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Case Report

Malignant intracerebral nerve sheath tumour of the brain presenting as a recurrent meningioma

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ABSTRACT

A malignant intracerebral nerve sheath tumour (MINST) is a variant of MPNST arising inside the brain. They are very rare tumours that are aggressive and have strong metastatic potential. We report a case of a recurrent lesion in the brain in a 39 years male who presented as recurrent meningioma. It was finally reported as MINST with histopathology and immunohistochemistry. Surgical excision is the treatment of choice. The patient is on follow-up after an uneventful surgical excision. To the best of our knowledge, very few cases of malignant intracerebral nerve sheath tumours have been reported so far in the literature. This case is presented not only for its rarity but also to highlight that this aggressive tumour needs to be kept in mind while diagnosing such lesions with these histomorphological findings.

1. Introduction

MPNST arising inside the brain are termed malignant intracerebral nerve sheath tumours (MINST). They are exceptionally rare malignancies with very few cases documented in the literature.[1] It generally presents between 1 to 75 years of age. The male-to-female ratio is 1.5:1 with a mean age of 29.7 ± 21.8 years.[2] There are 71% of cases of sporadic and 23% cases of NF type 1 related MINST.[3]These tumours have few risk factors like the presence of NF1 syndrome, along with a history of ionizing radiation exposure.[2] Schwannomas, the benign counterpart are also usually associated with neurofibromatosis.[4] It is very rare to find this tumour within the brain or cerebellar parenchyma.[5] Schwannomas do not undergo malignant transformation. Therefore it is preferable to label such lesions as malignant intracerebral nerve sheath tumours, and when rhabdomyoblastic differentiation is seen, it is called a Triton tumour.[5] Their cellular origin is not known.[4] Radioimaging findings are not specific. Therefore, histopathology with the help of immunohistochemistry remains the modality for the confirmative diagnosis. Molecular techniques can also act as an adjunct in its confirmation. These are highly aggressive lesions with recurrences. Surgery remains the mainstay of treatment as they are resistant to chemo and radiotherapy. Here we report a case of a young male presenting with a history of recurrent meningioma on CT and MRI. A final diagnosis of MINST was made on histopathological examination. We describe this rare but aggressive tumour with a review of the literature.

2. Case Report

A 39 -year-old male presented to the neurology department with complaints of headaches and visual problems. The patient had a previous craniotomy and resection of the tumour at the same site. There was no history of radiation

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therapy or clinical evidence of neurofibromatosis**.** Contrast-enhanced T1- weighted image revealed an extra-axial hyperdense mass of approximately 17x25 mm in the frontal region with bone erosion showing residual mitotic pathology suggestive of a recurrent meningioma/ glioma**.** A magnetic resonance imaging (MRI) examination revealed a heterogeneously intense tumour mass of 42x40x46 mm in the frontal region likely recurrence of meningioma with gliosis. (Figure 1). Thus based on the clinical and radiological examination a diagnosis of recurrent meningioma was made and excision of the tumour was performed and sent for histopathological examination.

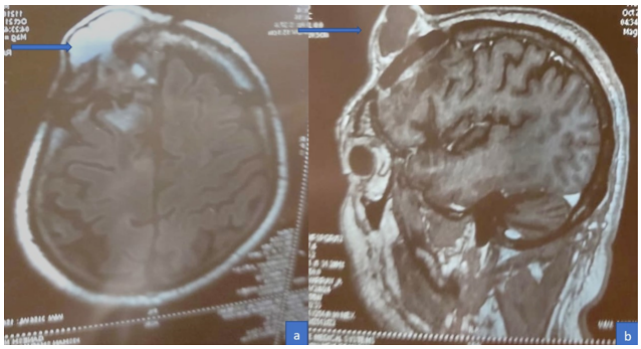

Fig. 1: MRI shows a hypoechoic lesion in the temporal region, eroding the skull plate.

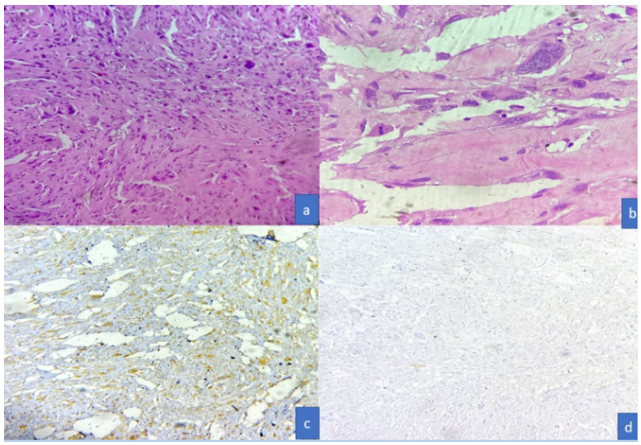

Fig. 2: Stained sections show intersecting fascicular loose arrangements of enlarged hyperchromatic nuclei with atypical mitotic figures and pleomorphic spindled cells with mitotic figures. H an E,40x,100x.(**a,b**) Immunohistochemistry for protein S-100 was strongly positive in neoplastic cells, and GFAP is negative 40x,100x.(**c,d**).

On gross, multiple grey-brown, friable tissue piece was found. On H and E stained section, the tumour showed a fascicular architecture with the spindle to pleomorphic cells with hyperchromatic nuclei and moderate to an abundant amount of eosinophilic cytoplasm and fibrillary stroma with few mitotic figures (Figure 2a,b). Foci of necrosis were present. Immunohistochemistry showed diffusely strong positives for Vimentin and S-100 (Figure 2c). They were negative for GFAP, neurofilament EMA, CD10, Desmin, synaptophysin, chromogranin and PR. (GFAP, Figure 2d). KI 67proliferation index was high. Therefore a final diagnosis of MINST was signed out. Since there was a history of old tumour resected almost 7 years later that showed no apparent infiltration of CNS parenchyma, as would be expected to be diagnosed for a meningioma. The patient is kept on follow-up after the surgery to track recurrence.

3. Discussion

MINST are extremely unusual tumours.[2] Similar to its MPNST counterpart, the diagnosis of MINST can be difficult and necessitates a thorough investigation of clinical, radiological and histopathological findings. The cell of origin of these tumours is uncertain. Some suggest an origin from Schwann cells of perivascular nerves while others suggest pluripotent mesenchymal cells.[4] Joshi. et al found a female child with signs of raised ICP and ataxia as well and he coined the term intracerebellar malignant nerve sheath tumour (ICMNST) to label the cerebellar location of such tumours.[6] Andrea reported the first ICMNST with rhabdomyoblastic differentiation.[5] A comprehensive histopathological analysis is needed along with, immunostaining, and electron microscopy for the confirmative diagnosis. Microscopy shows hypercellular spindle cells with marked cellular and nuclear pleomorphism, interlacing fascicular arrangement, signs of nerve sheath differentiation, presence of syncytial epithelioid tumour cells and multinucleated giant cells, along with foci of necrosis. MINST shows positivity for S-100, similar to our case, but it may be negative in a few cases. Differential diagnosis of MINST includes gliofibroma, gliosarcoma, meningioma, desmoplastic astrocytoma, rhabdomyosarcoma, malignant solitary fibrous tumour and gastrointestinal stromal tumour. These were ruled out by negative stains for GFAP, neurofilament, IDH-1, EMA, PR, CD10, and CD117.[3] Allison et al reported MINST with the help of immunostaining showing tumour cells negative for IDH1 and BRAF (V600E) mutation with ATRX retention and a Ki-67 labelling index of 20% to 30% with a differential diagnosis including gliosarcoma and anaplastic pleomorphic xanthoastrocytoma. It was finally found to be IDH1-R132 and IDH2-R172 wild type on Sanger sequencing, with the absence of mutations in histone H3F3Agene or telomerase reverse transcriptase (TERT) promoter genes.[7] MINST are aggressive tumours with varying outcomes with many having a dismal prognosis[8] Sharma et al. studied that the earlier the first recurrence, the worse the overall survival of the patient. Charles et al did a systematic review of 56 patients with

intracranial MPNSTs from 743 literature results. Median survival was 29 ± 22.1 months with a 1-year survival of 60%. Factors associated with reduced survival were older age, subtotal resection, rhabdomyoblastic histology, and early recurrence (≤6 months) (P = 0.018). They concluded that intracranial MPNSTs are not associated with a cranial nerve or the patient is kept on closed follow-up. Awareness of this tumour, its confirmatory histological examination and extensive surgical extirpation are needed in the definitive management.[2]They have poor prognoses and benefit from aggressive resection, multimodal treatment, and close follow-up. Next-generation sequencing can be helpful for potential targeted therapy.[9] Zaidi et al did another systemic review of 32 cases of cranial MPNST. The most involved cranial nerve was VIII (15/32), followed by the Vth (10/32) and the VIIth (5/32). 4 cases had NF type 1 and 2 had NF type 2. MPNST strongly express protein S-100 and collagen IV-laminin. There were 13 cases treated with radiotherapy for tumour recurrence and metastasis. Fatal outcomes occurred in 66% of patients whereas 19% were reported alive with or without complications. The seven cases were reported to have metastasis to the spine. It takes around a year for recurrence or metastasis. They concluded that MPNST of cranial nerves is very rare with a poor prognosis. A close postoperative follow-up is mandatory to eliminate recurrence.[10]It has high mortality and poor survival with the chances of survival ranging between 1 and 5 years after diagnosis, highlighting the need for a review of the literature.[2] The knowledge attained from this case may help in keeping MINST as a differential diagnosis for a patient with clinical and radiographic signs of an intracranial tumour as MINST have the potential to mimic.

4. Conclusion

MINSTs are rare but aggressive neoplasms with high malignant potential, multiple recurrences and poor prognosis. The similarity of MINST to their peripheral counterparts MPNST helps in the diagnosis which is confirmed through histopathological analysis. Surgery is the cornerstone of treatment. Considering this unusual tumour in a differential diagnosis of a heterogeneously enhancing intracerebral mass with such kind of histomorphology is helpful for the proper management and follow-up of the patient.

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6. Conflict of Interest

None Declared.

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