Craniospinal Dissemination of Clival Chondroid Chordoma

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ABSTRACT

Chondroid chordoma commonly presents as clival osseous and extradural mass. A 15-year-old boy presented with progressive visual deficit, headaches and diplopia since three years. Computed tomography (CT scan) showed a skull base tumour, but was wrongly reported at the time as chronic sphenoidal sinusitis and nasal polyps. In the past three months, he developed dysphagia, urinary retention and constipation. Terminally, he had weakness of all limbs. Fundoscopy showed optic atrophy. Temporal and spatial variation in symptoms led to a clinical diagnosis of multiple sclerosis with optic neuritis. Partial brain autopsy revealed small gelatious tumour nodules in the subarachnoid space of middle cranial fossa encasing base of brain like arachnoids. Tumour deposits extended down into the spinal cord along the subarachnoid space as far as vision allowed. Histopathology and immunohistochemistry confirmed a diagnosis of chondroid chordoma. Awareness of this rare mode of dissemination will avoid misdiagnosis and delay in treatment.

Case History

A 15-year-old boy presented three years ago with diplopia and bilateral lateral rectus palsy following a minor nasal trauma. Computed tomography (CT) scan at the time was reported as being unremarkable. In the following six months, the patient started having intermittent throbbing frontal headaches with progressive visual deficit. The CT scan showed a skull base tumour, but was reported at that time as nasal polyps and chronic sphenoidal sinusitis. During the previous three months, he had dysphagia and suffered from an inability to initiate urination and defecate. Terminally, he came to Mumbai with urinary retention, weakness of all limbs and projectile vomiting. Neurological examination revealed hypotonia of all limbs along with areflexia and sensory loss below thoracic (T)6 level. Cerebrospinal fluid (CSF) examination was unremarkable. Fundoscopy revealed bilateral optic atrophy and macular oedema. Complete haemogram revealed only mild anaemia (Haemoglobin 8.3 gm%). Ultrasonography (USG) of the abdomen revealed bilateral hydroureter, mild hydrourephrosis and cystitis. Urine examination showed trace proteins and sugar and few pus cells. Renal and liver function tests were normal. USG abdomen and x-ray chest did not show any focal lesion. Clinical diagnosis was Devic’s disease (Multiple Sclerosis with optic neuritis) with urethral stricture. The patient expired before Magnetic Resonance Imaging (MRI) could confirm the diagnosis.

Partial brain autopsy revealed diffuse gelatinous tumour nodules encasing the base of the brain in a flat sheet, like arachnoids (Figure 1) and causing local destruction of the optic nerve and pituitary. Tumour deposits extended down the foramen of Magnum along the subarachnoid space as far as vision allowed.

Histopathology showed a lobular tumour (Figure 2) with chondroid and chordoid areas. Chordoid foci showed typical physalipherous and cuboidal cells in cords and trabeculae. The cells were Periodic Acid Schiff (PAS) positive and diastase digestible. These areas were merging with chondroid matrix with atypical cells in lacunae (Figure 3), thus satisfying Heffelfinger’s criteria.[1] The matrix was Alcian blue positive. Psammoma bodies and metaplastic bone formation were seen. Immunohistochemistry found the tumour to be strongly positive for EMA (epithelial membrane antigen), S100, vimentin and cytokeratin, while GFAP (glial fibrillary acid protein) positivity was focal and weak, thus confirming the diagnosis of a chondroid chordoma.

Discussion

Chordomas are tumours arising from rests of primitive notochord along the craniospinal axis, especially at the cranial and sacral end. [1] Chondroid chordoma, first described by Heffelfinger in 1973, is a variant of chordoma that straddles
Figure 1: Chondroid Chordoma-Gross photograph showing subarachnoid nodular deposits of gelatinous tumor (arrow) encasing the base of the brain in a sheet-like manner resembling arachnoiditis.

Figure 2: Chondroid Chordoma- Distinctive lobular appearance (arrow) of the tumor on light microscopy. 50 x, Hematoxylin-Eosin stain (HE).

Figure 3: Craniospinal Dissemination of Clival Chondroid Chordoma.

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the bridge between conventional chordoma and chondrosarcoma. Twenty-eight to thirty-four per cent of skull base chordomas are of the chondroid variety.

Common clinical presentations of clival chordoma include diplopia, visual defect, headache, VI-XII cranial nerve palsies, hypopituitarism and intranasal mass. Rarely, does it present as pure intradural chordoma. Drop metastases and subarachnoid dissemination from clival origin to cervical and thoraco-lumbosacral regions usually occurs late in the course of the disease and after multiple surgical and radiation therapies. This route of spread is either due to surgical implantation or hematogenous dissemination and is more common with anaplastic chordomas.

On hindsight, our case had all the typical features of a clival space-occupying lesion, though they were initiated by minor trauma. Dysphagia and nasal polyps were probably due to nasopharyngeal spread of clival tumour. Weakness, hypotonia, areflexia could be attributed to cervico-thoraco-lumbar spread, while bladder and bowel symptoms were due to the sacral spread of the tumour. The cranial symptoms preceded quadripareis, which preceded urinary symptoms. This sequence suggests primary craniospinal subarachnoid progression of tumour, since our case had no surgical intervention. Surprisingly, the CSF examination did not detect any malignant cells. The patient had no symptoms of hypopituitarism and no metastases were detected by USG or on chest x-ray.

On CT scan, chordoma presents as a destructive bone lesion with periosteal elevation and a solid-cystic soft tissue mass with foci of calcification. The chondroid variety calcifies more frequently than the conventional type. On MRI, it is hyperintense and enhances heterogeneously with contrast material. Myelography is preferred to MRI for diagnosis of subarachnoid nodules, since there may be loss of contrast between lesions and CSF on MRI. In our case, the tumour did not form a tumour mass, but encased the base of brain like a flat sheet resembling arachnoiditis. Perhaps, this is why it was not picked up on CT scan.

Differential diagnoses of skull base chondroid chordoma include myxoid chondrosarcoma, chordoid meningioma, metastases of signet ring adenocarcinoma and chordoid glioma. Light microscopic features and immunohistochemistry can differentiate between these entities. Chondroid chordoma presents commonly in young females, has a lower recurrence rate and better survival as compared to conventional chordoma. This prognosis is attributed to chondroid differentiation in notochord. Our case however showed a relatively rapid decline.

The present case highlights a rare presentation of chondroid chordoma as flat sheet-like encasing of base of brain resembling arachnoiditis. The tumour disseminated through the
subarachnoid space along the craniospinal axis in the absence of any interventional procedures that could have initiated metastases. Compression and infiltration of the spinal cord with multiplicity of symptoms in time and space made it mimic a demyelinating disorder. Clinical awareness of this mode of presentation will prevent delay in diagnosis and treatment.

References