Child's Nervous System
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CASE REPORT

About a rare pediatric case of myxoid meningioma

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Abstract
Intracranial meningiomas are uncommon in the pediatric group. Myxoid meningioma is a subtype of metaplastic meningioma. It is the rarest WHO grade I meningioma. Only one case of pediatric myxoid meningioma was described in the literature. We report the case of 14-year-old boy who presented with a left parietal dura-inserted tumor. Histological and immunological studies concluded to a myxoid meningioma. We reviewed the literature for this rare pathology and draw attention to the necessity of distinguishing it from other mucinous tumors of the central nervous system, especially chordoid meningioma which has poorer prognosis.

Keywords Meningioma · Myxoid · Child · Neurosurgery

Introduction

Meningiomas are uncommon among children, as they account for only 0.4–4.1% of all pediatric intracranial tumors [15]. According to the latest World Health Organization (WHO) classification, myxoid meningioma (MM) is considered as a metaplastic meningioma (grade I).

To our knowledge, only ten cases of MMs have been described in English literature [9, 11, 13, 14], and only one occurred in a pediatric patient [5].

We present a new rare pediatric case of MM, highlight its histological features, and note relevant differential characteristics with other mucinous tumors.

Case description

A 14-year-old boy, with no medical history, presented in January 2020 with generalized seizures and mild headaches, without further symptoms. The neurological exam was normal. We did not find any café-au-lait spots in the cutaneous exam.

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Brain-computed tomography (CT) scan showed an extra axial left parietal hypodense lesion, with bone erosion (Fig. 1a). Brain magnetic resonance imaging (MRI) showed a lesion that measures 48 × 37.5 × 49 mm. It had a large insertion base on the dura, with a comet tail sign. It had a hyposignal on T1-weighted imaging (WI), a heterogeneous hypersignal on T2-WI, and a heterogeneous peripheral enhancement. The center of the tumor had a T1 hyposignal and T2 hypersignal and was not enhanced (Fig. 1b, c). FLAIR-WI showed a mild peritumoral edema (Fig. 1d).

The patient was operated through a large craniotomy, removing the infiltrated bone. We found a beige soft gelatinous extra-axial mass, well delineated from the adjacent brain (Fig. 2). A one-piece gross total resection (GTR) was performed (Simpson grade I resection). Blood loss was moderate, and no transfusion was needed. A tight duroplasty using a pericranium flap and a cranioplasty were performed.

Histological study found a well-delimited tumoral proliferation of spindle cells, with vesicular nuclei and eosinophilic cytoplasm. It was organized in a whorl formation within a mucoid matrix. There was no evidence of necrosis nor invasion of nearby glial tissue. The mitotic index was low (Ki67 of 3%). The tumor cells exhibited a significant epithelial membrane antigen (EMA) and Alcian blue staining (Fig. 3).

The diagnosis of myxoid meningioma was established.

The post-operative course was uneventful. Postoperative CT scan showed complete removal of the tumor without complications (Fig. 4). The patient was discharged 3 days after surgery under anti-epileptic drug. A genetic study was

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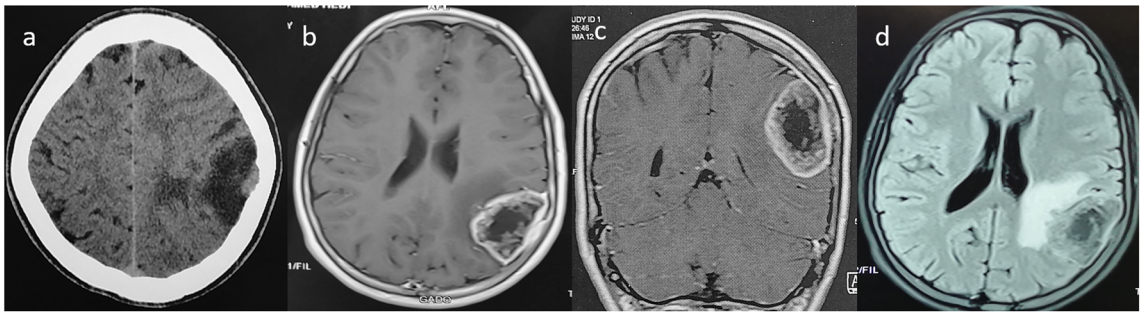

Fig. 1 Preoperative imaging. Brain CT scan (**a**) showing an extra axial left parietal hypodense lesion, eroding the inner table of the bone. Axial (**b**) and coronal (**c**) T1-WI brain MRI showing a hyposignal extra-axial tumor with a heterogeneous, mainly peripheral, enhancement, and a central non-enhanced zone, and with a large insertion base on the dura. Axial FLAIR-WI (**d**) showing a mild peritumoral edema

requested and is still in progress. Since MM is a grade I meningioma, no additional treatment was needed.

At 6-month follow-up, he was seizure free and showed no recurrence at the CT scan.

Discussion

Meningiomas are generally benign tumors arising from meningothelial cells. They are relatively common in adult patients but rare in pediatric population.

The WHO classification identifies metaplastic meningiomas as a subtype of grade I meningiomas [1, 12]. This subtype is characterized by various mesenchymal differentiations including osseous, cartilaginous, lipomatous, leiomatous, xanthomatous, and myxoid tissues, singly or in combinations [4, 13].

The myxoid subtype is among the rarest subtypes of meningiomas. To this day, only ten previous cases have been described [9, 11, 13, 14].

In children, meningiomas are the most common meningeal-based tumors. They represent only 0.4–4.1% of all pediatric intracranial tumors [15]. They usually develop in uncommon locations such as the posterior cerebral fossa, inside the ventricles, and in spinal epidural regions. The absence of dural attachment is more prevalent. Additionally, they are larger in size and more cystic. They tend to have higher intraoperative blood loss. Statistically, there is a slight male predominance.

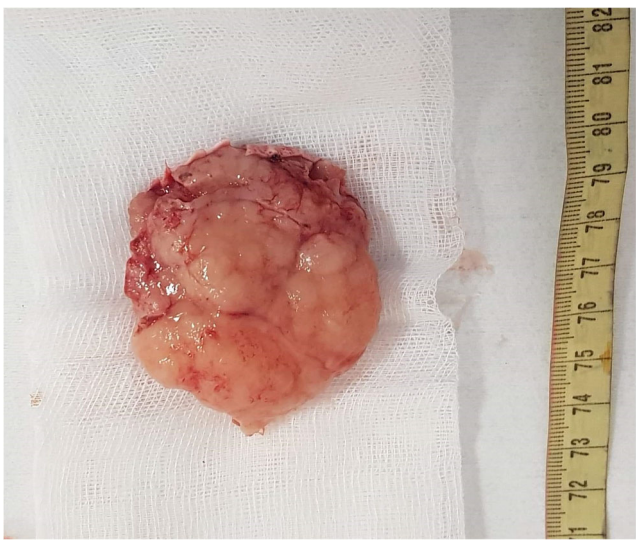

Fig. 2 Photo of operative specimen: a beige soft gelatinous mass

Two major risk factors are associated with meningioma development among children: prior low-dose radiation and type 2 neurofibromatosis (NF2). About 40% of child meningiomas are associated with NF2, which makes genetic testing for NF2 necessary for every child with meningioma [3, 5, 7].

The only published pediatric MM was intraventricular, recorded in a 7-year-old female by Dulai et al. in 2009 (7).

Due to their rarity, the imaging characteristics of MM are not completely identified. MRI found that most MMs were hyperintense on T2-WI [8]. Due to the abundant mucoid matrix and hypervascularization, the enhancement after gadolinium is important [13].

Histologically, the myxoid subtype is characterized by an abundant mucoid matrix in the cytoplasm and large amounts of acidic mucopolysaccharides that stain with Alcian blue [13].

MMs show positive immunological reactivity for vimentin and EMA, as well as type IV collagen, and negative reactivity for cytokeratin, HMB45, S-100, actin, desmin, CD117, neurofilaments, and glial fibrillary acidic protein (GFAP) [10, 13, 14]. As a grade I meningiomas, they have a low Ki-67 (MIB1) proliferation index (usually less than 5%) [6].

MMs should be distinguished from other myxoid or mucinous tumors of the central nervous system, such as metastatic mucinous adenocarcinoma, pilomyxoid astrocytoma, or myxoid chondrosarcoma. Other myxoid spindle cell neoplasms include schwannomas, fibromyxomas, and fibroxanthomas [2, 13].

Immunohistochemistry and ultrastructural analysis are essential to confirm the diagnosis. Radiological study associated to immunohistochemistry should eliminate a metastatic lesion or an embolism of a cardiac myxoma [14].

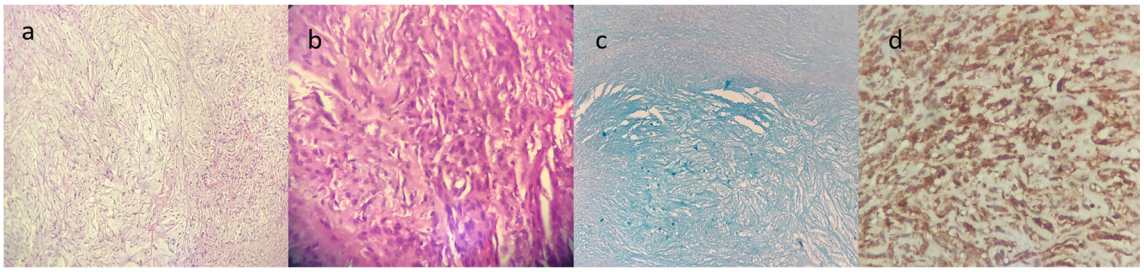

Fig. 3 Pathological examination. Low and high power micrographs showing spindle cell proliferation, with vesicular nuclei and eosinophilic cytoplasm, organized in a whorl formation (**a**, **b**). Alcian Blue staining showcasing the mucoid matrix (**c**). Tumor cells showing positive EMA staining (**d**)

The most difficult and important diagnosis that requires discrimination is chordoid meningioma, which is classified as a grade II neoplasm. Grade II meningiomas have higher recurrence rate and require further therapeutic methods even after GTR. Chordoid meningioma has a higher grade of dyskaryosis and has a reticular arrangement with less cytoplasmic vacuolation. The cells are rather polygonal. There could be an inflammatory infiltrate made of lymphocytes (B cells) [5, 13].

MM has a good prognosis with low recurrence rate, like other grade I meningiomas [5, 11]. Some authors report a higher recurrence rate of MM comparing to other grade I meningiomas [2]. However, there is not enough cases reported to make such conclusions.

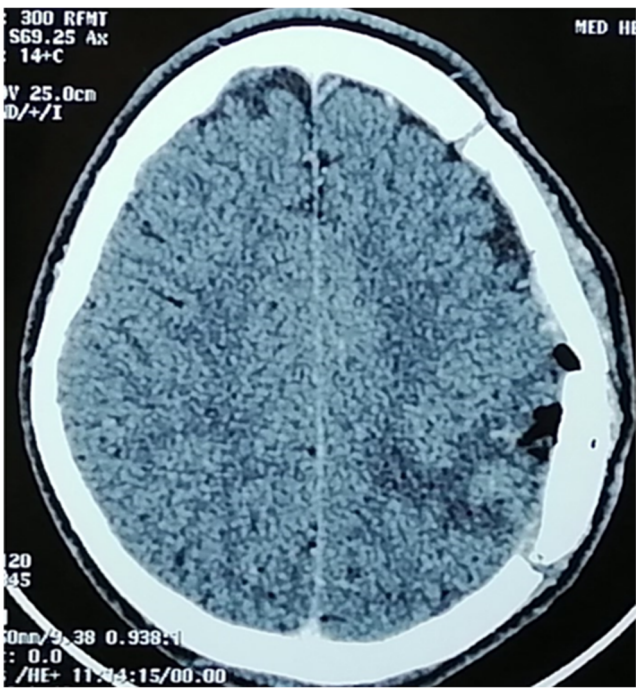

Fig. 4 Postoperative imaging. CT scan showing complete removal of the tumor

Conclusion

MM is a rare grade I meningioma. Only one pediatric case was reported in literature. It has particular histological and immunological characteristics. It has a low recurrence rate and low chance for malignant progression. It should be differentiated from other mucinous tumors, especially chordoid meningioma, a grade II tumor with higher recurrence rate. In pediatric population, NF2 must be excluded.

Abbreviations WHO, World Health Organization; MM, Myxoid meningioma; CT, Computed tomography; MRI, Magnetic resonance imaging; WI, Weighted imaging; GTR, Gross total resection; EMA, Epithelial membrane antigen; NF2, Type 2 neurofibromatosis

Declarations

Conflict of Interest No funds, grants, or other support was received. The authors have no relevant financial or non-financial interests to disclose. The authors have no conflicts of interest to declare that are relevant to the content of this article.

References

1. Barresi V, Caffo M, Tuccari G (2016) Classification of human meningiomas: lights, shadows, and future perspectives: Classification of Meningiomas: Value and Limits. J Neurosci Res 94:1604–1612. https://doi.org/10.1002/jnr.23801
2. Bégin LR (1990) Intriguing case: myxoid meningioma. Ultrastruct Pathol 14:367–374. https://doi.org/10.3109/01913129009032251
3. Caroli E, Russillo M, Ferrante L (2006) Intracranial meningiomas in children: report of 27 new cases and critical analysis of 440 cases reported in the literature. J Child Neurol 21:31–36. https://doi.org/10.1177/08830738060210010801
4. Choi Y-H, Choi C-Y, Lee C-H, Koo H-W, Chang S-H (2018) Metaplastic meningioma overspreading the cerebral convexity. Brain Tumor Res Treat 6:97. https://doi.org/10.14791/btrt.2018.6.e17
5. Dulai MS, Khan A-M, Edwards MSB, Vogel H (2009) Intraventricular metaplastic meningioma in a child: case report and review of the literature. Neuropathology 29:708–712. https://doi.org/10.1111/j.1440-1789.2009.01008.x
6. Gray F, Duyckaerts C, De Girolami U (2019) Escourolle & Poirier's manual of basic neuropathology, 6th edn. Oxford University Press, Oxford

7. Hong S, Usami K, Hirokawa D, Ogiwara H (2019) Pediatric meningiomas: a report of 5 cases and review of literature. Childs Nerv Syst 35:2219–2225. https://doi.org/10.1007/s00381-019-04142-y
8. Johnson MD, Hussain A (2019) Imaging and clinicopathologic features of myxoid meningiomas. Clin Neuropathol 38:238–244. https://doi.org/10.5414/NP301139
9. Johnson MD, Stevenson CB, Thompson RC, Atkinson J, Boyer P (2007) December 2006: 31-year-old woman with hemiparesis. Brain Pathol 17:255–257. https://doi.org/10.1111/j.1750-3639.2007.00068_3.x
10. Kimura Y, Matsumae M, Tsutsumi Y (1998) Pericellular deposition of basement membrane material in myxoid meningioma: immunohistochemical evidence for unbalanced production of type IV collagen and laminin. Pathol Int 48:53–57. https://doi.org/10.1111/j.1440-1827.1998.tb03828.x
11. Krisht KM, Altay T, Couldwell WT (2012) Myxoid meningioma: a rare metaplastic meningioma variant in a patient presenting with intratumoral hemorrhage. JNS 116:861–865. https://doi.org/10.3171/2011.12.JNS111020
12. Louis DN, Perry A, Reifenberger G, von Deimling A, Figarella-Branger D, Cavenee WK, Ohgaki H, Wiestler OD, Kleihues P, Ellison DW (2016) The 2016 World Health Organization Classification of Tumors of the Central Nervous System: a summary. Acta Neuropathol 131:803–820. https://doi.org/10.1007/s00401-016-1545-1
13. Marutani A, Nakano R, Nishi N, Yamada T (2019) Histological characteristics of myxoid (metaplastic) meningioma in a 44-year-old woman. Case Rep Oncol Med 2019:1–4. https://doi.org/10.1155/2019/9207632
14. Ortiz J, Ludeña MD, Gonçalves J, Carmen SD, Maillo Á, Bullon A (2013) Myxoid meningioma: an example of a rare brain tumour of difficult diagnosis. OJPathology 03:51–53. https://doi.org/10.4236/ojpathology.2013.31009
15. Stettner GM, Rostasy KM, Ludwig HC, Merkler D, Fahsold R, Gärtner J (2007) Infratentorial meningioma in an 8-year-old child as first sign of neurofibromatosis type 2. Neuropediatrics 38:29–31. https://doi.org/10.1055/s-2007-980204