Case Report

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Pleomorphic xanthoastrocytoma

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ABSTRACT

Pleomorphic Xanthoastrocytomas (PXAs) are rare neoplasm of all astrocytic glial tumors, which occurs commonly in children and young adulthood. We present a case of 75-year male who presented with right side involuntary movements, unconsciousness and tongue bite. On radiological examination CT Scan Head (plain) showed well circumscribed isodense lesion measuring 7.2 x 5.6 x 4.8 cm at the temporoparietal region left side causing displacement of basal ganglia. On histopathological examination, diagnosed as pleomorphic xanthoastrocytoma, of left temporoparietal region. We present this report for its presentation at late age, its clinical, radiological and histomorphological features.

Keywords: Astrocytoma, Histopathology, Supratentorial tumors

INTRODUCTION

As these tumours are rare, advances in the understanding of its natural history and prognosis have been lacking. PAXs are located superficially at the cerebral hemispheres and occurs in younger patients. These are usually low grade astrocytic glial neoplasms, first reported in 1979 by Kepes et al. PXAs has been considered to have a relatively favourable outcome as compared to other astrocytic tumors. ^{2,3}

CASE REPORT

A 75-year male patient presented to our hospital with complaints of sudden onset involuntary movement, unconsciousness and tongue bite. There was no history of fever, any drugs, vomiting, headache, etc. He was a known case of hypertension for last 30 years and on regular treatment of tablet Stamlo 5mg once a day. Previous history of cholecystectomy 5 years back. Presently prostatomegaly detected on USG. Patient was known alcoholic and tobacco chewer since last 35 years. On radiological study CT Scan Head (plain and contrast)

showed well circumscribed isodense lesion (HU 30-40), measuring 7.2 x 5.6 x 4.8 cm in temporoparietal region left side causing displacement of the basal ganglia.

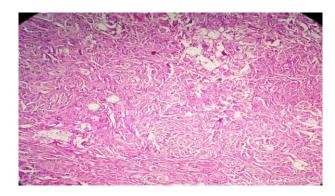


Figure 1: Tumour composed of pleomorphic astrocytes in sheets and diffuse pattern, (H and E stain, 100x)

No evidence of oedema, haemorrhage or calcification. Impression- S/O Neoplastic lesion-? Meningioma. Patient underwent left fronto-temperoparietal craniotomy with SOL excision. We received excised tissue from left temperoparietal lesion. Microscopic examination showed tumor composed of pleomorphic astrocytes in sheets and diffuse pattern, intermingled with spindle cells, xanthomatous cells, large giant cells and inflammatory cells. Tumor showed 2-3 mitosis/10 hpf. There was no area of necrosis. Scattered eosinophilic granular bodies were noted. On histopathological examination diagnosed as PAXs of left temporoparietal lobe.

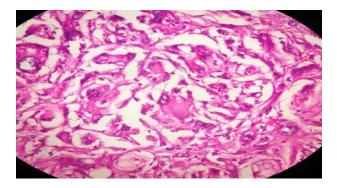


Figure 2: Numerous xanthoamatous foamy cells and multinucleated giant cells (H and E stain, 400 X)

DISCUSSION

PAXs is an uncommon tumor which constitute less than 1% of all astrocytic glial neoplasm. 4.5 About 99% cases showed tumor localization in supratentorial region by study of National Cancer Institute, Surveillance, Epidemiology and End Results, Database. 6 PXAs arises at cerebral hemispheres located superficially without dural involvement and in 40-50% cases it involves the temporal lobes. In our case it was situates in the left temperoparietal region.

The common age at presentation is in second decade of life, but PAXs can occur at any age ranging from infancy to ninth decade.^{6,7} In our case it presented at 75 years of age, which is rarely observed. PXA commonly occurs in young patients and manifests itself first as seizures followed by focal neurological deficits.

According to WHO (2007) classification PAXs are Grade II tumors of CNS.⁴ On histopathology our case showed tumor composed of pleomorphic astrocytes, intermingled with spindle cells, numerous xanthoamatous foamy cells and multinucleated giant cells. The mitotic activity was 2-3/10hpf. There were no areas of necrosis. The reticulin framework was noted in tumor. Also, eosinophilic granular bodies were noted.

The histological differential diagonosis includes glioblastoma, gliosarcoma, desmoplastic infantile ganglioma, desmoplastic astrocytoma of infancy. PAXs behave less malignant even with their highly pleomorphic histology. However, cases may have undergone transformation to anaplastic astrocytoma or glioblastoma.

A focus of small mitotically active tumor is considered to be sign of malignant transformation. Anaplastic PAxs are considered when mitosis is > 5/10hpf, necrosis and these are usually recurrence. The studies showed 9-20% PAXs may undergo malignant transformation.

PAXs have relatively favorable outcome with 5 years overall survival rate in 75-81% and 10 years in 67-70% cases. 6.7 PXA has been recently found to have high (up to 60%-78%) frequencies of BRAF V600E mutation in primary central nervous system (CNS) neoplasms. Usually the conventional PXAs demonstrate immunoreactivity for glial fibrillary acidic protein (100% of cases), S-100 protein (100%), class III beta-tubulin (73%), synaptophysin (38%), NF proteins (18 and 8%), and MAP2 (8%). 13

PAXs have relatively favorable outcome and treatment modalities are surgical excision. Additional chemotherapy and radiotherapy have been used.

CONCLUSION

We are presenting this case for its extreme rarity. It requires appropriate diagnosis as clinical course of patient is favorable. Also, these cases require regular follow up to detect any recurrences or malignant change.

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