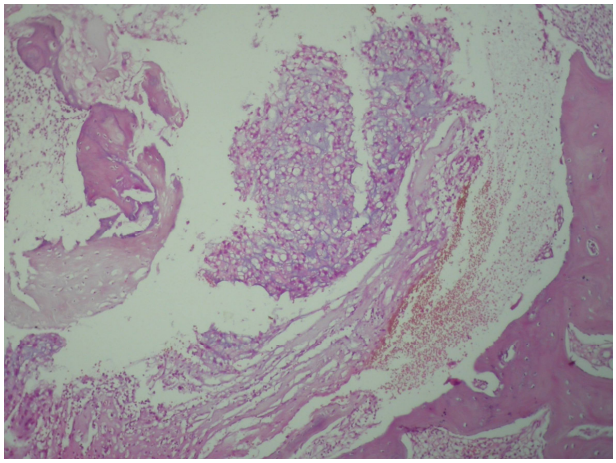


Fig.1:Showing cords and trabeculae of epithelial cells in the mucoid stroma with osseous spicules (H&E, 100X)

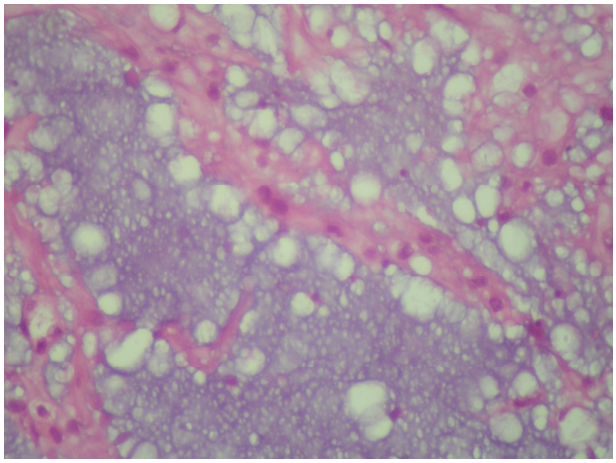


Fig 2: Showing highly vacuolated physaliferous cells. (H&E, 400X)

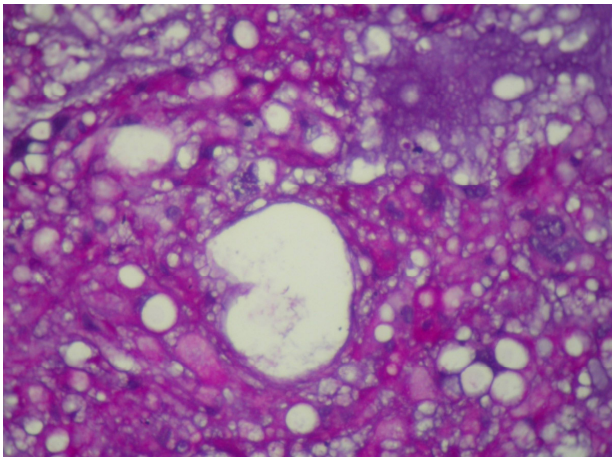


Fig 3: Showing PAS positive physaliphorous cells. (H&E, 400X)

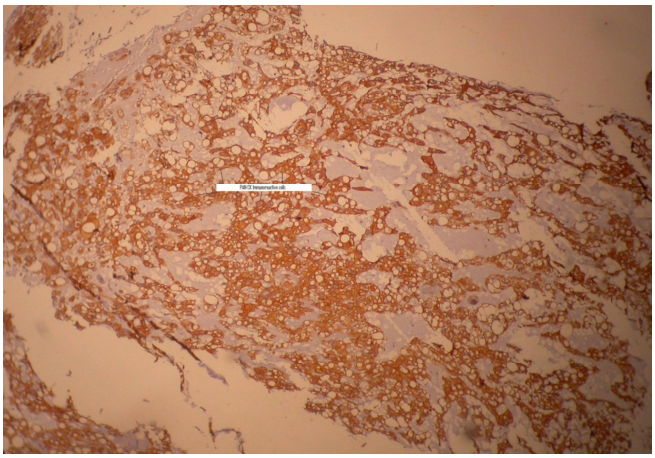
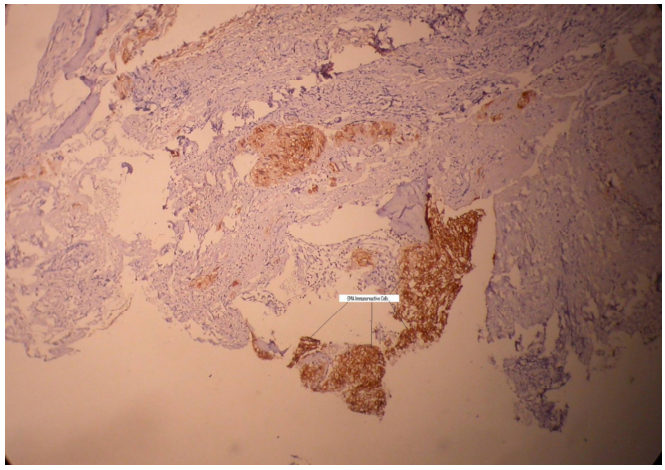


Fig 4: PAN CK immunoreactive cells. (100x)

Fig 5: EMA immunoreactive cells (100x)



Immunohistochemistry (IHC) revealed strong positivity for Pancytokeratin in the cytoplasm of tumor cells and Epithelial Membrane Antigen (EMA) in the tumor cell membrane and confirmed the diagnosis of CHORDOMA.

Discussion

Spinal chordoma accounts for 3-4% of all primary bone malignancies involving spine. The incidence is approximately 0.5 per million inhabitants [3]. The M/F ratio is 2:1 for spinal chordomas. About 15% of chordomas are observed in mobile vertebral column and more than one vertebral body may be involved [4]. As far as the embryogenesis is concerned, during the 4^{th}-6^{th} week of foetal development, a group of cells congregate together to form a structure called the notochord. The notochord defines the vertical midline of the body and spinal column develops around it. Normally, as development progresses, the notochord degenerates and disappears except for small bits of tissue becoming part of the discs (nucleus pulposus) between the spinal vertebrae.Chordoma is believed to develop from pieces of notochord that for some reason do not breakdown as they should. Over many years, these harmless bits of notochord transform and become malignant forming chordoma [5]. In the present case, L_4 and L_5 of vertebral bodies were involved.

Chordoma is a slow growing lesion and its symptoms are closely related with the localization of the tumor. These include compression to the root, spinal cord and paravertebral tissues. Compression of anterior column and root is predominantly seen; parasthesia and pain are the most common complaints in patients with lumbar chordomas. It is usually confused with more common tumors of the lumbar spine such as aneurysmal bone cyst, giant cell tumor, haemangioma, myeloma and metastasis. The present patient complained of low backache and left sciatica since 1 year with dragging pain in the left leg since six months.

The Criteria for Diagnosis of Chordoma: Lobulation is almost a *sine qua non* for the diagnosis of chordoma. Physaliferous cells are difficult to find at times and failure to identify them especially on a small biopsy specimen does not negate the diagnosis of chordoma.The role of special stains such as reticulin and glycogen in differentiating adenocarcinoma and chondrosarcoma are of little value because the reactions are similar.

Immunohistochemistry has proved a well adjunct to the diagnosis of chordoma, especially when chondrosarcoma is suspected and in distinguishing the presence of chondroid matrix in chordomas. Thus chordomas express epithelial markers such as cytokeratin & EMA, none of which are present in chondrosarcoma [6]. Differentiation from a metastasizing epithelial neoplasm can be difficult at times, because chordomas share epithelial markers with carcinomas [7]. Chordomas, however, express Vimentin, Fibronectin , S-100 protein & alpha-fetoprotien [8-9]. The most mucus producing carcinomas do not express these markers. The present case was positive for pancytokeratin, EMA and also S-100 protein. The presence of chondroitin sulfate immunoreactivity for vimentin and S-100 protein and areas of cartilaginous differentiation (Figs. 4,5) indicate a relationship both to chondromatous tumors and to normal notochord from which chordoma is believed to originate.

Conclusion

Lumbar spine is an uncommon localization for chordoma and the present case involving L_{4-5} vertebral bodies is reported. With the aid of modern imaging techniques and emphasis on immunohistochemical perspective study make the possible diagnosis of spinal chordoma (lumbar) pre-operatively. The immunohistochemical study confirms the utility of these markers in the differential diagnosis of chordoma and other tumors with similar histologic characteristics.

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