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Thalamic PXA with Extension to Midbrain

Mohammad Ali Fazeli[1*], Ali Mardani[2], Niloofar Fazeli[2], Ehsan Shafiei[2] and Nafise Sabzevari[2]

*[*1]Functional Neurosurgery Research Center, Shohada Tajrish Hospital, Shahid Beheshti University of Medical Sciences, Tehran, Iran*

[2]Student Research Committee, Ahvaz Jundishapour University of Medical Sciences, Ahvaz, Iran.

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ABSTRACT

Pleomorphic xanthoastrocytoma (PXA) is uncommon slow growing tumor which is responsible for <1% of all astrocytic tumors. PXA incidence is frequently in the second decade of life and has a favorable outcome. We present the clinical, radiological and histopathological features of PXA involving the right thalamus and midbrain in a 9 year old girl with right upper motor neuron facial paresis of three weeks duration. Histopathology confirmed the diagnosis of PXA. Our case is unique in that our patient is the first one reported in the literature with midbrain and thalamus involvement. Radiotherapy may have a role to play in patients with incomplete resection. PXA usually presents with seizures, dizziness, and headache or rarely patients are asymptomatic. Patterns of presentation can be purely cystic, mixed cystic-solid and purely solid

Keywords: Pleomorphic xanthoastrocytoma, Midbrain, Thalamus

INTRODUCTION

Pleomorphic xanthoastrocytoma (PXA) accounts for <1% of all astrocytic neoplasms. PXA was first described by Kepeset [1]. In 1979 as a distinct clinic pathological entity in 12 young patients and was subsequently classified as Grade 2 astrocytoma by WHO in 1993 [2]. PXAs developed gradually and usually happen in the second decade of life with no sex preference [3,4]. Headache and long-lasting seizures are the presenting properties [5,6]. PXAs are superficial tumors with a preference for the temporal lobe followed by parietal, frontal and occipital lobes. Other uncommon sites are cerebellum, spinal cord and retina [3]. In spite of favorable prognosis, PXA malignant transformation has been depicted as an anaplastic variant [3,7,8].

Although PXA commonly affects young patients, this entity has been depicted in elderly patients with variable prognosis [5,6,9]. We present a rare case of PXA involving the right thalamus and midbrain a 9 year old Iranian girl with right upper-motor neuron (UMN) facial paresis. Our case is unique in that our patient is the first one reported in the literature with midbrain and diencephalic PXA.

CASE REPORT

A 9 year old girl presented to the neurosurgery department of Shohada Tajrish hospital with right sided facial paresis of three weeks duration left simple partial motor seizure lasting about one year. She also complained of headache in occipital area. There was no history of decreased sensations over the face or similar episodes in the past. She had no history of trauma, vomiting, fever or other associated symptoms.

On examination, higher mental functions were intact. There was an UMN type of facial paresis on the right side. Rest of the cranial nerves and neurological examination was intact.

Computed tomography scan of the brain with and without contrast revealed an intra-axial heterogeneously hyper dense mass in the right thalamus with extension to cerebral peduncle and quadrigeminal cistern. Brain MRI revealed an intra-axial solid lesion in the right thalamus extending into cerebral peduncle and quadrigeminal cistern. The lesion was hypo to isointense on T1-weighted images, heterogeneously hyper intense on T2-weighted and showed heterogeneous enhancement on gadolinium contrast.

The patient underwent a streotactic biopsy of thalamic lesion. The tumor was intra-axial in location, grayish white in color and partially firm consistency **(Figure 1)**.

Corresponding author: Mohammad Ali Fazeli, Functional Neurosurgery Research Center, Shohada Tajrish Hospital, Shahid Beheshti University of Medical Sciences, Tehran, Iran, E-mail: ma.ali.fazeli@gmail.com

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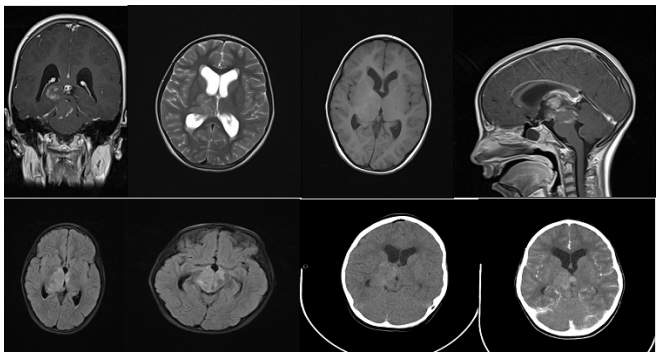

Figure 1. Streotactic biopsy of thalamic lesion.

Histolopathological examination revealed brain tissue involved by a low moderately neoplasm composed of neoplastic astroglial cells with fibrillary background. Occasional large bizarre cells were identified .no evidence of increased mitotic activity. No evidence of increased mitotic activity, necrosis or vascular endothelial proliferation was seen. IHC study revealed diffusely positive CD34 and Ki-67 was positive in less than 1% of tumoral cells. Pathological findings were in favor of low grade glioma compatible with pleomorphic xanthoastrocytoma **(Figure 2)**.

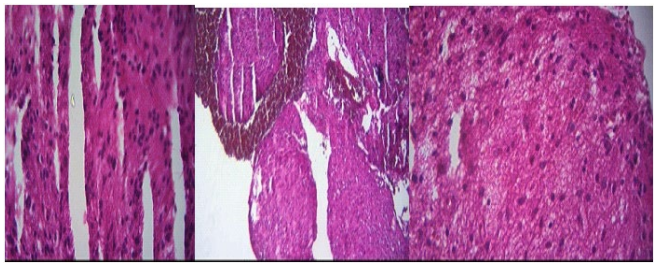

Figure 2. Pathological findings.

DISCUSSION AND CONCLUSION

Pleomorphic xanthoastrocytoma is an uncommon slow growing astrocytic tumor. These tumors frequently involve the supratentorial compartment with a preference for the temporal lobe [3,6]. They are thought to originate from the subpial astrocytes as a result of presence of "basal lamina", which is a typical property of these astrocytes [1,2,10,11]. Affected patients are frequently in their first three decades of life [3,5,9,12]. Reported a left fronto-temporal PXA in a 76 year old female, who underwent subtotal excision and palliative radiation therapy [6]. However, the patient died 6 months later as result of the rapid progression of the tumor. Perry et al. [9] described a composite PXA and ganglioglioma in an 82 year old male patient involving the left frontal lobe, who was subsequently lost to follow-up. Similarly, Bucciero et al. [5] reported an atypical PXA in a 65 year old man affecting the left capsulo-thalamic region that was treated with subtotal excision and fractionated external beam radiotherapy. The patient died 22 months after the excision as result of a massive recurrence of the tumor. Our patient was a 9 year old Iranian girl who was treated with brain stereotactic biopsy.

The prominent histopathological features of PXA as described by Kepes et al. [1] includes marked cellular pleomorphism with spindle cells, multinucleated giant cells with bizarre nuclei, prominent lipid droplets, eosinophilic granular bodies, perivascular lymphocytic infiltration and dense reticulin network. Mitotic figures are rare and necrosis is frequently absent. On immunohistochemical analysis, GFAP and S-100 is positive in all tumors [12]. Synaptophysin reactivity ranges from 38% to 100% and neuro filament protein expression is seen in 8-71% of tumors [3,12,13]. Although PXA has been known as a tumor with a favorable prognosis, 20% of these tumors can progress to malignancy and categorized anaplastic PXA [3,6-8,14]. Various factors such as increased mitotic activity, high Ki-67, MIB-1, proliferating cell nuclear antigen labeling index, endothelial proliferation, presence of necrosis, diffuse proliferation of monomorphic cells, decreased reticulin fibers and the extent of surgical resection, have been described to predict the unfavorable outcome in patients with PXA [3,6,7,9,12,15].

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