



Case Report

Chondroid chordoma- A rare tumor diagnosed on cytology: A case report

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ABSTRACT

Chordomas are rare, malignant and locally aggressive tumors that are derived from the remnants of primitive notochord, out of which chondroid chordomas are even rarer. Very few case reports have described this variant, which is difficult to pick up on cytopathology alone and has a number of other differentials too. We report here a case of chondroid chordoma at the sacrococcygeal region that was diagnosed on FNAC in an elderly male patient emphasizing on its cytomorphology and how to differentiate from its cytological mimickers.

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1. Introduction

Chordomas are rare, locally aggressive malignant bone tumors arising from the primitive notochord remnants that account for 1%–4% of all malignant primary bone tumors.¹ Most common sites are sacrococcygeal and sphenooccipital regions, followed by cervico thoracic and coccyx, though they can occur anywhere along the axial skeleton.^{2,3} Extra axial sites and soft tissue involvement are also reported.⁴ Though the most common age group involved are between 5th to 6th decades, person of any age can be involved. Amongst the axial Chordomas, in adults they are commonly seen in sacrococcygeal region and in children in the sphenooccipital region.⁵

Histologically, it is of three types, conventional [NOS], chondroid and dedifferentiated. Chondroid chordoma is a very rare variant that accounts for about 14% of all chordomas and is said to have a better prognosis than conventional chordoma.⁶ Though there are a few case reports describing Chondroid chordomas on histopathology,^{6–12} case reports of these diagnosed on cytology are hardly present.^{13,14} Hence, we present here a case of sacrococcygeal chondroid chordoma emphasizing on the cytopathological features and

its differential diagnosis as well as how to differentiate between them.

2. Case Report

A 65 years old male presented to the Out Patient Department of our institute with a gradually increasing painless lump on the sacrococcygeal region for 2 years. There were no any other associated complaints.

On clinical examination, swelling was 4 x 4 cm in size, non-tender, non-mobile and firm to hard in consistency. Overlying skin was normal [Figure 1]. CT scan showed a large heterogenous hyperdense lobulated mass in the sacrococcygeal region with intraspinal extension as well as extension anteriorly to the pelvic cavity and laterally to bilateral ischioanal fossae and pelvic sidewalls with multiple calcific foci. Infiltration to right multifidus and gluteus medius was also present [Figure 2].

Fine needle aspiration cytology was performed and stained with May Grunwald Giemsa and Pap stains. Microscopically, smears were moderately cellular and showed abundant myxoid stroma with focal areas of chondroid matrix that were encircling the tumour cells which were large in size, moderately pleomorphic, had round to oval nuclei, inconspicuous to prominent nucleoli

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Fig. 1: Sacrococcygeal lump



Fig. 2: CT scan film showing large heterogenous hyperdense lobulated mass in the sacrococcygeal region

and abundant bubbly cytoplasm (Physaliphorous cells). Few bi and multinucleated cells were also seen. Clusters of epithelial cells and cells with signet ring like morphology were also noted at many places [Figure 3].

On the basis of cytomorphology, a differential diagnosis of Chondroid chordoma and Chondrosarcoma was thought of. Patient was advised for biopsy and histopathological examination. We received whole of the tumor as multiple grayish white pieces of soft tissue, largest and smallest measuring 8 x 6 x 3 cm and 2.5 x 2.5 x 1 cm

respectively, along with few bony pieces. Extensive sectioning of the tumor tissue was done and submitted for processing. Histologically sections showed tumour cells arranged in lobules separated by fibrous bands. These tumour cells were large in size with round nuclei and prominent nucleoli with abundant vacuolated cytoplasm (physaliphorous cells) in a prominent chondroid and chondromyxoid background. Admixture of these tumour cells with smaller cells having moderate amount of eosinophilic cytoplasm (Epithelial cells) was also seen at many places. Occasional multinucleated tumour giant cells and at places signet ring like cells were also noted. Mitotic activity was not high. Sections examined from bony tissue showed invasion by the tumour cells [Figure 4]. We went ahead with the two differentials that we had made during cytological examination, i.e. Chondroid chordoma and chondrosarcoma and advised IHC panel of EMA, CK8, CK19 and Brachyury for confirmation. All of these markers turned out to be positive confirming the diagnosis of Chondroid Chordoma [Figure 5]. Following surgery our patient underwent 25 fractions of external beam radiotherapy for 5 weeks followed by oral Imatinib. At the time of writing this paper, patient was responding well to the treatment.

3. Discussion

Chordomas are locally aggressive rare malignant tumours originating from embryonic notochord vestiges that account for 1-4% of all primary malignant bone tumors, out of which maximum cases are seen in the sacrococcygeal region (50%) followed by sphenoccipital region(35%) and rest in vertebral region (15%).¹ Incidence estimated annually is around 0.1% per 1 lakh population. The mean age involved are 5th-6th decades of life, with male to female ratio of 2-3:1, though they can be seen at any age.² They can occur anywhere along the axial skeleton, with the sphenoccipital involvement more commonly reported in children and young adults and the sacrococcygeal involvement in the elderly.³ Few case reports have mentioned them in extra-axial skeleton and soft tissues as well.⁴

Three histologic types, conventional, chondroid and dedifferentiated have been described, amongst which chondroid are believed to have better and dedifferentiated to have worse prognosis than the conventional ones.⁵

The concept of chondroid chordoma was first proposed by Heffelfinger et al. in 1973. Since then, there are few case reports where this entity has been described. Most of these have mentioned the histopathological features⁶⁻¹² with only a handful of them mentioning the cytopathology.^{13,14}

On cytopathology, though conventional chordomas are easy to diagnose because of presence of typical physaliphorous in a background of abundant myxoid matrix, diagnosis of chondroid chordoma is very tricky because of simulation with other tumors like chondrosarcomas,

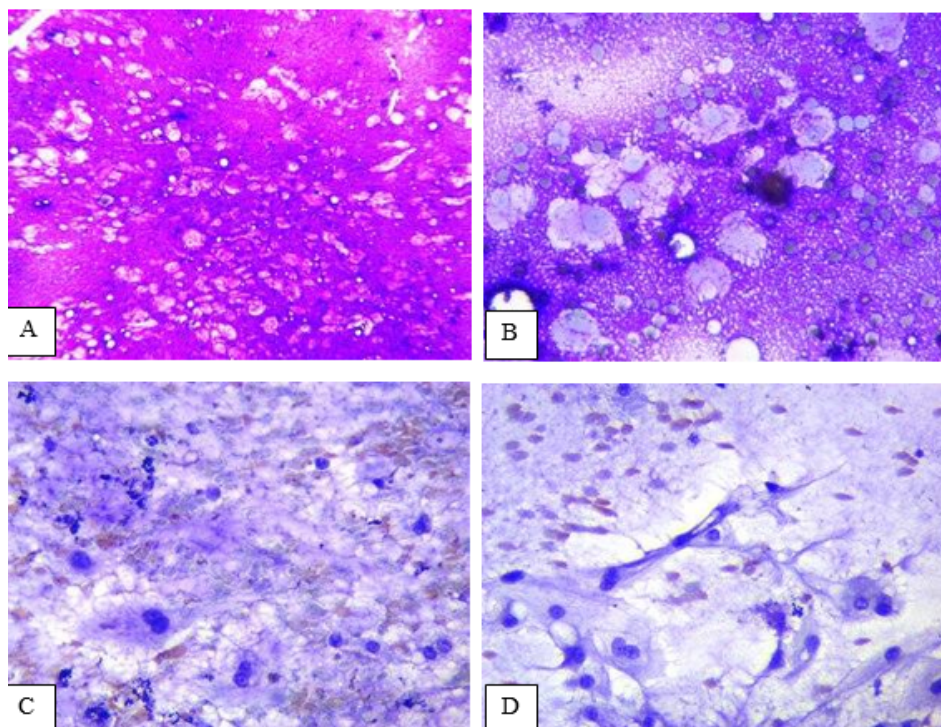


Fig. 3: **A):** Large cells with bubbly cytoplasm in chondroid background (physaliphorous cells), MGG 10x; **B):** Physaliphorous cells with bubbly cytoplasm, MGG 40x; **C):** Binucleated cell in myxoid background, Pap (10x); **D):** Epithelial cells with few signet cells in myxoid background, Pap (40x)

chondromas and chordoid meningiomas.

In Chondroid chordoma mixture of both chordoid and chondroid cells are seen. Frequently epithelial cells and bi-multi nucleated cells are also noted, as was seen in this case. In chondromas, few free lying monomorphic cells with indistinct cell borders and weakly stained cell nuclei are seen lying within lacunary structures in a background of purplish thick myxoid material. Chondrosarcomas show cells which are pleomorphic, hyperchromatic and have multiple prominent nuclei, the cells may be vacuolated but rarely present with the large cytoplasmic vacuoles as seen in chordoma where it is a characteristic feature.¹⁴ Chordoid meningiomas contain cords of polygonal tumor cells with bland nuclei, nuclear pseudoinclusions and vacuolated eosinophilic to clear cytoplasm resembling physaliphorous cells in mucin rich stroma (myxoid stroma). However, occasional loose pink purple cells without cytoplasmic vacuoles and admixed lymphoplasmacytic infiltrates are helpful identifying features.¹⁵

On histology, Chondroid Chordomas show the typical morphology of chordomas [physaliphorous cells admixed with epithelial cells] along with a focal /extensive cartilaginous background, as was seen in this case. These are generally low grade tumors bereft of the pleomorphism and high mitotic activity that are usually seen in Chondrosarcomas. Chordoid Meningiomas usually show

areas of typical meningiomas along with the chordoid areas. Chondromas are rare in the pelvis and usually hypocellular, but may sometimes be cellular and cytologically atypical. Areas of ischemic necrosis are frequently seen.

With the help of Immunohistochemistry [IHC], these close differentials can be distinguished with high degree of certainty. IHC demonstrates that tumor cells of chordoma are reactive to epithelial markers like Epithelial Marker Antigen (EMA) and Cytokeratins (CKs) especially CK 8 and CK 19, whereas Chondromas and Chondrosarcomas are negative for CKs and EMA and are positive for Vimentin. Chordoid meningiomas are positive for EMA and variably positive for S100 protein but negative for cytokeratins. Brachury has emerged as the most sensitive as well as specific maker for chordomas as it is positive only for this and negative in the rest. However, it's important to note that the positivity for all these IHCs is lost in the dedifferentiated chordomas.

Because of an indolent nature of the chondroid chordoma, by the time the diagnosis is made, tumor usually attains a large size with extensive local spread as was seen in this case too where there was intraspinal extension as well as bony infiltration. Prognosis depends on the extent of spread the treatment option chosen. Since, Chordomas are reported to be usually resistant to chemotherapy, the best treatment remains complete surgical excision in blocks along with

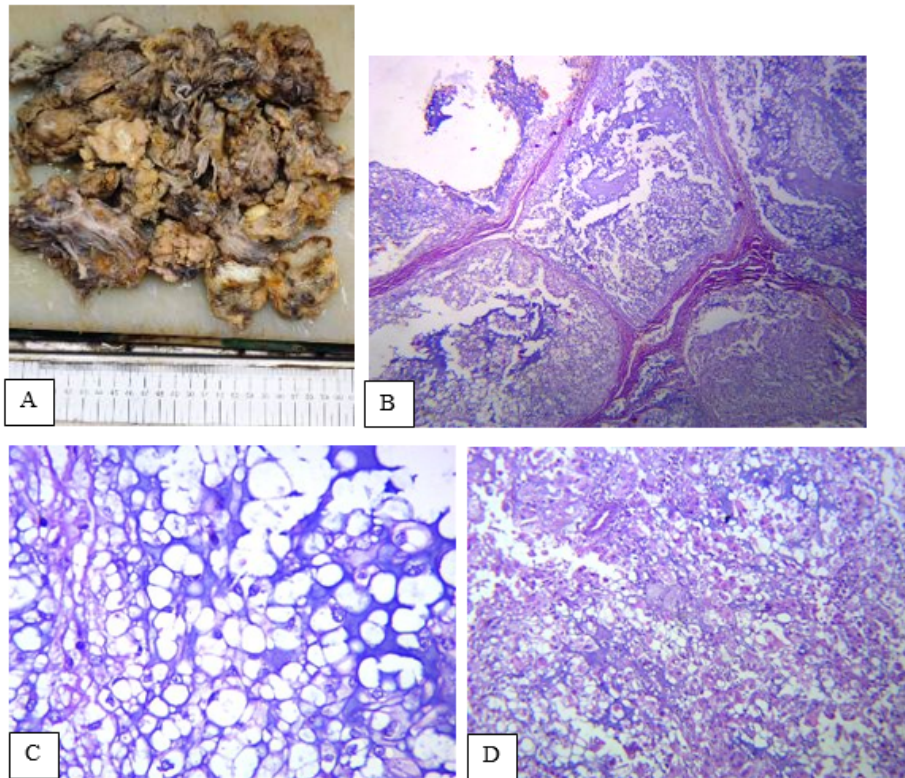


Fig. 4: **A):** Gross specimen, multiple greyish white pieces of tissue; **B):** Lobulated tumor pattern with chondroid and chondromyxoid areas, H&E (10x); **C):** Extensive physaliphorous cells in chondroid background H&E (40x); **D):** Admixture of physaliphorous cells and epithelial cells with eosinophilic cytoplasm, H&E(10x)

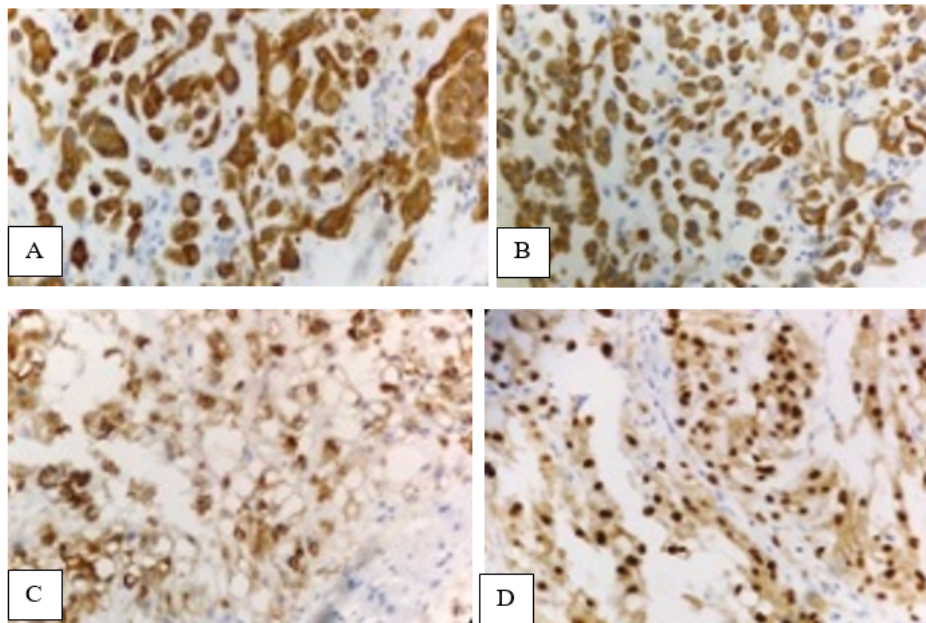


Fig. 5: **A:** Strong cytoplasmic positivity for CK 8 (IHC), 10x; **B):** Strong cytoplasmic positivity for CK 19 (IHC), 10x; **C:** Strong membranous positivity for EMA (IHC), 10x; **D:** Strong nuclear positivity for brachyury (IHC), 10x

adjuvant radiotherapy by various modalities, out of which Proton beam therapy has proven to be the most effective. The probability of recurrence remains high even after total resection, with the reported recurrence rate as high as 68% and the rate of distant metastasis is reported between 5%-29%.⁶

4. Conclusion

Chondroid chordomas are a very rare type of chordomas which are said to have a better prognosis than the conventional chordomas. Diagnosing them preoperatively by FNAC is very important because of its different prognosis and treatment approach with respect to those tumors with which its cytomorphological features overlap. A high index of clinical suspicion and an extremely cautious and well trained cytopathologist can clinch the diagnosis.

5. Source of Funding

None.

6. Conflict of Interest

None.

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