CHONDROSARCOMA OF THE SKULL BASE - A CASE REPORT

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Abstract:
Chondrosarcoma of the skull base is a rare slow-growing tumor presenting with symptoms due to compression of adjacent structures. Total surgical excision is often difficult due to local anatomical limitations. The differential diagnosis among chondrosarcoma, chordoma, and chondroid chordoma is important due to their different prognoses. We describe a case of chondrosarcoma of skull base presenting with headache, visual dysfunction and seizures. Imaging revealed a skull base tumor. After tumor excision, histopathologic examination and immunostaining showed a grade II chondrosarcoma. In conclusion, accurate diagnosis and careful surgical treatment play important roles in the management of chondrosarcoma.

Keyword: Chondrosarcoma, Skull base, Intracranial, Temporal bone

INTRODUCTION:
Chondrosarcoma of the skull base is an uncommon neoplasm comprising 0.15% of all intracranial tumors and 6% of skull base neoplasms. They are slow-growing tumors. Diagnosis is only made after biopsy since clinical signs and symptoms and radiological findings are not pathognomonic. Symptomatology mainly derives from tumour encroachment and infiltration of adjacent intracranial structures. We present a case of chondrosarcoma of base of skull presenting with pressure symptoms.

CASE REPORT:
A 45 year old female patient presented to the outpatient department with complaints of headache for past 3 years, blurring of vision in the left eye and 3 episodes of seizures during the past one year. Imaging revealed a 5x5x5 cm well enhancing lobulated tumour with calcifications in the left temporal lobe compressing the left cerebral peduncle with adjacent temporal edema and midline shift towards right. [Figure 1] On sagittal view the tumour was seen to be attached to the base of the skull. [Figure 2]
Figure 1: CT Scan showing a lobulated well enhancing tumour with surrounding temporal edema and midline shift towards right

Figure 2: MRI on saggital view, the tumour was seen to be attached to the base of the skull with pressure effect on the left temporal lobe

The patient underwent left frontotemporal craniotomy and excision of the tumour was done and submitted for histopathological examination. Grossly, the tumour was received as multiple gray white soft tissue fragments varying in size from 1x1x1 cm to 3x3x2 cm. The external surface appeared nodular and the cut surface showed a firm gray white appearance with gelatinous areas, focal calcified and hemorrhagic areas. [Figure 3]

Hematoxylin and eosin stained tissue sections revealed a cellular neoplasm composed of sheets of cells with moderate eosinophilic cytoplasm and oval to elongated hyperchromatic nuclei with moderate nuclear pleomorphism with occasional mitotic figures. The cells are separated by abundant chondroid and myxoid matrix. The tumour is traversed by dense bands of fibrocollagenous tissue. [Figures 4 to 6] A diagnosis of chondrosarcoma grade II was made.
Immunohistochemical staining showed positivity for S100 in 60% of the cells, [Figure 7] and negativity for EMA, Cytokeratin 19 and Glial Fibrillary Acidic Protein, thus confirming the diagnosis of chondrosarcoma.

**Figure 7: S100 immunostain positivity**

**DISCUSSION:**

Chondrosarcomas are malignant mesenchymal tumours occurring only rarely in the bones of the cranium. Less than 5% of all chondrosarcomas are located in the head and neck area and their commonest location is the ethmoids and the sphenoid sinus. The prevailing hypothesis is that they arise from cartilaginous remnants in the petro-clival, sphenoid-occipital and fronto-nasal synchondroses. Chondrosarcoma can be subclassified, in order of frequency, into the conventional (hyaline or myxoid), dedifferentiated, clear cell, and mesenchymal subtypes. According to a review of 200 cases by Rosenberg et al, conventional chondrosarcoma is the most common subtype. Dedifferentiated chondrosarcoma is the most malignant with a high risk of metastasis. The most important differential diagnosis for chondrosarcoma of the skull base is chordoma. Although they are similar in management, distinction between chordoma and chondrosarcoma is important due to different prognoses and outcomes. They are difficult to differentiate with imaging alone and a misdiagnosis may be made even on histological examination. A chordoma typically contains cohesive nests and cords of large cells with bubbly eosinophilic cytoplasm called physaliphorous cells. The cells in chondrosarcoma are smaller and have less cytoplasm than those seen in a chordoma, and lack cohesive nests or cords. Chondroid chordoma has features of chondrosarcoma and chordoma. Chondroid chordoma is similar to chondrosarcoma in the cartilaginous areas and contain the cohesive nests and cords of physaliphorous cells that are typical features of chordoma.

Immunohistochemical study is helpful in confusing cases. Chondrosarcoma is usually positive for S-100 and negative for epithelial membrane antigen (EMA) and cytokeratin (CK). Chordoma, in contrast, is usually positive for EMA, CK and S-100. A diffuse chordoma background with areas of chondroid patterns immunonegative for CK staining is more suggestive of chondroid chordoma.

Computed tomography (CT) is useful in evaluating bone destruction and showing the characteristic calcified rim formation of chondrosarcoma. The soft tissue part of the skull base and chondrosarcoma are well demonstrated in magnetic resonance imaging. Surgery is the treatment of choice, while radiotherapy has an adjunctive role. Chemotherapy is not effective.
CONCLUSION
Chondrosarcoma of the skull base is a rare, slowly growing tumor. It should be differentiated from chordoma due to different clinical outcomes. Once the diagnosis of chondrosarcoma is made, careful preoperative image evaluation and radical excision with radiotherapy is the currently accepted management.

REFERENCES


