SELLAR CHONDROID CHORDOMA

(A Case Report)

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Introduction

Chordomas are uncommon neoplasms. They arise from the remnants of the notochord and hence are seen in close relation to the axial skeleton. Sites usually involved are the sacrococcygeal and the sphenoccipital regions [1,2]. Chordomas containing prominent chondroid elements have been termed as chondroid chordoma [3]. They are slow growing tumours which infiltrate adjacent structures and stubbornly recur after excision. Distant metastases are rare and occur late in the disease [3,4].

Case Report

A 10-year-old boy presented with squint of the right eye. Over the next few months the squint worsened and in 9 months he had complete ophthalmoplegia. There was pain with ptosis and pupillary dilatation in the left eye and abducens and trochlear nerve palsy on the right side. There were no visual field defects, limb weakness or symptoms pertaining to any lesion in the posterior cranial fossa. There was no other neurological deficit.

Skull radiogram showed stippled calcification in the sellar, suprasellar and retrosellar regions. Sphenoid sinus was hazy. CT scan revealed a partly calcified lesion in the same region with formation of small lucent cystic areas (Fig 1). This lesion had a parasellar extension. The patient underwent a right frontal craniotomy. A partial excision of the mass was possible. The postoperative period was uneventful. Movements were seen in the left eye. Patient was advised radiotherapy.

On histopathology, the tissue bits showed cells arranged in cords in a myxomatous stroma. The cells in places were compact, while other foci showed cells with characteristic bubbly (physaliphorous) cytoplasm (Fig 2). No significant pleomorphism, hyperchromasia or mitosis was observed. Foci of chondroid differentiation with calcification were also observed (Fig 3). The cytoplasm of the physaliphorous cells was PAS positive. The matrix was Alcian blue positive and negative for phosphotungstic acid haematoxylin (PTAH). A diagnosis of

chondroid chordoma was established.

Discussion

Intracranial chordomas originate mostly from the sphenoccipital synchondrosis [3,5]. Tumours have an average age of onset at 48 years, with age

Fig. 1 : CT scan. Coronal section through sella showing calcified tumour.

Fig. 2 : Tissue section showing characteristic physaliphorous cells. (H&E 200 X).

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Fig. 3 : Foci of chondroid differentiation seen in the tumour (H&E 200 X).

group ranging from 8-83 years. The male:female ratio is 2:1. The common complaint is diplopia [3].

These tumours form partially or nonencapsulated masses. Often they extend into inaccessible regions making total excision impossible as was the case in our patient. The mainstay of diagnosis is the demonstration of physaliphorous cells with areas of chondroid differentiation on histopathology. The physaliphorous cells are large cells with a round central nucleus and multivacuolated cytoplasm and are seen only in chordomas. Typical physaliphorous cells and areas of chondroid differentiation with calcification were observed in our case. Calcification is well known in chondroid chordomas [2,3,5]. Occasionally these tumours may be confused with mucin secreting adenocarcinomas or cartilaginous tumours. Adenocarci-

nomas, however, do not show physaliphorous cells while the matrix is PTAH positive in chondromas. These tumours were excluded in our patient morphologically and with the use of special stains as PTAH.

The importance of recognizing chondroid chordomas as a distinct group lies in their better survival rate. The malignant potential of chordomas appears to be decreased when cartilaginous foci are present [3,4].

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