# 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 petroclival 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.<sup>[1]</sup>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 indivuals and can affect in males and females equally.<sup>[2]</sup> It has favourable post-surgical outcome.<sup>[3]</sup>

# 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[Figure 1]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 T1W1 [Figure 2], heterogeneously hyperintense on T2WI [Figure 3]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 [[Figure 4] No significant perifocal oedema was noted. The lesion was causing compression and rotation of brainstem with mass effect on aqueduct, 4<sup>th</sup> 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

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**Figure 3:** Axial T2WI MRI scan reveals ill-defined heterogeneously hyperintense mass lesion at right cerebellopontine angle cistern infiltrating right cerebellum and right half of the brainstem extending anteriorly into suprasellar and right parasellar region and medially into right temporal lobe.

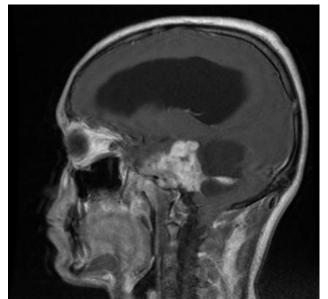


Figure 4: Sagittal T1 + C (contrast study) MRI scan reveals heterogenous post contrast enhancement of the solid components of mass

#### 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.<sup>[4]</sup>Pleomorphic xanthoastrocytoma is categorised as grade II, benign neoplasm according to WHO classification for astrocytic tumours.<sup>[5]</sup>It was first described in 1979.<sup>[6]</sup>It comprises less than 1% of all astrocytic neoplasms and is predominantly found in children and young indivuals with no sex predilection. However rare incidence in older age group has also been reported.<sup>[5][7]</sup>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.<sup>[8]</sup>

Pleomorphic xanthoastrocytoma are usually supratentorial in location. Rare cases of infratentorial location of tumour has been described.<sup>[9]</sup>Computed tomography and Magnetic resonance imaging play an important role in diagnosis and planning of surgical management in patients. The tumour appears has 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.[10][11]The differential diagnosis for this tumour includes meningioma, hemangiopericytoma, another low grade or high-grade glioma etc. <sup>[10]</sup>The histopathological features of this neoplasm include presence of pleomorphic cells intermixed with spindle cells. Presence of necrosis suggest anaplastic nature of pleomorphic xanthoastrocytoma.<sup>[11</sup>

Bucciero A *et al.* <sup>[8]</sup> studied 4 cases of pleomorphic xanthoastrocytoma and found predominance of superficially located menigo-cerebral mass in temporo-parietal region on imaging studies which were confirmed on histology. Abid Met al. <sup>[9]</sup> 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.<sup>[3]</sup>

# 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

- [1] Choudry UK, Khan SA, Qureshi A, Bari E. Primary anaplastic pleomorphic xanthoastrocytoma in adults. Case report and review of literature. International journal of surgery case reports. 2016 Jan 1;27:183-8.
- [2] Ksantoastrositom İP. Intraventricular pleomorphic xanthoastrocytoma: a case report. Turk Neurosurg. 2014;24(6):987-91.
- [3] Fouladi M, Jenkins J, Burger P, Langston J, Merchant T, Heideman R, Thompson S, Sanford A, Kun L, Gajjar A. Pleomorphic xanthoastrocytoma: favorable outcome

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after complete surgical resection. Neuro-oncology. 2001 Jul 1;3(3):184-92.

- [4] Johnson DR, Guerin JB, Giannini C, Morris JM, Eckel LJ, Kaufmann TJ. 2016 updates to the WHO brain tumor classification system: what the radiologist needs to know. Radiographics. 2017 Oct 13;37(7):2164-80.
- [5] Giannini C, Scheithauer BW, Burger PC, Brat DJ, Wollan PC, Lach B, O'Neill BP. Pleomorphic xanthoastrocytoma: what do we really know about it?. Cancer: Interdisciplinary International Journal of the American Cancer Society. 1999 May 1;85(9):2033-45.
- [6] Kepes JJ, Rubinstein LJ, Eng LF. Pleomorphic xanthoastrocytoma: A distinctive meningocerebral glioma of young subjects with relatively favorable prognosis A study of 12 cases. Cancer. 1979 Nov;44(5):1839-52.
- [7] Marton E, Feletti A, Orvieto E, Longatti P. Malignant progression in pleomorphic xanthoastrocytoma: personal experience and review of the literature. Journal of the neurological sciences. 2007 Jan 31;252(2):144-53.
- [8] Bucciero A, De Caro M, De Stefano V, Tedeschi E, Monticelli A, Siciliano A, Cappabianca P, Vizioli L, Cerillo A. Pleomorphic xanthoastrocytoma: clinical, imaging and pathological features of four cases. Clinical neurology and neurosurgery. 1997 Feb 1;99(1):40-5.
- [9] Huang Y, Zhu H, Gan W, Wang Z, Zhou Y. Infratentorial pleomorphic xanthoastrocytoma: report of two cases and review of literature. Int J Clin Exp Pathol. 2016 Jan 1;9(6):6391-6.
- [10] Tien RD, Cardenas CA, Rajagopalan S. Pleomorphic xanthoastrocytoma of the brain: MR findings in six patients. AJR. American journal of roentgenology. 1992 Dec;159(6):1287-90.
- [11] Sharma A, Sharma DN, Julka PK, Rath GK. Pleomorphic xanthoastrocytoma–a clinico-pathological review. Neurologia i neurochirurgia polska. 2011 Jan 1;45(4):379-86.
- [12] Abid M, Haroon S, Memon AH, Ahmad Z, Hasan SH. Pleomorphic Xanthoastrocytoma; Clinicopathological spectrum of An Intriguing neoplasm. Pakistan journal of medical sciences. 2018 Mar;34(2): 277

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