Chondroid Chordoma: An Uncommon Bone Neoplasm

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Abstract

Chordomas are rare, slowly growing, locally aggressive neoplasms of bone that arise from embryonic remnants of the notochord. Chondroid-chordoma is an uncommon variant of chordoma. These tumors typically occur in the axial skeleton. Here we report a rare occurrence of chondroid variant of chordoma at sacrococcygeal region in a 60 year old female. Magnetic resonance imaging revealed expansile mass involvingsacrococcygeal region.Gross and microscopic examination confirmed chondroid chordoma.Later patient underwent radiotherapy, doingwell,on a one year follow up. Chondroid chordoma is discussed due to its predilection for occurrence at sacrococcygealregion and its more favorable prognosis compared with that of conventional chordoma.

Keywords: *Chondroid-chordoma, notochord, sacrococcygeal.*

Introduction

Chordoma is rare tumor that grows slowly,locally aggressive, invades surrounding soft tissue and metastasizes infrequently. It accounts for 1%– 4% of all primary malignant bone [(1,2)] Chordoma usually occurs in the sacrococcygeal (50%), spheno-occipital region (35%) and (15%) in the true vertebrae. [(3,4)] Nearly, 33% chordomas in the base of the skull are chondroid in type. [(5)] It is only malignant tumor arising from notochordal elements, more frequent in fifth and sixth decades, affect much more often men than women. Chondroid chordoma is slower growing than conventional chordoma and shows foci of chondroid (cartilaginous) differentiation. It has a better prognosis than classic (non-chondroid) chordoma. Heffelfinger *et al*. described this variant of chordoma indistinguishable from hyaline type chondrosarcoma.[(6)]

Case Report: A 60-year-old female was admitted in orthopaedicward of this hospital with a complaint of numbness and pain in left hip and has increased since 4-8 months period. Magnetic resonance imaging (MRI) findings are showed aapproximately 5.8x4.2x4.8cm, expansile mass involving S4-S5 bodies and coccyx protruding in presacral space. Operative findings revealed tumor mass eroding sacrum below S2 level. It was pink, friable and soft to firm and vascular mass. We received multiple grayish-white to brown pieces which had soft to hard consistency. Hematoxylin and eosin stained sections studied from multiple bits show tumor cells arranged in sheets, cords and nest embedded in myxoid to chondroid matrix. These are small round cells having eosinophilic to vacuolated cytoplasm giving bubbly appearance known as physaliferous cells and round nuclei. Areas of hemorrhage and necrosis seen.

Discussion

Chordomas are usually relatively slow-growing, low-grade malignancies. They arise from the sacrum in approximately 50%–60% of cases, skull base region (spheno-occipital/nasal) in approximately 25%–35% of cases, cervical vertebrae in approximately 10% of cases, and thoracolumbar vertebrae in approximately 5% of cases.[(2)]Genetic studies performed on chordomas include chromosome analysis, telomere reduction and telomere

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activity, DNA microsatellite, loss of heterozygosity (LOH), and clonality studies.Clonality studies on eight cases of sacral chordomas indicated a polyclonal origin of the tumor.[7]

Clinical presentation is usually with pain as the cardinal symptom, whereas neurologic deficits tend to vary based on the location of the lesion. Chordomas are divided into conventional, chondroid, and dedifferentiated types. Conventional chordomas are the most common. They are characterized by the absence of cartilaginous or additional mesenchymal components. Chondroid chordomas contain both chordomatous and chondromatous features. The cartilaginous foci of chondroid chordomas resemble those of chondroma or low grade chondrosarcoma. Histologically, they display lobules and vacuolated (physaliphorous), moderately atypical, neoplastic cells across a myxoid stroma separated by fibrous bands.[8]

They have an ectodermal origin, chordomas are not sarcomas; however, they are traditionally classified and approached as sarcomas on the basis of being a primary bone tumor.[9]

In case of diagnositicdelimma, immunohistochemistry stain demonstrates that tumor cells of chondroid chordoma are reactive to epithelial markers like epithelial marker antigen (EMA) and cytokeratin (CK). Chondroma and chondrosarcomas are negative for CK and EMA and are positive for vimentin.[5]

MRI and computed tomography scan are important for pre-operative planning and staging of disease. Tumor can be visualized as midline, lobular, osteolytic foci or an expansile concomitant soft-tissue mass. Technetium-99m bone scan demonstrates hot areas within the tumor showing destructive changes with sclerotic rim and calcification in pre or paraspinal soft-tissue mass.[10]

Though chordoma has been considered of low metastatic potential; but distant metastasis to lung, bone, soft tissue, lymph node, liver, and skin has been reported in up to 43% of patients.[11,12,13] Metastatic disease may be considered of adverse prognostic significance because the median survival time was reported to be <12 months in a series of 28 chordoma patients after the development of distant metastasis.[9]

Surgery continues to be the primary modality in the management of chordomas. Chordomas are considered radioresistant tumors and require doses in excess of 60 Gy. Patients with spinal and sacral chordomas at the Massachusetts General Hospital, with a 5-year local control rate of 53%.[14]Chordomas are not reported to be sensitive to chemotherapy, similar to many other low-grade malignancies. Accordingly, chemotherapy response has been reported in patients with high-grade dedifferentiated chordomas, which represent <5% of all chordomas.[15]

Conclusion

Chondroid Chordomas are uncommon primary bone tumors with a high risk for local recurrence and modest propensity for distant metastasis. Surgery is the primary modality to achieve the best long-term control therapeutic value. Newer techniques and charged particle radiotherapy allow for better dose delivery, and hence better disease control. Chondroid chordoma has better prognosis than conventional chordoma.

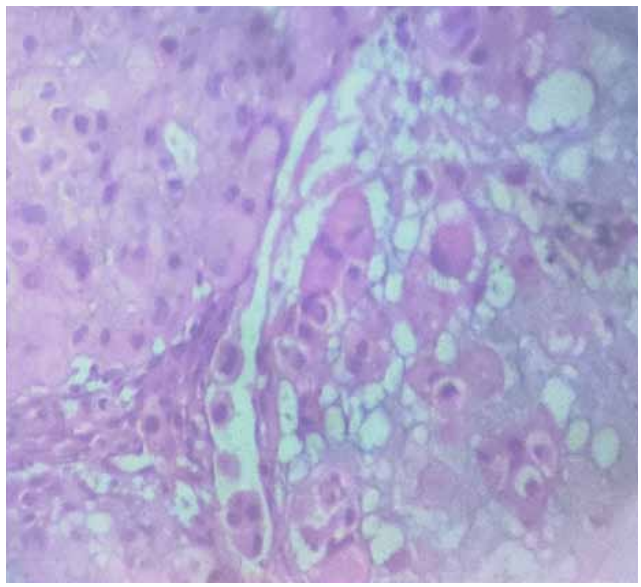

Figure 1: High power view showing small round cells have eosinophilic to vacuolated cytoplasm giving bubbly appearance known as physaliferous cells and round nuclei embedded in myxoid to chondroid matrix. (H and E).

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