PATHOLOGICAL DIAGNOSIS

Pleomorphic Xanthoastrocytoma (PXA)

Subhash Bhardwaj*, M. R. Attri, Meera Bhardwaj**

Abstract

PXA is a rare form of supratentorial astrocytoma which occurs in the young individuals and involves the leptomeninges. Microscopically the tumour shows pleomorphism without any evidence of necrosis and vascular endothelial proliferation. Tumour cells contain intracytoplasmic fat. The present case is a 20 years old male who presented with history of Grand Mal seizures and the CT scan showed a tumour in the supratentorial region. Histopathologically tumour turned out to be a PXA.

Keywords

PXA, Supratentorial, GFAP, Glioblastoma multiforme.

Introduction

The Pleomorphic Xanthoastrocytoma is an uncommon brain tumour arising in children and young adults and is characterised by a superficial cortical location, cellular pleomorphism and variable xanthomatous changes in the neoplastic astrocytes (1-2). The expression of glial fibrillary acidic proteins (GFAP) in the tumour cells has confirmed the astrocytic nature of these tumours. The tumour usually is located in the temporal lobes and clinically the patients have a relatively benign couse characterised by history of complex seizures and signs and symptoms of a mass lesion in the brain. In view of the rarity of this entity, a case report is presented.

Case Report

A 20 years old male presented with complains of sudden onset grandmal seizures. Neuroradiological examination revealed a 2.0 cms. low density lesion located in the right temproparietal region of the brain. The brain biopsy done, consisted of multiple, soft yellowish, tumour tissue pieces measuring together about 2×1.5×1 cms. The tissues were fixed in 10% buffered formalin and cut at 5μ thick sections after being processed for paraffin embedding. Subsequently the sections were stained by hematoxylin and eosin stains and reticulin stains. Sections showed a tumour composed of spindle and pleomorphic astrocytes, many of which contained intracytoplasmic lipid vacoules (Fig. 1). Occasional large hyperchromatic cells were also noticed. No evidence of tumour necrosis or vascular endothelial cell proliferation was seen in any of the sections studied. The reticulin stained sections revealed a reticulin rich tumour where the individual tumour cells were surrounded by reticulin fibres.

From the Departments of *Pathology and **Surgery, Government Medical College, Jammu (J&K) India.
Correspondence to : Dr. Subhash Bhardwaj, Lecturer, Pathology Department, Govt. Medical College, Jammu Tawi - 180001 (J&K) India

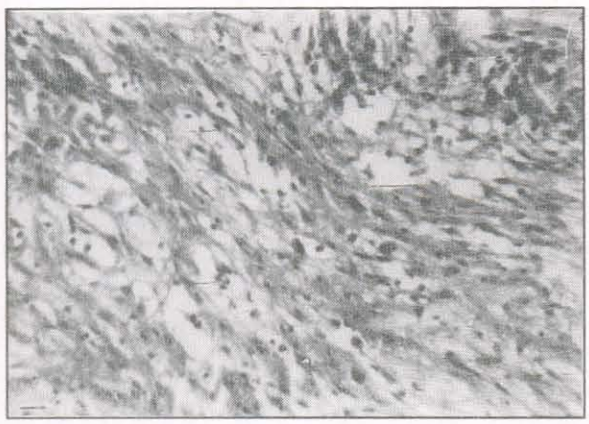

Fig. Photomicrograph from the tumour showing malignant astrocytes having vacuolated cytoplasm. H&E X40

Discussion

PXA displays distinct histopathological features, first described by Kepes *et. al.* (1). Before this, these lesions were referred to either as atypical fibroxanthomas or xanthosarcomas. PXA is a tumour of young adults usually occuring in the 2nd decade of life. Majority of cases are supratentorial in location, usually in the temporal lobes. Occasional case reports with infratentorial location of tumours have also been reported in the literature (2). A history of epileptic seizures is usually elicited in almost all the cases. In any given tumour, the lipidization of the neoplastic astrocytes and the amount of reticulin deposition reflects the elaboration of encircling basal lamina material by the tumour cells. The astrocytic lineage of PXA has been confirmed by immunohistochemical demonstration of GFAP in the tumour cells (3). GFAP expression also helps in segregating PXA from the lesions of mesenchymal origin e. g. malignant fibroushistiocytoma of brain. As far as cell of origin is concerned, the PXA is thought to arise from the subpial cortical astrocytes (1). The absence of necrosis and vascular endothelial cell proliferation in PXA is an important diagnostic point. It helps to differentiate PXA from a pleomorphic glioma and lipidized glioblastoma multiforme (4). Also, the absence of necrosis, paucity of mitotic figures and lack of vascular endothelial cell proliferation have been regarded as indices of slow growth of the PXA and perhaps this explains the indolent nature of the PXA inspite of it being histologically a pleomorphic tumour (4,5).

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