CHONDROID CHORDOMA OF THE $L_5 S_2$ SPINOUS PROCESS BODY: A RARE CASE REPORT

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ABSTRACT

Chordoma are slow growing, locally destructive tumors derived from remnants of notochord. They occur mostly along axial skeleton that is basiocciput and sacrococcygeal area. are biphasic malignant neoplasm possessing elements of both chordoma and cartilaginous tissue, an entity which has better prognosis than classical Chordoma. The tumor is likely to recur and hence diagnostically important for appropriate management. Histopathologically, tumor showed chords or nests of cells with partly vacuolated cytoplasm (physaliferous cells) embedded in a myxoid matrix and extensive cartilage formation with degenerative calcification was seen. Immunohistochemically, tumor was positive for Cytokeratin and EMA and negative for S-100 except for Cartilaginous areas which were S-100 positive. We report a case of Chondroid chordoma in a 50 year male presented with intermittent radiating pain in both leg and backache for 1 year. MRI lumbosacral showing the tumor in posterior elements if $L_5 S_2$ vertebra. Chondroid chordoma is a distinct entity to be discriminated from typical type of Chordoma because of its better prognosis. MRI cannot differentiate between Chondroid chordoma and typical chordoma. Awareness of this rare tumor will avoid misdiagnosis and improve the prognosis. A wide surgical excision coupled with adjuvant radiation is the best treatment in the present case.

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Key words: Chordoma, Bone tumor, Lumbar Spine, Prognosis.

INTRODUCTION

Chordomas are locally invasive primary malignant bone tumors, that occurs in midline axial skeleton, which arises from remnants of the notochord and have high risk for recurrence. (1) Chordomas include 1-4% of all primary bone tumors. (2) About 50% of chordomas develop in the sacrococcygeal region, 35% in spheno-occipital region and 15% in the vertebrae. (3)They are rarely found in the scapula and transverse process of L3. (4) Chordomas are usually occur in adults with peak age of 55-65 years whereas Chondroid chordoma is seen at a slightly younger age group.(5) Both genders are equally affected by cranial chordomas, whereas in sacrococcygeal tumors, the male and female ratio is approximately 2:1.(6) Its Annual incidence is 0.1% per 1 lakh population. Prevalence of chordoma is approximately 8 per million or 1 in 125,000 people. Population based study showed that overall median survival for chordoma is 7 years. Prognosis for each Chordoma patient is different, depends on the age, site and the size of the tumor, histological subtype, method of the treatment, extent of resection and other factors. There are 3histologically subtypes histologically divided into : Classical (conventional), Chondroid and Dedifferentiated .(7)
Chondroid chordoma is slow growing which is rare with incidence of 0.1% cases in million population annually (8).They were described by *Heffel finger*, as biphasic malignant tumor which have elements of both chordoma and cartilaginous tissue with better prognosis than classic (Non chondroid) chordomas. While Dedifferentiated Chordomas are more aggressive, faster growing and more likely to metastasize. (9)

We report a rare occurrence of chondroid variant of chordoma at sacrococcygeal region). We have discussed this lesion due to its prediction for occurrence at sacrococcygeal region and its more favourable prognosis compound with that of conventional chordoma. During skeletal development the notochord degenerates and remains as the nucleus pulposus (compound of the vertebral disc). It has been proposal that incomplete regeneration of the residual notochord can become neoplastic, but it is mostly seen that the tumor arises within the vertebral body and generally affects more than half of the cases. (6)

CASE HISTORY

A 50 year male presented with intermittent radiating pain in both leg and backache for 1 yr. He had history of hypertension but no history of weakness, numbness, or incontinence. On local examination vertebral spinous

process was prominent, but no tenderness was present. No neurological deficit was present. All routine investigations were within normal limit. On radiological examination, MRI of Lumbosacral-T2 weighted sagittal coronal and axial images show well defined, hyperintense lesion involving bodies of L5S1 & S2. This appears hypointense on T1 weighted images. On diffusion weighted- No diffusion restriction is seen. Extension of tumor was seen into the neural foramina of L5 & S1. There was scalloping of posterior body of L5-S2 (Figure 1a,b,c,d).

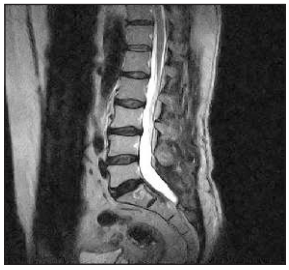

Figure 1(a)

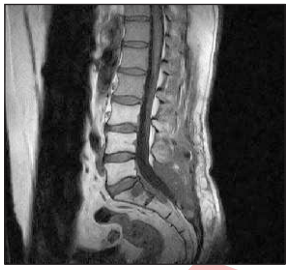

Figure 1(b)

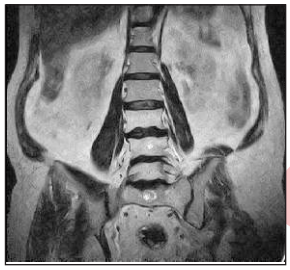

Figure 1(c)

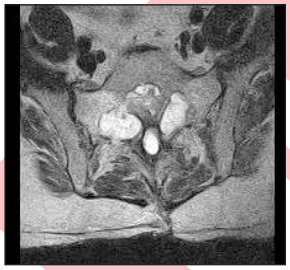

Figure 1(d)

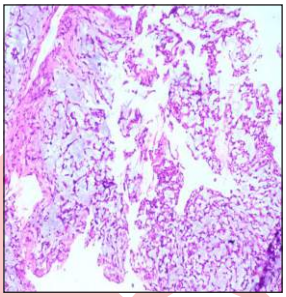

Figure 2(a)

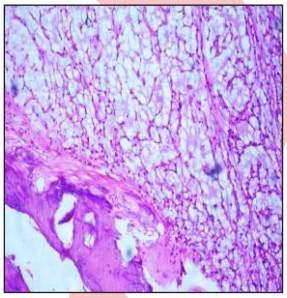

Figure 2(b)

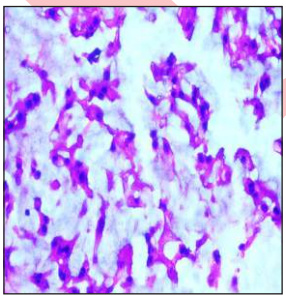

Figure 2(c)

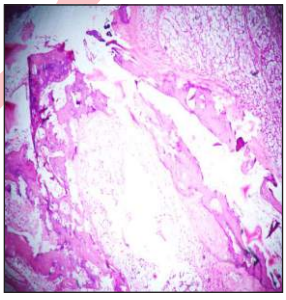

Figure 2(d)

Laminectomy with gross total extradural tumor excision was done. On histological examination, grossly multiple grey white soft tissue piece altogether measuring 2 x 2cm was received. Microscopically, tumor showed cords or nests of cells with partly vacuolated cytoplasm (physaliferous cells) embedded in a myxoid matrix and foci of cartilage formation with degenerative calcification were seen. (Figure 2 a,b,c,d). Immunohistochemically tumor was positive for cytokeratin and EMA and negative for S-100 except for cartilaginous areas which were S-100 positive. A diagnosis of Chondroid chordoma was made. On follow up- The patient was relieved of most of the symptoms with mild parasthesia in right leg. Repeat MRI showed small foraminal residual tumor. The patient was reported for Radiation therapy.

DISCUSSION

Chordomas usually occur sporadically, but can be familial in rare cases with autosomal dominance mode of inheritance. Chordomas are rare malignant tumor which originates from remnants of notochord, during skeleton development the notochord degenerate and remains as nucleus pulposis (compartment of the vertebral disc). It has been proposed that incomplete regeneration of the residual notochord can become neoplastic but it is mostly seen that the tumor arises in vertebral body and generally affects more than 50%.that represents 1-4% of all malignant primary bone tumors in which 50% and are usually noted at dorsal and chordal end (baso-occiput).

Chordomas can arise from base in the skull base and anywhere along the spine. The two most common locations are cranially at the clivus and in the sacrum at the bottom of the spine. Chondroid chordomas described as biphasic malignant neoplasm which contain both chordomatous and chondromatous features. (9)

The complete surgical resection followed by radiation therapy is the best treatment for long term control (10). Incomplete resection of the tumor makes controlling the disease more difficult and increases the odds of recurrence (10).

Chordomas are relatively radioresistant, requiring high doses of radiation to be controlled. The proximity of chordomas to vital neurological structures such as the brain stem and nerves limits the dose of radiation that can safely be delivered. Microscopically tumor resembles to normal notochord tissue which grows in cell cords and lobules, separated by mucoid intercellular tissue and fibrous septa, few tumor cells (physaliferous) are large, with vacuolated cytoplasm and prominent vesicular nucleus; but some shows cytoplasmic vacuoles contain glycogen. Other tumor cells are small, with inconspicuous nuclei and no visible nucleoli. Mitotic figures are scanty or absent. Areas of cartilage and bone may be present.

Histologically chordomas are subclassified into three variants of chordoma: classical ("conventional"), Chondroid and differentiated. Microscopically the differential diagnosis includes: Chondrosarcoma, Myxopapillary ependymoma, and Chordoid meningioma. (9) In Chondrosarcoma's the conventional type have tumor cells produces cartilaginous matrix either well, moderately or poorly differentiated with focal atypia, intracytoplasmic hyaline globules (11),while the Myxopapillary ependymoma shows well differentiated cuboidal to elongated tumor cells around vascularised myxoid cores with a myxopapillary in appearance which is similar to Chondroid chordoma (12), Chordoid meningioma resembles chordoma due to presence of cords and trabeculae of epitheloid cells. (13)Small vertebral chordomas are confined to the body with extension of tumor posteriorly to compress the spinal cord. The lesions may spread along the posterior longitudinal ligament or by direct extension through the intervertebral disc. The posterior element involvement of the spinal canal.(14)On Immunohistochemistry, the cells are positive for S-100 protein, keratin, epithelial membrane antigen [EMA]. In chondrosarcoma the D2-40 positive, EMA, pan CK & GFAP are negative, in the myxopapillary ependymoma the GFAP are positive but EMA is negative and in chordoid meningioma, EMA are positive and pan CK is negative. Huse JT et al showed that D2-40 which is a monoclonal antibody, behaves as a chondroid marker differentiating true chondroid neoplasm's from chordoma.(9)

CONCLUSION

Chondroid chordoma is a distinct entity to be discriminated from typical type of chordoma because of its better prognosis. MRI cannot differentiate between Chondroid chordoma and typical chordoma. Awareness of this rare tumor will avoid misdiagnosis and improve the prognosis. A wide surgical excision coupled with adjuvant radiation is the best treatment in the present care.

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