Pleomorphic Xanthoastrocytoma - A Rare Neoplasm of Central Nervous System

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Abstract: Pleomorphic xanthoastrocytoma is a rare variant of cerebral glioma. We present a case of 21-year-old male with history of seizure and diminution of vision of right eye. Computed tomography (CT) and Magnetic resonance imaging (MRI) were performed revealing a mass at right cerebellopontine region infiltrating into right cerebellum and right half of brainstem with supratentorial petro-clival extension. Diagnosis of pleomorphic xanthoastrocytoma was considered. Patient underwent tumour resection. Histopathology confirmed the diagnosis.

Keywords: Pleomorphic xanthoastrocytoma, seizure, Computed tomography, Magnetic resonance imaging

1. Introduction

Pleomorphic xanthoastrocytoma is one of the rarest benign neoplasm of central nervous system accounting for less than 1% of all astrocytomas. It is a superficially located neoplasm in cerebral hemisphere with seizure as the initial presenting clinical complaint. It is usually found in children and young individuals and can affect in males and females equally. It has favourable post-surgical outcome.

2. Case Report

A 21-year-old male presented to casualty in our hospital with complaints of sudden onset one episode of seizure and decreased vision in right eye in the last 1 week. Neurological examination did not reveal any focal motor and sensory deficit. Fundoscopic examination revealed papilledema in both eyes. CT and MRI brain revealed a large ill-defined mass lesion in right cerebellopontine angle cistern with petro-clival pressure erosion and infiltration of right cerebellum and right half of the brainstem. The lesion was seen extending anteriorly into suprasellar and right parasellar region and medially into right temporal lobe. Lesion measured 7.8 x 7.5x 5.3 cms in size, appearing hypointense in T1WI and heterogeneously hyperintense on T2WI and FLAIR with few foci on blooming on GRE. A large peripheral fluid signal intensity component was noted predominantly in right cerebellar hemisphere. The mass showed heterogenous pattern of enhancement on post contrast study. No significant perifocal oedema was noted. The lesion was causing compression and rotation of brainstem with mass effect on aqueduct, 4th ventricle with subsequent dilatation of bilateral lateral and third ventricles. Possibility of pleomorphic xanthoastrocytoma was raised which was further confirmed on histo-pathology.

Figure 1: Axial CT scan-bone window view reveals pressure erosion changes at petro-clival region on right side.

Figure 2: Axial T1WI MRI scan reveals ill-defined mass lesion at right cerebellopontine angle cistern infiltrating right cerebellum and right half of the brainstem with supratentorial petro-clival extension appears iso-hypointense on T1WI.
Pleomorphic xanthoastrocytoma are usually supratentorial in location. Rare cases of infratentorial location of tumour has been described. Computed tomography and Magnetic resonance imaging play an important role in diagnosis and planning of surgical management in patients. The tumour appears well defined, sometimes poorly defined iso to hypodense mass lesion with no significant perifocal oedema. Since the tumour is superficial, cortical based and slowly growing it may cause scalloping or remodelling of overlying bone. On MRI, the mass has solid -cystic appearance. The cystic component appears hypointense on T1WI, hyperintense on T2WI while the solid component shows iso to hypointense signals on T1 sequence and iso-hyperintense on T2 weighted imaging. The lesion may exhibit variable or mixed signal intensity on FLAIR imaging with enhancement of solid components on post contrast study. The differential diagnosis for this tumour includes meningioma, hemangiopericytoma, another low grade or high-grade glioma etc. The histopathological features of this neoplasm include presence of pleomorphic cells intermixed with spindle cells. Presence of necrosis suggest anaplastic nature of pleomorphic xanthoastrocytoma.

Bucciero A et al. studied 4 cases of pleomorphic xanthoastrocytoma and found predominance of superficially located meningo-cerebral mass in temporoparietal region on imaging studies which were confirmed on histology. Abid Met al. studied clinico-pathological spectrum of pleomorphic xanthoastrocytoma cases and found headache as the main presenting complaint with temporal lobe as the common site of occurrence. Histological characteristics of tumour includes pleomorphic cells, xanthomatous cells and spindle cells.

The tumour has favourable prognosis. Surgical resection is the mainstay treatment with relatively less chance of recurrence.

4. Conclusion

Pleomorphic xanthoastrocytoma is rare low grade astrocytic tumor with excellent survival rate post -surgery. Imaging modalities like CT and MRI aids in appropriate diagnosis, formulation of treatment protocol and post-operative follow up of patients. Radiologist should considered this neoplasm as differential diagnosis while imaging solid-cystic lesion in region.

References


3. Discussion

Gliomas constitute a large group of central nervous system neoplasms which can further be classified on the basis of histological pattern or characteristics. Pleomorphic xanthoastrocytoma is categorised as grade II, benign neoplasm according to WHO classification for astrocytic tumours. It was first described in 1979. It comprises less than 1% of all astrocytic neoplasms and is predominantly found in children and young individuals with no sex predilection. However rare incidence in older age group has also been reported. Patient may initially present with history of seizures. Signs and symptoms of raised intracranial tension may be present due to mass effect of the neoplasm. Pleomorphic xanthoastrocytoma are usually supratentorial in location. Rare cases of infratentorial location of tumour has been described. Computed tomography and Magnetic resonance imaging play an important role in diagnosis and planning of surgical management in patients. The tumour appears well defined, sometimes poorly defined iso to hypodense mass lesion with no significant perifocal oedema. Since the tumour is superficial, cortical based and slowly growing it may cause scalloping or remodelling of overlying bone. On MRI, the mass has solid -cystic appearance. The cystic component appears hypointense on T1WI, hyperintense on T2WI while the solid component shows iso to hypointense signals on T1 sequence and iso-hyperintense on T2 weighted imaging. The lesion may exhibit variable or mixed signal intensity on FLAIR imaging with enhancement of solid components on post contrast study. The differential diagnosis for this tumour includes meningioma, hemangiopericytoma, another low grade or high-grade glioma etc. The histopathological features of this neoplasm include presence of pleomorphic cells intermixed with spindle cells. Presence of necrosis suggest anaplastic nature of pleomorphic xanthoastrocytoma.

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