Case Report

Sacrococcygeal chondroid chordoma: A case report with brief literature review

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ABSTRACT

Chondroid-chordoma is an uncommon variant of chordoma that originates from the remnants of notochord. These tumors have axial distribution particularly at the upper and lower ends of the vertebral column. However, they may occur in unusual sites in ectopic notochordal tissue. This paper reports a rare occurrence of chondroid variant of chordoma at sacrococcygeal region. The characteristic presentation, radiological appearance, findings at surgery, pathology and treatment of this lesion are discussed due to its predilection for occurrence at the sacrococcygeal region and its more favorable prognosis compared with that of conventional chordoma.

Key words: Chondroid-chordoma, notochord, sacro-coccygeal

INTRODUCTION

Chordoma is a rare malignant tumor arising in the axial skeleton from notochordal remnants. Notochord is a phylogenetic structure representing a primitive spine, which in higher animals replaced by the vertebrae and sacrum. Remnants of notochord are found in human within the vertebral bodies and intervertibral discs and more rarely in presacral soft tissue. Chordomas represent one to four percent of all primary bone tumors in various series.[1,2] It occurs mostly in adults with an overall peak in patients aged 55-65 years. Chondroid chordoma occurs in slightly younger individuals with a mean age of 40 years.

In 1973, Heffelfinger *et al*. described chordomas at the base of the skull with prominent chondroid differentiaton. It has a better prognosis than conventional chordoma. Chondroid chordoma may occur in spheno-occipital and sacrococeygeal areas.[3]

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We report a case of sacrococcygeal tumor, which was diagnosed as chondroid chordoma, with review of the literature.

CASE REPORT

A 60-year-old male was admitted in surgery ward of this hospital with a complaint of numbness and pain left hip. Pain has increased since 4-8 months period. Pain radiated until left foot which increased with physical activity. Patient also gave a history of constipation off and on. On rectal examination a mass was felt on posterior side. Magnetic resonance imaging (MRI) findings are showed a presacral mass, which was pushing the rectum and eroding the sacrococcygeal bony tissue.

Operative findings revealed tumor mass eroding sacrum below S2 level and going anteriorly up to lateral rectal margin. It was pink, friable and soft to firm and vascular mass intraoperatively.

We received multiple grayish-white to brown pieces which had soft to hard consistency.

Microscopic examination

Multiple hematoxylene and eosin stained microsections studied, which revealed tumor cells in lobulated pattern. The lobules are separated by fibrous septa. Within lobules two populations of cells were seen, small round

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cells arranged in cords and trabeculae with eosinophilic cytoplasm and round nuclei, second types of cells show vacuolization of cytoplasm giving bubbly appearance known as physaliphorous cells. Stroma was myxoid and showed areas of chondroid differentiation.

With these histomorphologic findings diagnosis of chondroid chordoma was made [Figures 1 and 2].

DISCUSSION

Chordoma is locally aggressive tumor that grows slowly, invades surrounding soft tissue and metastasizes infrequently. It is only malignant tumor arising from notochordal elements, more frequent in fifth and sixth decades, affect much more often men than women. Chordoma usually occurs in the sacrococcygeal (50%), spheno-occipital region (35%) and (15%) in the true vertebrae.[4] Nearly, 33% chordomas in the base of the skull are chondroid in type. They are rarely evident at other sites such as sacrococcygeal region.[5,6]

Clinical manifestations depend on location of tumor. In cranial region, they can cause cranial nerves palsies, hydrocephalous and torticollis.[7] The sacral lesions remain asymptomatic for a long time and presents with variety of non-specific symptoms which include progressive perineal pain, constipation, frequency of micturition and urinary incontinence. Pressure on emerging nerve roots may lead to paresthesia and anesthesia.

Histomorphological chordomas have been divided in to three subtypes; conventional, chondroid and dedifferentiated. Conventional chordoma is the most common type, slow growing, soft, tan, malignant tumor. 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.[3]

Dedifferentiated chordomas are rapidly growing tumors and are biphasic, with areas of high-grade sarcoma, alongside conventional or chondroid chordoma. They comprise 1% to 8% of all chordomas. Most studies suggest that they arise from sarcomatous transformation of chordoma, with some reporting their occurrence following post-operative radiotherapy for conventional chordoma. The prognosis is very poor with most patients dying of tumor related complications within 1 year.[8,9]

The cartilaginous foci of chondroid chordomas resemble those of chondroma or low grade chondrosarcoma. Immunohistochemistry stain demonstrates that tumor cells of 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.[4] Therefore diffuse expression of CK and EMA in a tumor is diagnostic of chondroid chordoma in case of diagnostic dilemma.

Imaging studies like MRI and computed tomography scan are vital 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]

Other tests procedures include fine needle aspiration cytology or true-cut needle biopsy, intra-operative cytology or a Frozen section, which can be performed for diagnostic purposes and to determine the excision of tumor borders.

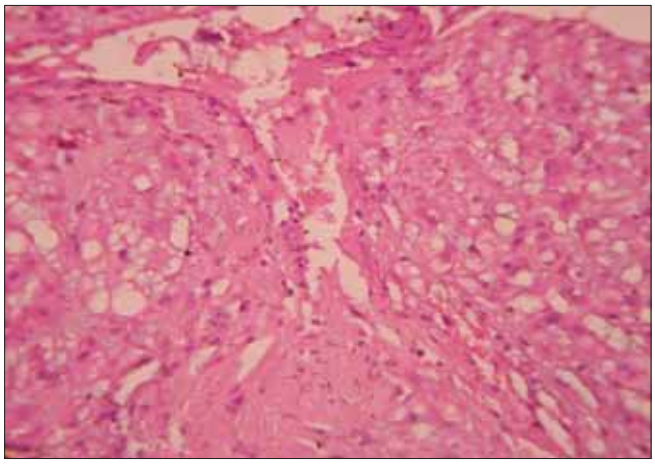

Figure 1: Low power view of chondroid chordoma showing tumor cells in lobulated pattern. The lobules are separated by fibrous septa (H and E)

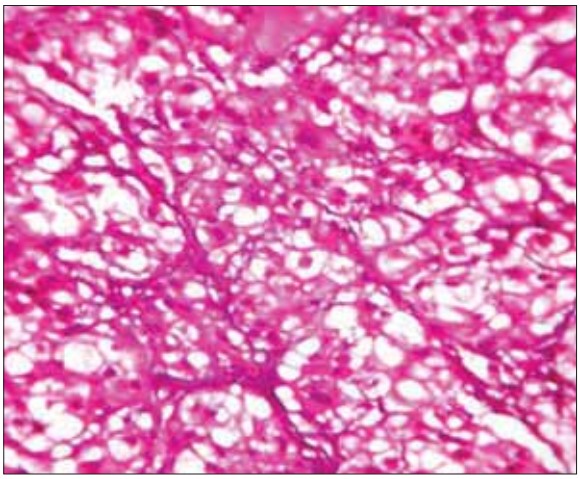

Figure 2: High power view showing, within lobules two populations of cells, small round cells arranged in cords and trabeculae with eosinophilic cytoplasm and round nuclei, second types of cells show vacuolization of cytoplasm giving bubbly appearance known as physaliphorous cells (H and E)

Unfortunately, many chordomas are massive when initially evaluated and because of their locally aggressive behavior have already infiltrated adjacent structures. Treatment options include low-dose or high-voltage radiation therapy, combined radiation and surgery and surgical excision alone. In some instances, debulking procedures are only option. Use of photon radiation therapy may be beneficial especially for spheno-occipital lesions.[11] Tumors detected and diagnosed early have a favorable prognosis if treated with a complete or en bloc excision. The disease-specific survival rate of 88.9% in a study suggests that cyber knife stereotactic radiosurgery offers promise in the treatment of chordomas. The proposed increase to 40 Gy to the margin in five sessions with relative sparing of the adjacent critical structures and atleast 1 cm margin gives a favorable response to the chordomas. The 5 year and 10 year survival rates of conventional chordomas are approximately 50% and 25-30% respectively. Conversely, chondroid chordoma has 5 years 10 years survival rate of approximately 80%.[12]

All three types of chordomas can metastasize, usually latter in the course of disease, to skin, bone, lungs and lymph nodes except dedifferentiated chordoma, which metastasize early.

Vertebral body chordomas have a higher incidence of metastasis than do those arising in clivus or sacrum.[13]

Sacrococcygeal chondroid chordoma is a rare slow growing malignant tumor, with a high rate of local recurrence, which presents a difficult diagnostic and therapeutic problem. Differentiation from other variants of chordomas and chondrosarcoma is important. High index of suspicion is required for early diagnosis and help to achieve adequate surgical margin for good outcome.

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