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Giant hypervascular Nauta type IV cystic atypical meningioma in a young adult: Radiologic-pathologic correlation

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ABSTRACT

Cystic meningiomas are uncommon and may mimic intra-axial neoplasms, particularly in young patients. An 18-year-old patient presented with progressive morning headaches, vomiting, blurred vision, and left-sided weakness. Magnetic resonance imaging demonstrated a 71 × 63 × 69 mm multilobulated right frontal convexity extra-axial tumor with a broad dural attachment, falcine extension, an intensely enhancing hypervascular solid component, internal necrotic change, and an inferior peritumoral cyst located between the tumor and the brain, consistent with Nauta type IV. Despite limited vasogenic edema, the lesion produced a 19-mm midline shift, subfalcine herniation, early right uncal herniation, and obstructive dilatation of the left lateral ventricle with transependymal cerebrospinal fluid flow. Simpson grade II resection was achieved. Histopathology showed syncytial meningothelial proliferation, eight mitoses per 10 high-power fields, and microscopic brain invasion, establishing an atypical meningioma, central nervous system World Health Organization grade 2. This case emphasizes the diagnostic value of recognizing extra-axial signs and the cyst-tumor-brain relationship, while using heterogeneous enhancement, necrosis, marked vascularity, and an indistinct tumor-brain interface as warning features rather than as surrogates for histologic grade.

KEYWORDS

Atypical meningioma; cystic meningioma; brain invasion; Nauta classification; magnetic resonance imaging

MAIN ARTICLE

INTRODUCTION

Meningiomas are usually recognized by a broad dural attachment, intense enhancement, a cerebrospinal fluid cleft, and inward displacement of the adjacent cortex. Cystic change is uncommon and can obscure these features, producing an appearance that resembles a high-grade glioma, metastasis, or another solid-cystic tumor.[1]

Preoperative magnetic resonance imaging (MRI) cannot assign a definitive meningioma grade, but irregular morphology, heterogeneous enhancement, necrosis, extensive vascularity, edema, and loss of the tumor-brain interface may raise concern for higher-grade behavior. We report a giant Nauta type IV cystic meningioma in a young adult, with radiologic-surgical-pathologic correlation confirming central nervous system World Health Organization (CNS WHO) grade 2.[4]

CASE REPORT

An 18-year-old patient followed for axial spondyloarthritis presented with a 6-week history of progressive frontal headaches, worse in the morning, recurrent vomiting, intermittent blurred vision, and gradually increasing weakness of the left upper and lower limbs. There was no history of seizures, cranial irradiation, or known malignancy. The patient was alert and oriented. Examination demonstrated bilateral papilledema and a mild left pyramidal syndrome, with muscle strength of 4/5 in the left upper limb and 4+/5 in the left lower limb.

MRI demonstrated a large right frontal convexity extra-axial mass measuring 71 × 63 × 69 mm. The lesion was markedly multilobulated, had a broad dural attachment with falcine extension, and contained solid, cystic, and necrotic components. The viable solid tissue was heterogeneously hypointense on T1-weighted images and mildly hyperintense on T2-weighted and fluid-attenuated inversion recovery images. It did not show marked diffusion restriction. Numerous intratumoral and peritumoral flow voids were present (Figure 1).

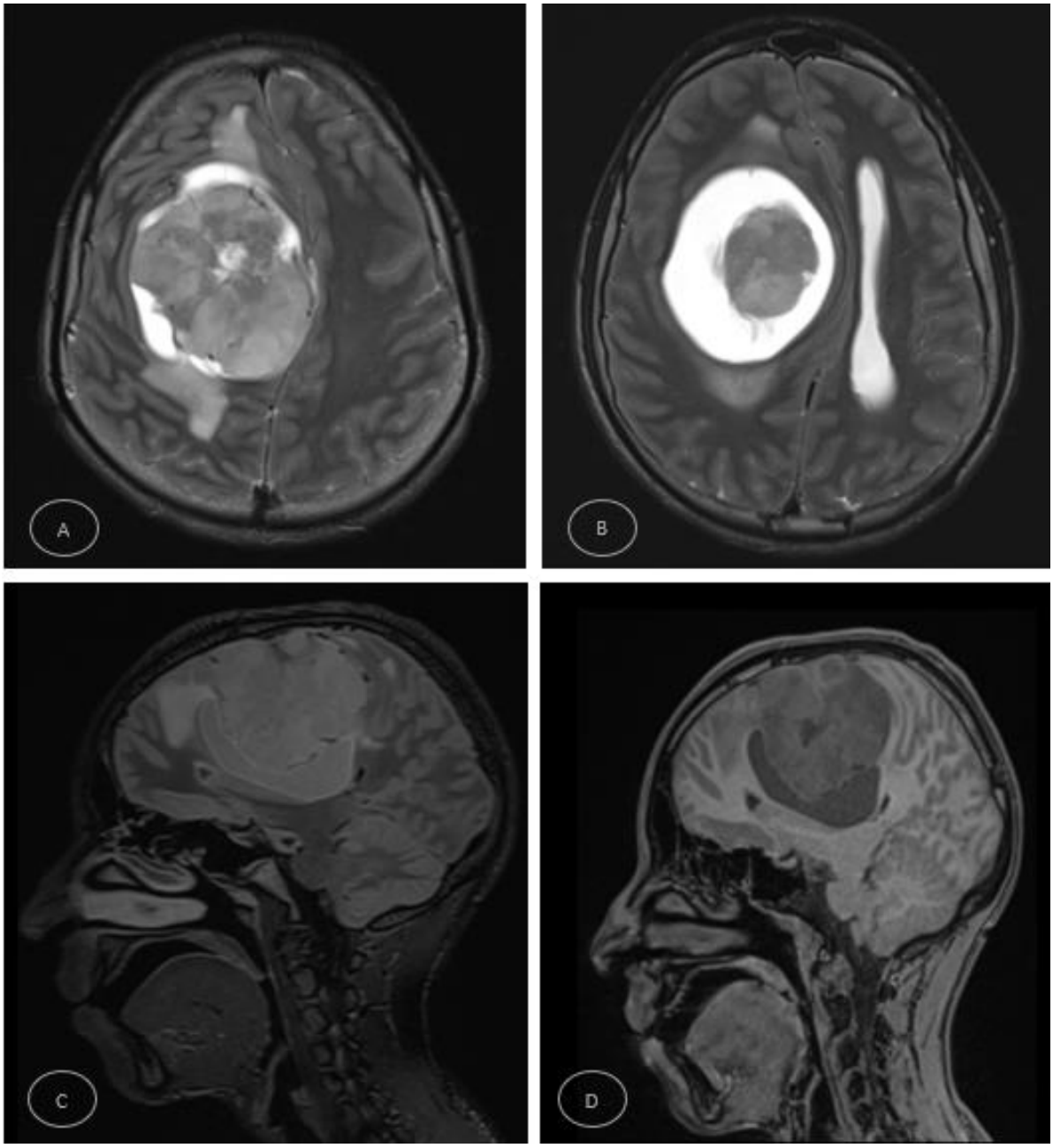


Figure 1: (a and b) Axial T2-weighted images show a giant multilobulated right frontal convexity extra-axial mass with solid and cystic components, severe mass effect, and contralateral ventricular dilatation. (c) Sagittal fluid-attenuated inversion recovery and (d) sagittal T1-weighted images demonstrate the inferior peritumoral cyst located between the tumor and compressed frontal lobe, supporting a Nauta type IV configuration.

After gadolinium administration, the viable component enhanced intensely and heterogeneously, with adjacent dural-tail enhancement. The inferior cyst was located between the solid tumor and the compressed frontal cortex, approached cerebrospinal fluid signal, and

had no definite enhancing wall. This tumor-cyst-brain relationship was consistent with Nauta type IV. Susceptibility-weighted imaging demonstrated intratumoral vascular structures, and perfusion imaging showed markedly increased relative cerebral blood volume. Proton MR spectroscopy showed increased choline with reduced creatine and no convincing alanine peak (Figure 2).[3]

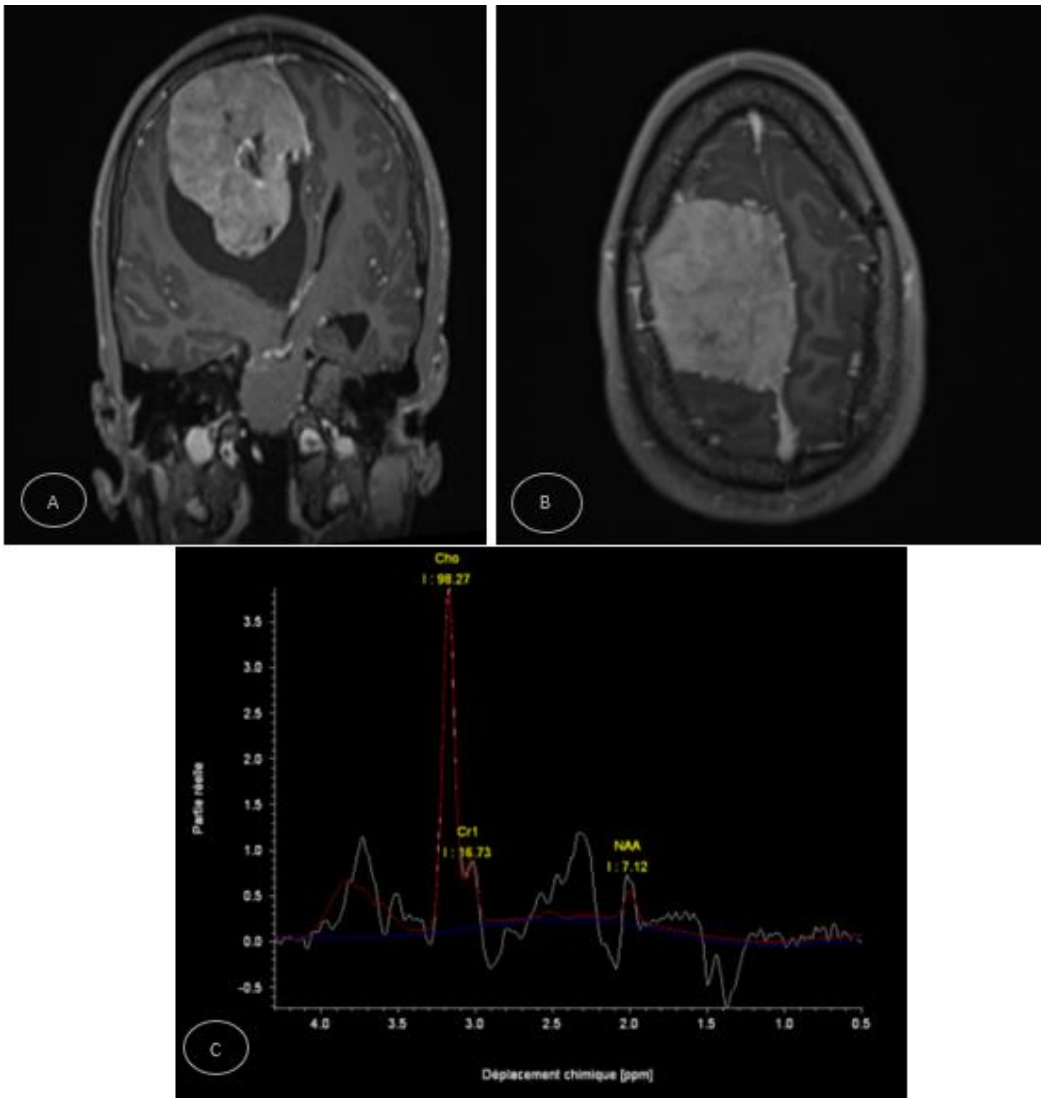


Figure 2: (a and b) Axial and coronal contrast-enhanced T1-weighted images demonstrate intense heterogeneous enhancement, a broad right frontal dural attachment, falcine extension, and adjacent dural-tail enhancement. (c) Proton MR spectrum shows increased choline with reduced creatine and no convincing alanine peak.

Although surrounding vasogenic edema was limited, the tumor caused approximately 19 mm of leftward midline shift, subfalcine herniation, early right uncal herniation, compression of the

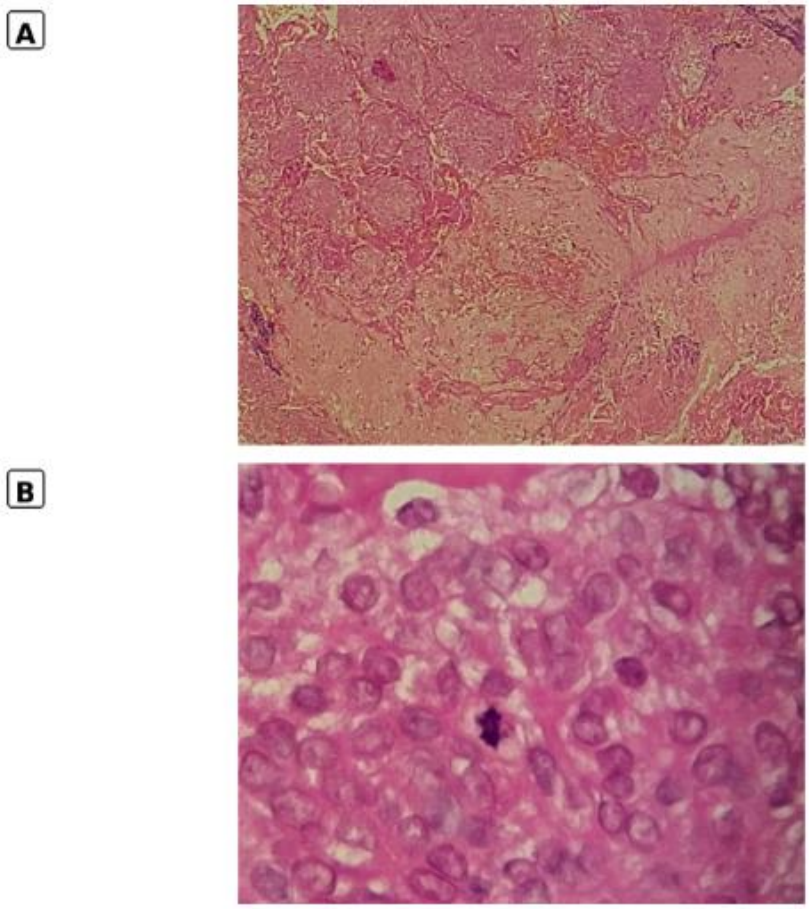
right lateral ventricle, and obstructive dilatation of the left lateral ventricle with transependymal cerebrospinal fluid permeation. The superior sagittal sinus remained patent, without definite intraluminal extension.[6]

Urgent right frontal craniotomy was performed. A firm, multilobulated, highly vascular extra-axial tumor with a broad convexity attachment and falcine extension was identified. The inferior cyst contained clear xanthochromic fluid. Macroscopically complete resection was achieved; the operative report classified the resection as Simpson grade II.

Histopathologic examination showed broad sheets of syncytial meningothelial cells with occasional whorls. Mitotic activity reached eight mitoses per 10 high-power fields. Tumor nests infiltrated adjacent cerebral parenchyma, confirming microscopic brain invasion (Figure 3). These findings established an atypical meningioma, CNS WHO grade 2.[7]

Postoperative clinical and imaging follow-up were not included in the material available for preparation of this report.

Figure 3: Histopathology of the resected tumor. (a) Low-power hematoxylin and eosin microphotograph shows syncytial meningothelial proliferation infiltrating adjacent cerebral parenchyma (original magnification ×10). (b) High-power hematoxylin and eosin microphotograph demonstrates a mitotic figure in a tumor with increased mitotic activity (eight mitoses per 10 high-power fields; original magnification ×40).



DISCUSSION

The main diagnostic challenge was the combination of young age, giant size, and a prominent cystic component. Nauta type IV cysts are peritumoral, situated between the tumor and brain, with a wall formed by arachnoid rather than tumor. Correct classification is surgically relevant because the cyst wall does not necessarily require the same resection strategy as an intratumoral neoplastic wall.[2]

In this case, the broad dural base, dural tail, cerebrospinal fluid cleft, and inward cortical displacement established an extra-axial origin. Marked lobulation, heterogeneous enhancement, internal necrotic change, prominent vascular channels, and a focally indistinct tumor-brain interface raised suspicion for atypical behavior. However, the absence of marked diffusion restriction did not exclude grade 2 disease because apparent diffusion coefficient values overlap among meningioma grades.[4]

Perfusion confirmed hypervascularity and helped anticipate operative blood loss, but high relative cerebral blood volume is not independently diagnostic of grade. Likewise, an absent alanine peak does not exclude meningioma. Histopathology therefore remained decisive. In the present tumor, both eight mitoses per 10 high-power fields and microscopic brain invasion supported CNS WHO grade 2.[9]

Meningiomas are uncommon in children and adolescents, and published cohorts suggest a greater proportion of atypical locations, higher-grade tumors, or aggressive presentations than in typical adult practice. The severe clinical deterioration in this patient was driven by tumor volume and ventricular obstruction despite relatively limited surrounding edema.[8]

Differential diagnosis

The principal alternatives were a solitary fibrous tumor, high-grade glioma, dural metastasis, and pleomorphic xanthoastrocytoma. The extra-axial signs and broad dural attachment favored meningioma, while definitive distinction from another dural tumor required histopathology.

The main differentiating features are summarized in Table 1.

CONCLUSION

A giant cystic meningioma in a young adult may mimic an intra-axial aggressive neoplasm. Recognition of the extra-axial origin and the Nauta type IV relationship between cyst, tumor, and brain narrowed the diagnosis. Heterogeneous enhancement, necrosis, hypervascularity, severe mass effect, and an indistinct tumor-brain interface should prompt careful pathologic sampling, but CNS WHO grading must remain histologic.

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Conflicts of interest: There are no conflicts of interest.

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Data availability: Data supporting this case are available from the corresponding author subject to institutional and patient-privacy restrictions.

TABLE

Table 1: Differential diagnosis of a giant solid-cystic dural-based tumor in a young patient

Diagnosis	Potentially supportive features	Features favoring the reported diagnosis
Solitary fibrous tumor	Dural-based, lobulated, strongly enhancing, and hypervascular	Cystic appearance is less typical; definitive distinction requires STAT6 testing
High-grade glioma	Necrosis, heterogeneous enhancement, severe mass effect	Broad dural base, cerebrospinal fluid cleft, and inward cortical displacement indicate extra-axial origin
Dural metastasis	Dural attachment, necrosis, and edema	No known primary malignancy, young age, and solitary non-destructive lesion
Pleomorphic xanthoastrocytoma	Young age, superficial solid-cystic tumor	Usually intra-axial and cortically based without convincing extra-axial signs
Supratentorial ependymoma	Large heterogeneous solid-cystic tumor in a young patient	Usually intra-axial; broad dural attachment and dural-tail enhancement are atypical

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