CASE REPORT

CHORDOMAS; PRESENTATION OF THREE CASES OF PARA SELLAR, CLIVAL, AND VERTEBRAL REGION

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ABSTRACT: Chordomas are rare, locally aggressive tumours that probably originate from embryonic remnants of the notochord. The sacrum is the most common site followed by the base of the skull/clival region. We present three cases with different sites of presentation of supra and parasellar, vertebral and clival chordomas that are managed at department of Neurosurgery Government General Hospital, Kurnool, Andhra Pradesh, India. Histopathologically chordomas are divided into 3 subtypes: conventional, chondroid and dedifferentiated. Of these three cases one is chondroid type histopathologically. The chondroid chordoma is having a significantly favorable outlook than the typical physaliphorous chordoma and the pertinent literature is reviewed.

KEYWORDS: Chordomas; Presentation of three case reports of para sellar, Clival and vertebral region and the literature reviewed.

INTRODUCTION: Chordomas are generally slow growing malignant neoplasms of presumed notochordal origin.¹ Cranial chordomas account for less than 1% of all intracranial neoplasms² and 4% of all primary bone tumours. Muller (1858) was the first to recognize its origin from notochordal cells, and Ribbert (1894) first introduced the term chordoma. Nearly all chordomas arising in the skull are related to the clivus. Those tumours related to the most rostral extension of the notochord in the dorsum sellae will present as sellar and parasellar chordomas.³ The term chondroid chordoma was introduced in 1973 by Heffelfinger et al.¹ It represented a sub-group of chordoma that straddles the bridge between conventional chordoma and chondrosarcoma. It has a better prognosis than the conventional type.^{4,5}

CASE REPORT 1: A 58 years old female presented with a history of headache for 2 months, diplopia for one month, ptosis of left eyelid and paraesthesia of left side of face of recent onset. Neurological examination revealed left sided III, IV and VI nerve palsy with hyperalgesia of maxillary division of left side of face. X-ray film of skull revealed calcification in the sellar region. Computed tomography demonstrated 2.7x2.6x2.5cm. Well-defined round partially enhancing lesion with peripheral calcification in the left side of sellar and suprasellar region and flattening of the left clinoid processes with thinned out bony wall in the superior aspect of the left sphenoid sinus (Fig. 1). Left pterional craniotomy was carried out. Lesion was approached intradurally. Microscopic examination of the tumor revealed lesion composed of large myxoid areas with lobules of cartilage. There are fragments composed of large polygonal cells with clear cytoplasm and small nucleus consistent with chondroid chordoma (Fig. 2). There was no post-operative complications. The patient received post-operative conventional radiotherapy with 5000 rads in divided doses over a period 5 weeks. We followed the case for a period of 6 months after surgery and radiotherapy. She did well with relief of headache.

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CASE REPORT 2: A male aged twenty two complained of pain in the thoracolumbar region of 6 months duration with swelling at that region. On examination showed tenderness at thoracolumbar region with gait disturbance. CT scan showed lytic lesion of the T12-L1 vertebral bodies with epidural cord compression (Fig. 3). T12-L2 laminectomy done, tumour decompressed and spine stabilized. Microscopic examination revealed the classic physaliphorous type and the radiotherapy delivered and the patient got relief of pain.

CASE REPORT 3: A 40 year old female presented with a sudden onset of head ache. MRI of brain revealed a large mass solid and partially cystic involving the brain stem with erosion of clivus (figure 4). Neurological examination showed depressed gag reflex on left side and unsteady gait. By median suboccipital approach tumor partially decompressed. Microscopic examination revealed myxoid matrix with round cells with occasionally larger cell containing multivacuolated cobweb-like cytoplasm, so called physaliphorous cells and the patient is subjected to radiotherapy.

DISCUSSION: Sinuous nature of the notochord remnants at the base of the skull explains the various sites of predilection of cranial chordomas. Chordomas are found along the axial skeleton and are relatively evenly distributed among three locations, sacro-coccygeal 30-50%, sphenoid-occipital 30-35% and vertebral body 15-30%.⁶ Sacro-coccygeal is the most common location of all chordomas and involving particularly the fourth and fifth sacral segments. With male; female ratio of 2:1. Sphenoid-occipital is the next most common the mass typically projects in the mid-line posteriorly indenting the pons. Vertebral chordomas are rare commonly involve the cervical spine (C2) followed by the lumbar spine then the thoracic spine. They commonly extend across the intervertebral disc space involving the more than one vertebral segment and extend into the epidural space compressing the spinal cord. Those tumours related to the most rostral extension of the notochord in the dorsum sellae will present as sellar and parasellar chordomas.³

Forking at the rostral end of the notochord has been demonstrated in embryo and would be presumed as embryological source of the lateral skull base tumours.⁷ Cranial chordomas are most frequent in men than in women 2:1 and may present at any age with mean age at diagnosis being 38 years. These lesions present clinically as destructive bony masses with soft tissue involvement. They erode and impinge upon adjacent structures giving rise to a wide variety of clinical symptoms. In the cranial region they can cause cranial nerve palsies, hydrocephalus and torticollis. The sacral lesions can remain asymptomatic for a long time and present with a variety of nonspecific symptoms. These symptoms may involve back pain, changes in bowel habits or a feeling of fullness in the rectal area. Physical examination must include a rectal examination to exclude a presacral mass.⁸ An early clinical suspicion of skull base tumour is important, when total excision with preservation of neurovascular structures is the goal.

Diplopia is an early feature of parasellar region tumours. Headache, proptosis and other cranial nerve signs occur at a later stage. Calcification has been reported in 34 to 90% of tumours. High resolution C.T. gives essential information about the site, extent and nature of the lesion. MRI provides the 3-dimensional analysis of the posterior fossa, sella turcica, cavernous sinus and middle cranial fossa. Clear cut bone destruction without sclerosis, punctate calcification and non-contrast enhancement on C.T. are characteristic features. Angiography is indicated to demonstrate the relationship of the internal carotid artery to the lesion and it is important when considering surgery in both parasellar and other skull base tumours.

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Metastatic spread of chordoma is observed in 7-14% of patients and includes nodal, pulmonary, bone, cerebral or abdominal visceral involvement predominantly from massive tumours. Surgical excision is the first line of treatment followed by the radiotherapy. These tumours are approached by transcranial subfrontal, pterional, and subtemporal routes to be considered for sellar or parasellar tumours. Surgical resection followed by radiation therapy because such treatment is associated with best survival than is surgery or radiation alone.^{8,9,10} Percutaneous radiofrequency ablation has been trialed as an adjunct. Recurrence; including seeding along the operative tract is common. Prognosis is typically poor, due to the locally aggressive nature of these tumours, with 10 year survival of 40% differentail diagnosis include chondrosarcoma, plamacytoma, giant cell tumour and spinal metastasis.

Chondroid chordoma was originally defined by Heffelfinger¹ and co-workers in 1973 as an otherwise typical chordoma that contained areas resembling hyaline cartilage and also found it to have a more benign clinical course. However the existence of a chondroid chordoma has been recently questioned. In 1994 Ishida and Dorffman developed an organography and provide criteria for differential diagnosis of chondroid chordomas.^{5,11,12} Tumours with (1) predominantly chondroid pattern and small chondroid foci. (2) equal volumes of chondroid and chordoma components (3) cytokeratin and epithelial membrane antigen, that when positive were classified as chondroid chordoma. On the contrary tumours that were predominantly cartilagenous with small chordoid elements and negative staining in both areas were classified as chondrosarcoma. Rosenberg et al^{13,14} using Heffelfinger's criteria found that all chondroid and non chondroid chordomas were immunopositive for cytokeratin and that the majority were also immunopositive for epithelial membrane antigen (E.M.A.) In contrast none of the chondrosarcoma was immunoreactive to cytokeratin, E.M.A. or C. E. A. Vimentin and S-100 Protein were immunopositive in more than 95% of the both classic and chondroid chordomas and chondrosarcomas.

Morphologically chordomas consist of sheets, nests and cords of large cohesive cells that were surrounded by myxoid matrix and physaliphorous cells with vacuolated cytoplasm. Chondroid chordoma consists of areas of typical chordoma that are adjacent to or merge with foci that resemble hyaline cartilage. Chondrosarcomas were of the mixed hyaline and myxoid type. Cytologically the cells of chondrosarcoma were significantly smaller and the nuclei were more uniform with less prominent or absent nucleoli in comparison to chordoma cells.

CONCLUSIONS: Our diagnosis is based on morphological appearance and immunohistochemical staining was reserved for certain cases in which the differential diagnosis between chondroid chordoma and chondrosarcoma was not possible using only haemotoxylin and eosin stain. So chondroid chordoma is a variant of chordoma reported to have better prognosis and should not be confused with chondrosarcoma. Multidisciplinary approach is the important pre-requisite in succeeding to improve quality of life and outcome.

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Fig. 1: CT scan of case 1 coronal and axial sections showing supra and parasellar mass with punctate calcification.

Fig. 1

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Fig. 2: Chondroid chordoma showing large polygonal cells with small nucleus. Chondroid chordoma with lobules of cartilage and chondroid tissue in a myxoid stroma histopathologically.

Fig. 2

Fig. 3: CT scan of case 3 dorsolumbar spine axial and sagittal showing osteolytic lesion in vertebral body T12-L2 with epidural extension into spinal canal causing cord compression.

Fig. 3

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Fig. 4: MRI scan of case 3 sagittal and axial showing large clival mass causing brain stem compression.

Fig. 4

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