

## Primary intracranial extraosseus myxoid chondrosarcoma of dominant frontal lobe

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### Introduction

Primary intracranial extraosseus myxoid chondrosarcoma are an extremely rare neoplasm. Till date there are only nine cases of this entity has been reported. The authors present a case of primary intracranial extraosseus myxoid chondrosarcoma of the dominant frontal lobe.

### Case report

A 40 years old male was admitted to our department with complaints of headache on and off and recurrent seizures for last five years. Patient was on antiepileptic drugs as advised by his treating physician and he responded well earlier but for last one year seizures were not in control hence he was investigated. On imaging, NCCT brain shows an ill-defined heterogenous mass in left frontal region with focal areas of calcification. MRI brain shows a well circumscribed intra-axial heterogeneously hypointense mass on T1W and hyperintense on T2W and FLAIR MRI sequence. There was minimal perilesional edema. No restriction of diffusion seen on DWI and diffusion coefficient was increased on ADC sequence. Faint patchy enhancement seen on contrast images.

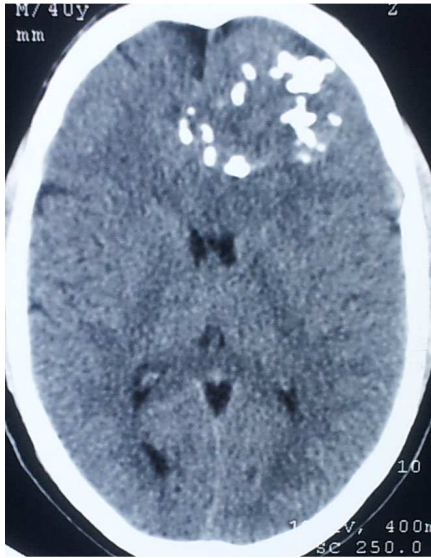
Left frontotemporoparietal craniotomy with complete excision of mass done. Tumor was well circumscribed greyish white in

colour, firm to hard in consistency, relatively avascular, loosely adherent to surrounding brain parenchyma. A plane of cleavage was present at the lesion brain interface.

On histopathology, sections shows circumscribed lobulated growth pattern with areas having hyaline cartilage. The lobules are variable in size. The capsule is moderately thickened and is showing mild lymphocytic infiltrate. The tumor has low to moderate cellularity and nuclear pleomorphism. Predominantly the lacunae are mononucleated however binucleated and multinucleated lacunae are also seen. Focal areas of calcification and fibromyxoid changes are also seen. Overall morphology is in favour of chondrosarcoma/chondroma grade -1.

On immunohistochemistry tumor cells are strongly positive for vimentin. 50-75 percent cells shows Bcl-2 expression. CD99, P-53, E-cadherine, EMA, CK8/18 and CK PAN (AE1-AE3) are negative in the tumor cells.

Postoperative period was uneventful and patient was discharged on fifth postoperative day without any neurologic deficit. Antiepileptics were initially prescribed for one month and then tapered gradually over fifteen days. On follow up of six months patient had no complaints and seizures were under control on phenytoin 300mg per day.



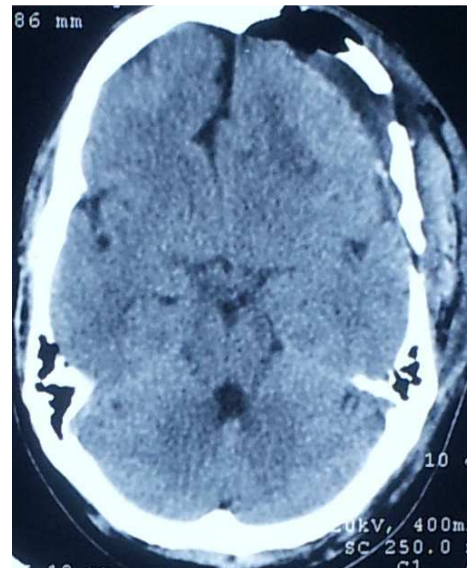
**Figure 1** - NCCT brain shows an ill defined heterogenous mass in left frontal region with focal areas of calcification



**Figure 2** - MRI brain shows a well circumscribed intra-axial heterogeneously hyperintense on T2W imaging



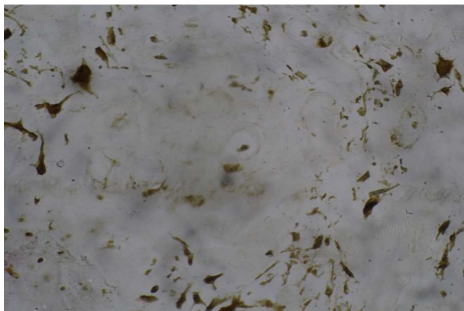
**Figure 3** - Tumor was well circumscribed greyish white in colour, firm to hard in consistency, relatively avascular



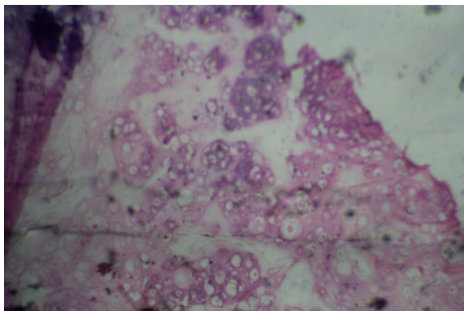
**Figure 4** - Postoperative NCCT brain suggestive of left frontotemporoparietal craniotomy with complete excision of mass



**Figure 5** - showing cells are Bcl-2 positive



**Figure 6** - showing cells are S-100 positive (high power)



**Figure 7** - There is blue myxoid matrix located in lacunae. Tumor cells are lobulated and double nucleated cells are present with foci of calcification

## Discussion

Since Enziner and Shiraki first described myxoid chondrosarcoma in 1972 as a deep soft tissue tumor of the extremities only nine cases

of primary intracranial extraosseus myxoid chondrosarcoma have been reported as per authors' knowledge. (2, 4, 5, 9, 10, 13, 15, 16, 17). Intracranial myxoid chondrosarcoma is extremely rare and is thought to arise from the choroid plexus, dura or in rare instances the pineal region. (1, 3, 6, 9, 14, 17). Histologically only three subtypes of intracranial chondrosarcomas have been described: classic chondrosarcoma, mesenchymal chondrosarcoma and myxoid chondrosarcoma. (5, 8, 11). Most of the Primary intracranial extraosseus chondrosarcoma show a dural involvement. However those within the brain parenchyma without any attachment to the cranium or the meninges are very rare. The case of author is of myxoid chondrosarcoma having no dural involvement.

The classic cranial and intracranial chondrosarcoma usually arise at the skull base and most frequently affect adults. (8, 15). The classic subtype has a better prognosis than the mesenchymal subtype. (15). Mesenchymal type usually occurs in the frontoparietal region and is highly vascular (15) it is the most aggressive subtype with a tendency for recurrence and metastasis. (8, 15). Calcification was more common in cranial myxoid chondrosarcoma and it lacks hyaline.

The optimal treatment for intracranial myxoid chondrosarcoma is radical excision, and total removal of the tumor is critical. (5, 8, 12, 14, 15). Adjuvant therapies, including radiotherapy, brachytherapy, and proton beam treatment, have been found to improve patient outcomes for this rare cancer. (5, 11, 15). However there have been no reports on the advantages of adjuvant chemotherapy for patients with intracranial extraskeletal myxoid chondrosarcomas till date. In addition, intracranial myxoid chondrosarcomas are not associated with a good prognosis after treatment.

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**References**

1. Bourgouin PM, Tampieri D, Robitaille Y, Robert F, Bergeron D, del Carpio R, et al.: J Comput Assist Tomogr 16: 268-273, 1992.10
2. Chaskis C, Michotte A, Goossens A, Stadnik T, Koerts G, D'Haens J: Primary intracerebral myxoid chondrosarcoma. Case illustration. J Neurosurg 97: 228, 2002.2
3. Cummings TJ, Bridge JA, Fukushima T: Extraskelatal myxoid chondrosarcoma of the jugular foramen. Clin Neuropathol 23: 232-237, 2004 . 11
4. Enzinger FM, Shiraki M: Extraskelatal myxoid chondrosarcoma. An analysis of 34 cases. Hum Pathol 3: 421-435, 1972 . 1
5. González-Lois C, Cuevas C, Abdullah O, Ricoy JR: Intracranial extraskelatal myxoid chondrosarcoma: case report and review of the literature. Acta Neurochir (Wien) 144: 735-740, 2002 .3
6. Grossman RI, Davis KR: Cranial computed tomographic appearance of chondrosarcoma of the base of the skull. Radiology 141: 403-408, 1981.12
7. Harsh GR IV, Wilson CB. Central nervous system mesenchymal chondrosarcoma: case report. J Neurosurg 1984; 61:375-81.16
8. Hassounah M, Al-Mefty O, Akhtar M, Jinkins JR, Fox JL: Primary cranial and intracranial chondrosarcoma. A survey. Acta Neurochir (Wien) 78: 123-132, 1985 .14
9. Im SH, Kim DG, Park IA, Chi JG: Primary intracranial myxoid chondrosarcoma: report of a case and review of the literature. J Korean Med Sci 18: 301-307, 2003.4
10. Jin Hoon Park, Mi-Jung Kim, Chang Jin Kim, Jeong Hoon Kim, J Korean Neurosurg Soc 52: 246-249, 2012.8
11. Korten AG, ter Berg HJ, Spincemaille GH, van der Laan RT, Van de Wel AM: Intracranial chondrosarcoma: review of the literature and report of 15 cases. J Neuro Neurosurg Psychiatry 65: 88-92, 1998 .15
12. O'Brien J, Thornton J, Cawley D, Farrell M, Keohane K, Kaar G, et al.: Extraskelatal myxoid chondrosarcoma of the cerebellopontine angle presenting during pregnancy. Br J Neurosurg 22: 429-432, 2008 .17
13. Peker, Bettül C. MD\*; Saral, Hatice D. MD\*; Baloğlu, Murat MD†; Ataizi, Serdar MD†Yurdasiper, Alper MD‡ Neurosurgery Quarterly: August 2013 - Volume 23 - Issue 3 - p 153-155.9
14. Sala F, Talacchi A, Beltramello A, Iuzzolino P, Bricolo A: Intracranial myxoid chondrosarcoma with early intradural growth. J Neurosurg Sci 42: 159-163, 1998.13
15. Salcman M, Scholtz H, Kristt D, Numaguchi Y: Extraskelatal myxoid chondrosarcoma of the falx. Neurosurgery 31: 344-348, 1992 .5
16. Scott RM, Dickersin R, Wolpert SM, Twitchell T: Myxochondrosarcoma of the fourth ventricle. Case report. J Neurosurg 44: 386-389, 1976.6
17. Sorimachi T, Sasaki O, Nakazato S, Koike T, Shibuya H: Myxoid chondrosarcoma in the pineal region. J Neurosurg 109: 904-907, 2008.7