Quiz

Correct answer to the quiz. Check your diagnosis

Case report

Pigmented lesion of the central nervous system: what is it?

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Meningiomas are the most common primary central nervous system (CNS) neoplasms in adults, arising from arachnoid cells as extra-axial tumours. We report a rare case of World Health Organization (WHO) grade 2 metaplastic meningioma with lipomatous differentiation in a 70-year-old man presenting with persistent headaches. Magnetic resonance imaging revealed a dyshomogeneous left parieto-occipital extra-axial mass with calcific components and dural thickening suggestive of infiltration, confirmed histologically by brain parenchyma invasion, atypical cell proliferation with hemosiderin-containing brownish pigmentation (Prussian blue-positive), and lipoblast-like elements. Immunohistochemistry showed positivity for epithelial membrane antigen and progesterone receptor, with negativity for S100, glial fibrillary acidic protein, and Olig2, confirming meningothelial lineage and excluding melanocytic or glial neoplasms. According to WHO CNS 2021, brain parenchyma invasion warranted a grade 2 designation; however, the prognostic implications of this criterion in metaplastic meningioma remain to be fully established.

Key words: meningioma, metaplastic meningioma, lipomatous meningioma, brain invasion, hemosiderin pigmentation, WHO CNS 2021, parenchymal infiltration.

Introduction

Meningioma is the most common primary neoplasm of the CNS in adults, originating from arachnoid cells and thus classified as extra-axial. However, meningiomas can rarely infiltrate the glial parenchyma of the brain, which warrants the assignment of grade 2 according to WHO-CNS 2021, as seen in this case. This infiltration complicates the neuroradiological identification of the extra-axial origin.

Currently, 15 subtypes of meningioma are recognized, with the metaplastic subtype being one of the rarest. It is characterized by differentiation into osseous, cartilaginous, lipomatous, myxoid, or xanthomatous components [1].

Case report

A 70-year-old male presented with persistent headaches. A magnetic resonance imaging revealed a left parieto-occipital extra-axial lesion characterized by a dyshomogenous iso-intense signal in T1, isointense signal in T2, and a large central and peripheral component in T2 with low signal, possibly indicating a partly calcific matrix. The lesion exhibited dyshomogenous contrast enhancement, with

areas along the ependymal surface of the ventricle in the peritrigonal location, and showed evident thickening of the dural lining in the posterior third of the falx cerebri and superior sagittal sinus, suggestive of infiltrative behavior.

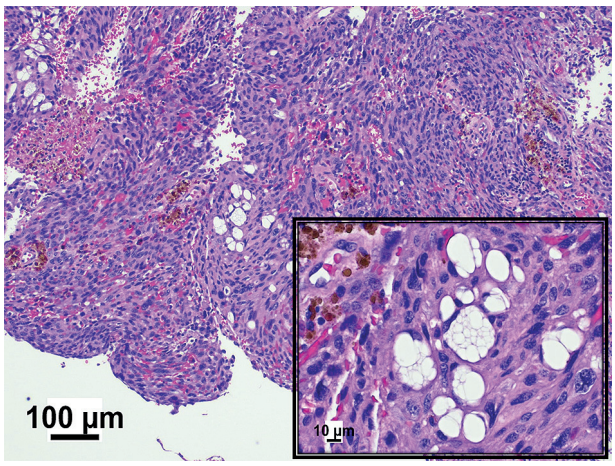

Figure 1. Photomicrograph showing a compact proliferation of atypical elements associated with areas of brownish pigmentation and areas of clear cytoplasmic cells (HE, 10×)
The latter elements show a cytologic lipoblast-like morphology at higher magnification (inset, HE, 60×).

Histological examination revealed a proliferation with markedly increased cellularity and a compact pattern of medium to large, slightly elongated cells with discrete nuclear pleomorphism and focal brownish pigmentation. Scattered lipoblastic/lipoblast-like elements were also noted (Figure 1, inset). The lesion demonstrated brain parenchyma infiltration and leptomeningeal spread, confirming the neuroradiological suspicion. A further microscopic study with histochemical staining for Prussian blue was positive (Figure 2A), and immunohistochemical analyses revealed positivity for epithelial membrane antigen (EMA) and progesterone (Figure 2B, C), while S100 was negative (Figure 2D). The lesion was also negative for glial fibrillary acidic protein and Olig2.

Results

The final diagnosis is a meningioma, metaplastic (lipomatous) subtype, according to the WHO classification of central nervous system tumours of 2021 (WHO-CNS 2021). While the metaplastic lipomatous subtype does not inherently indicate worse behavior [2], in this instance, it exhibited an infiltrative pattern.

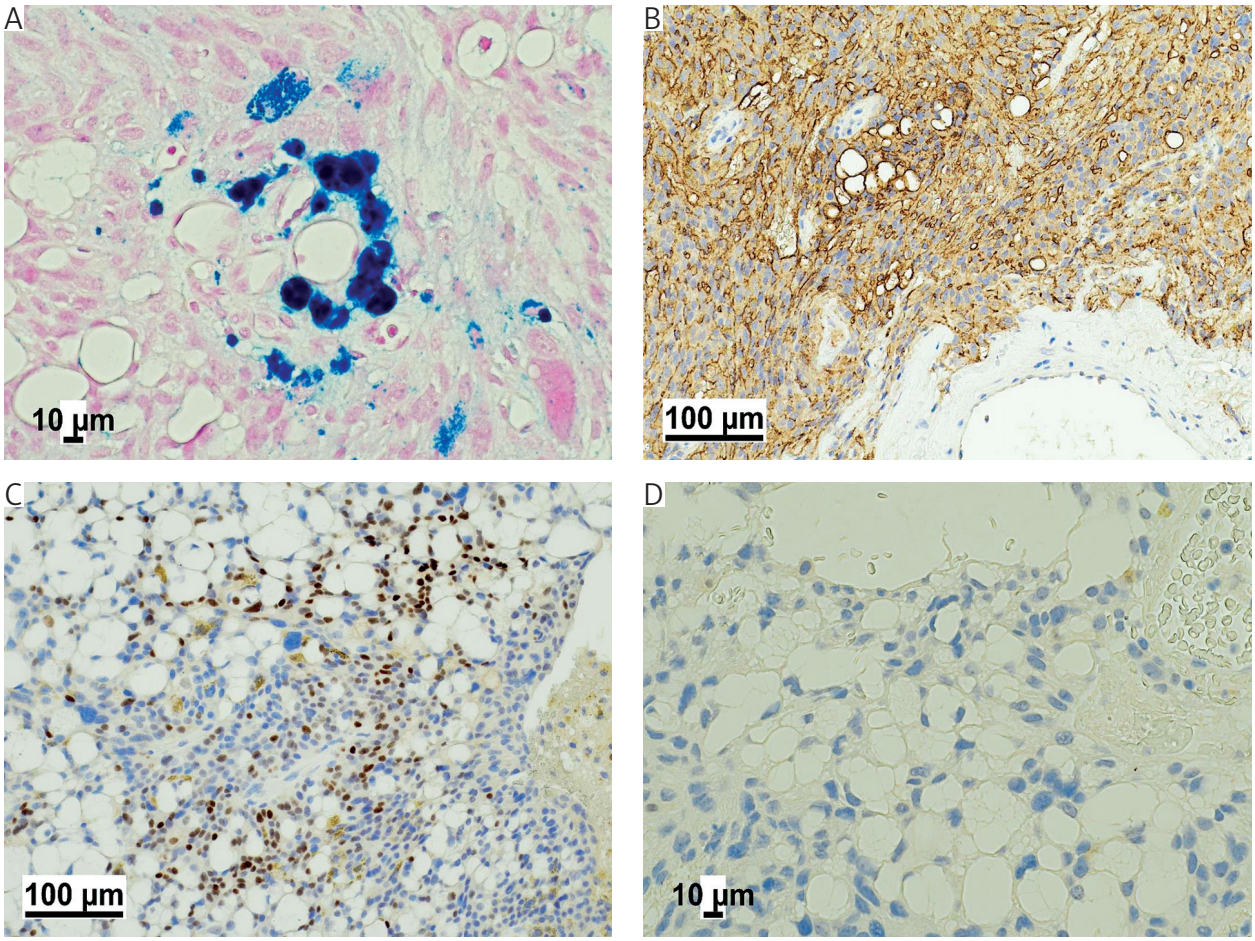

Figure 2. Histochemistry and immunohistochemistry. **A**) The pigmented deposits are positive for Prussian blue staining. **B**) Diffuse positivity for epithelial membrane antigen is noted. **C**) Nuclear positivity for progesterone is observed in scattered elements. **D**) Widespread negativity for S100 is evident

Discussion

Meningiomas may show pigmentation as well as other non-melanocytic primitive CNS neoplasms (e.g. gliomas, schwannomas, medulloblastomas), which are part of the differential diagnosis [3, 4]. It is also important to consider true melanocytic lesions in the differential diagnosis, both metastatic (even if in this case there was no suspicion of melanoma on admission) and primary CNS lesions [5], especially as the leptomeningeal location of the latter complicates the differential diagnosis and makes the microscopic distinction between pigmented entities based on hematoxylin-eosin staining alone very difficult.

Further histochemistry and immunohistochemistry clarified the issue:

- positivity for Prussian blue indicated that the pigmentation was due to hemosiderin, not melanin,
- negativity for S100 excluded a melanocytic lesion (either primary CNS or metastatic),
- negativity for glial fibrillary acidic protein and Olig2 excluded a glial metastatic),
- positivity for EMA and progesterone confirmed the diagnosis of a meningioma, albeit in its exceedingly rare form as a grade 2 metaplastic/lipomatous subtype.

One final point about grading: according to the WHO-CNS 2021 classification system, invasion of the brain parenchyma is recognized as an independent histopathological criterion sufficient to assign a grade 2 (G2) designation to any meningioma subtype, including metaplastic subtype [1]. This criterion is applied regardless of other histological features, reflecting the consensus that brain invasion signifies a higher risk of recurrence and warrants a grade 2 designation. However, the neuro-oncology community continues to debate whether meningiomas with brain parenchyma invasion, including metaplastic meningiomas, which have traditionally been considered biologically indolent, consistently demonstrate the aggressive clinical behavior associated with grade 2 lesions [6].

Conclusions

Although we have classified this case as grade 2 in accordance with the WHO-CNS 2021 guidelines, we acknowledge the need for further studies to clarify the prognostic and predictive implications of brain invasion in this histotype.

Disclosures

1. Institutional review board statement: Not applicable.
2. Assistance with the article: None.
3. Financial support and sponsorship: None.
4. Conflicts of interest: None.

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