PRIMARY LIPOSARCOMA OF THE MENINGES: A CASE REPORT

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ABSTRACT

INTRODUCTION: Liposarcoma of meninges is extremely rare pathology which is hardly reported in scientific literature. We present a interesting extraaxial lesion of brain which was proved to be primary liposarcoma of meninges. **CASE REPORT:** A 49 years old gentleman presented with gradual headache for 3 months. He was found to be have extraxial mass with heterogenous signal intensity on cross sectional imaging and given the provisional diagnosis of meningioma. On pathology and immunohistochemistry, it showed features of liposarcoma. Screening of extremities and abdomen showed no primary elsewhere. **CONCLUSION:** Though extremely rare, Liposarcoma of meninges should be considered as differential diagnosis to meningioma in extraaxial mass with heterogenous signal intensity as prognosis and treatment plan are different .

Keywords: primary, liposarcoma, meninges

Introduction

Liposarcoma of the meninges is a rare disease, with an incidence of approximately 0.5 - 3% out of all intracranial meningeal tumors. Meningeal liposarcoma usually does not have noticeable clinical symptoms even when they become large. Little is known about this clinical entity. There are hardly any case reports about such cases. Here we are reporting such a case who had no primary liposarcoma elsewhere and had no exposure to radiation. We found only two case reports of primary liposarcoma, one was reported by Sima A. et al, which was published in Acta Patho Microbiol Journal[1] and other more recent one was by Mumert et al,which was published in Journal of Neurooncology.[2] However this is the first case to be reported in myxoid variety of intracranial primary meningeal liposarcoma. Most known cases of liposarcoma of meninges till date have been described as a metastasis from the soft tissue sarcomas of other sites.

Case Report

A 49-year-old male was admitted to the hospital due to a gradual onset of intermittent type of headache that had started from past 3 months. Physical examination showed decrease in the power of the muscle of the limbs. No other abnormal neurological signs were elicited. All routine investigations were done which did not show any striking abnormality. Computed tomography angiography (CTA) exa-mination showed no intracranial artery malformation. The right anterior cerebral artery was displaced towards the left side by some mass lesion which was supplied by the anterior and the middle cerebral arteries.

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Maximum intensity projection clearly revealed arteries from the right anterior circulation of the brain encircling the mass, and the surrounding brain tissues were mildly compressed. Magnetic resonance imaging (MRI) showed that the size of the lesion was 5.8 cm x 5.9 cm x 5.6 cm with mixed T1WI and T2WI signals. The lesion was heterogeneous in signal intensity with midline shift towards the left and there was bilateral lateral ventricle compression. In contrast enhanced MRI, the lesion showed a heterogeneous type of enhancement (Fig: 1).

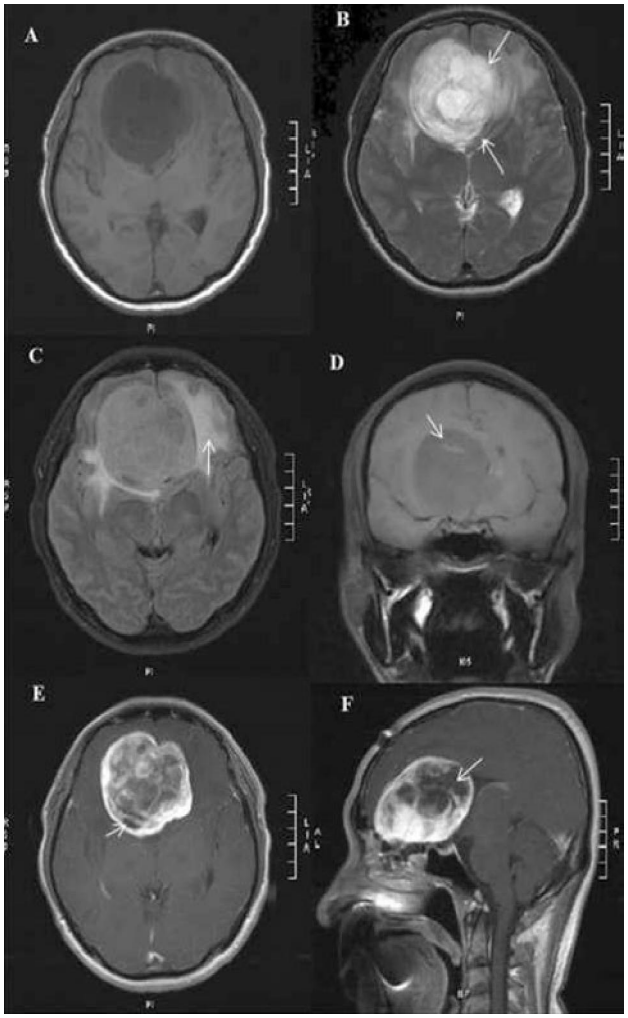

Figure 1: Myxoid liposarcoma with a typical appearance and heterogeneous enhancement. (A, D) Axial and Coronal T1-weighted MR images show lacy and amorphous foci within a mass (arrow in image D) (B) T2-weighted MR image showing edema and midline shift (arrow) (C) FLAIR MR image showing edema (arrow) (E, F) Gadolinium-enhanced axial and sagittal T1weighted MR images show heterogeneous enhancement throughout the mass with non-enhancing areas of necrosis and/or mucinous material (arrow).

An initial diagnosis of meningioma was given based on the medical history, physical examinations, and radiological examinations. However, the nature of the lesion remained undetermined, and lesion resection was scheduled. Intraoperatively, a white colored, well encapsulated mass of size 6.8 cm x 6.9 cm x 7.6 cm was resected. The tumor had an abundant blood supply, and was slightly adherent to the anterior clinoid process. Postoperative pathological results showed deeply stained spindle shaped and oval shaped mesenchymal cells within myxoid background. In addition, infarcted tissues were visible within the tissues. Immunohistochemistry was also done which showed Ki-67 (+) <1%, s-100(+), Desmin(-), EMA(-), PR(-) Fig: 2).

Thus, on the basis of pathological and immunohistochemistry report the mass was diagnosed to be the myxoid (mucinous) type of liposarcoma of meninges. MRI of lower extremities and abdomen revealed no mass lesion/primary. After treatment the patient is in good health condition and no signs of recurrence have been reported.

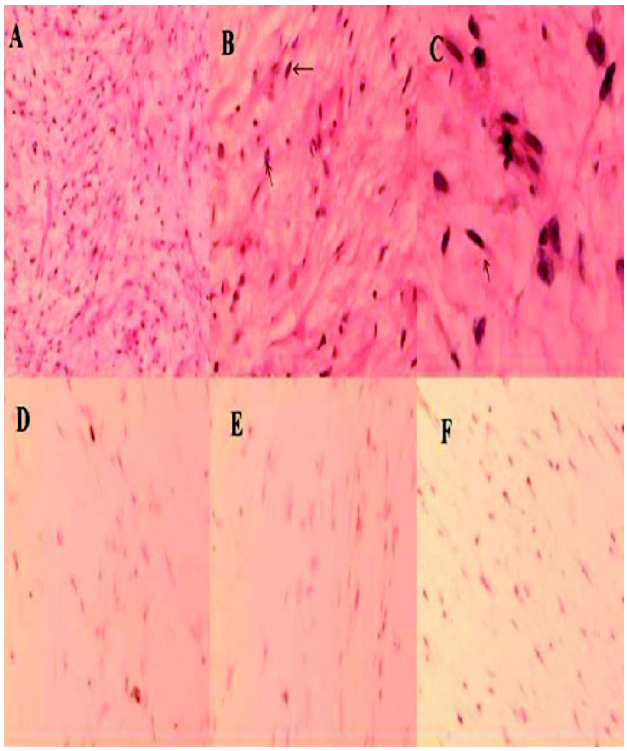

Figure 2: Pathology images (A, B, C) showing deeply stained spindle shaped and oval shaped cells (arrow in B &C) with myxoid background. Immunohistochemistry pictures (D, E, F) showing K-67 (+) (D&E) and S-100 (+) (F).

Discussion

Liposarcomas are exceedingly rare in the CNS and has been classified by WHO under intracranial mesenchymal tumor. CNS mesenchymal tumors itself are very rare. According to Brain Tumor Central Registry of the United States (CBTRUS), they comprise <1% of CNS tumors.[3] Most case reports of liposarcomas involving the meninges have reported metastases from soft tissue primaries. So detail work up is required before concluding the lesion is primary.[4,5]

Here we report a case of primary intracranial mass which was initially diagnosed as meningioma on the basis of preoperative imaging findings but on further pathological and immunohistochemistry examination the tumor was confirmed to be myxoid liposarcoma of meningeal origin.

Liposarcomas originate from primitive mesenchymal cells rather than from mature fat cells. Histologically they can be divided into: well-differentiated, dedifferentiated, myxoid, round-cell, and pleomorphic types.[6] Liposarcoma can be diagnosed in MR imaging due to presence of fat signal. However, myxoid liposarcoma are difficult to detect on MRI due to relative lack of fat signal,especially high grade have no fat component.[7] MR imaging findings depends on multiple factors including; amount of fat and myxoid material, the degree of cellularity and vascularity, and the presence of necrosis of the tumor.[6-8] Most myxoid liposarcomas have little fat which are in linear/lacy, amorphous foci. Some myxoid liposarcomas may be cystic at MRI without contrast, although they usually enhance like other solid masses at contrast enhanced MR imaging.[1] So without the administration of gadolinium contrast material, the tumor may even mimic a cystic tumor.[1] In our case, MRI examination revealed the tumors to be encapsulated, non-infiltrating and lobulated in shape. On T1-weighted sequence, we found few areas of lacy, amorphous foci of high signal within a predo-minantly low signal mass. These foci are due to fat within the tumor which helps to suspect these lesion even in unusual areas like brain. The mass of low signal intensity with few lacy high signal areas on T1-weighted sequences was clue towards our diagnosis of intracranial myxoid liposarcoma. On Gadolinium enhanced MRI imaging the lesion showed a heterogeneous type of enhancement. The enhan-cing areas within the tumor represent cellular soft tissue area the non-enhancing areas represent necrosis and accumulated mucinous material. So contrast MRI helps to differentiate benign cystic lesion from liposarcoma.[1,4,9]

The diagnosis in our case is further enforced by a pathological report which showed deeply stained spindle shaped mesenchymal cells in the myxoid background with areas of necrosis in the tissue which is characteristic for sarcoma. Immunohistochemistry of the tumor showed Ki-67 (+) <1%, S-100 (+), Desmin (-), EMA (-), PR (-) which also strongly suggests intracranial origin of myxoid type of liposarcoma.[10] The low level of ki-67 also indicates the lesion being intracranial in origin. Workup for primary should be always searched for in any liposarcoma of atypical site. We found no primary with expensive evaluation of abdomen and extremities. Due to the marked predilection of this tumor for the lower extremities,[8] MRI of the lower extremities was also done, which revealed no abnormality.

Conclusion

On the basis of preoperative and postoperative evaluation the intracranial lesion is finally diagnosed to be myxoid liposarcoma of intracranial and extraaxial origin. This patient presented with the first reported case of primary myxoid liposarcoma originating intracranially. The previous cases of primary intracranial liposarcoma reported were of well differentiated and pleomorphic type. Given the rarity of presentation, the clinical course and surgical outcome are difficult to predict. So when mixed signal intensity mass with probable fat component is detected on imaging, Knowledge about possibility of liposarcoma of intracranium may help the reporting radiologists and operating surgeons to plan and prognosticate the treatment.

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References

1. Sima A, Kindblom LG, Pellettieri L. Liposarcoma of the meninges. A case report. Acta pathologica et microbiologica Scandinavica Section A, Pathology. 1976; **84(4):** 306-10.

2. Mumert ML, Walsh MT, Jensen EM, Jensen RL. Pleomorphic liposarcoma originating from intracranial dura mater. Journal of neuro-oncology. 2010; **97(1):** 149-53.

3. Tarik T, Scott EK. Mesenchymal Tumors of Central Nervous System. In Perry A, Brat DJ, editors. Practical Surgical Neuropathology: A Diagnostic Approach. Philadelphia: Elsevier Health Sciences; 2010. 219-20

4. Blum A, Henrot P, Sirveaux F, Rios M, Verhaeghe JL, Molé D, Stines J, Vignaud JM: Diagnostic des lipomes des parties molles et des liposarcomes chez l'adulte. In Conduite à tenir devant une image osseuse ou des parties molles d'allure tumorale. Monographie du GETROA. Volume 31. Edited by Laredo JD. Sauramps medical, Paris; 2004: 401-16.

5. Kilpatrick SE, Doyon J, Choong PFM, Sim FH, Nascimento AG: The clinicopathologic spectrum of myxoid and round cell liposarcoma. Cancer 1996; **77:** 1450-8.

6. Sung MS, Kang HS, Suh FS, Lee FH. Myxoid Liposarcoma:Appearance at MRImaging with Histologic Correlation. RadioGraphics 2000; **20:** 1007-19.

7. Lowenthal D, Zeile M, Niederhagen M, Fehlberg S, Schnapauff D, Pink D, et al. Differentiation of myxoid liposarcoma by magnetic resonance imaging: a histopathologic correlation. Acta Radiol. 2014; **55(8):** 952-60.

8. Barile A, Zugaro L, Catalucci A, Caulo M, Di Cesare E, Splendiani A, et al. Soft tissue liposarcoma: histological subtypes, MRI and CT findings. La Radiologia medica. 2002; **104(3):** 140-9

9. Loubignac F, Bourtoul C, Chapel F. Myxoid liposarcoma: a rare soft-tissue tumor with a misleading benign appearance. World journal of surgical oncology. 2009; **7:** 42-7.

10. Philipps B, Lörken M, Manegold E, Kasperk R, Schumpelick V. (Primary liposarcoma of the stomach wall--a rare mesenchymal tumor). Der Chirurg; Zeitschrift fur alle Gebiete der operativen Medizen. 2000; **71(3):** 334-6.