Supratentorial Ectopic Ependymoma

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ABSTRACT: Background: Ependymomas usually arise from the ventricular surface. Methods: We report an 11-year-old female who presented with a supratentorial ectopic ependymoma. Results: The patient presented with a two-month-history of progressive headache, nausea and vomiting. Examination revealed papilledema, horizontal nystagmus, diplopia on upward gaze, and right pronator drift. CT scan showed an enhancing left precentral subcortical lesion measuring 3 cm in diameter with associated edema and mass effect. Its medial border was located 3 cm from the ependymal surface of the ventricle. A firm tumour was dissected from the centrum semiovale white matter, and removed in toto as confirmed on MRI. Pathological examination revealed histological, immuno-histochemical and electron microscopic features consistent with an ependymoma. Spine MRI and bone marrow aspirate, as well as lumbar puncture of cytology failed to show any dissemination. Conclusion: From the literature review, this represents an exceptional ependymoma located at the distance from the ventricular system or cisterns. Different pathogenic alternatives are discussed.

On gross inspection, the surgical specimen was composed of multiple fragments of hemorrhagic, white and grey soft tissue. Histological sections revealed that the lesion was very cellular and well vascularized. There were numerous large pseudorosette formations. The cells of these formations had their nuclei aligned around central vessel, with the eosinophilic cytoplasmic processes directed toward the vessel. The nuclei were quite uniformly oval with mild polymorphism and hyperchromaticity. Rare mitotic figures and true rosettes were seen. Large areas of necrosis were visualized in the tumour (Figure 2). Within the tumour, immuno-chemistry showed that neuron-specific enolase (NSE) was equivocal, synaptophysin was negative, and glial fibrillary acidic protein (GFAP) was positive in some tumour cells. Cytokeratin and epithelial membrane antigen (EMA) had negative reactivity. Ultrastructural study was done on a fragment of tumour fixed in gluteraldehyde. It showed the tumour cells to be made of large nuclei, often cleaved, with thin chromatin at the periphery. The cytoplasm contained numerous organelles, rough endoplasmic reticulum, mitochondriae, lysosomes, numerous microfilaments, long cytoplasmic processes filled with microfilaments and microtubules. There were some cells with centrioles, others with immature ciliary bodies, and numerous very long junctional complexes at the apex of cells (“zipper-like” junctions). Some cells surrounded a lumen and projected microvilli in the lumen (Figure 3). These findings suggested a Grade II-IV ependymoma according to the WHO revised classification of 1993. Necrosis is not an infrequent finding in ependymoma and is not necessarily indicative of a more aggressive behaviour.

The post-operative course was uneventful with complete resolution of symptoms present on admission. Papilledema, however, persisted during the hospitalization. Post-operative CT scan and MRI confirmed the complete tumour resection. Further work-up, including spine MRI, bone marrow aspirate as well as lumbar puncture for cytology failed to show any dissemination.

**DISCUSSION**

The radiological differential diagnosis of such a lesion included a high grade glioma, a primitive neuro-ectodermal tumour, a metastasis, as well as an atypical meningioma, given the superficial extension noted on several sections of the CT scan. A hemangioblastoma, as well as a ganglioglioma, were also part of the differential diagnosis.

This case involves a supratentorial ependymoma of unusual location and, in particular, without attachment to the ventricular system. Our review of the literature reveals that in older series, the occurrence of supratentorial ependymomas, without any visible attachment to the ventricular system, seemed to be a frequent feature, reported in more than one-half of the cases. However, these studies were all conducted during the pre-CT era.

**Figure 1:** Contrast-enhanced computerized tomography scan demonstrating a 3 cm diameter contrast-enhancing lesion in the left frontal region with large amount of associated edema (A). There is no continuity between the lesion and the ventricular system (B).

**Figure 2:** Photomicrograph showing cellular proliferation in pseudo rosette arrangement (HPS, 20X).
The uniform and differentiated histological features of this tumour, and the relatively old age of the patient, make this possibility unlikely. Finally, a more tempting hypothesis is to postulate that the lesion described here might be the result of a neoplastic growth within an ectopic ependymal cell of the frontal cortex or adjacent leptomeninges and, therefore, at least in part, a consequence of a migrational error.

In conclusion, this report points out that although exceptional in this location, ependymoma has to be included in the radiological differential diagnosis of a contrast-enhancing parenchymal mass lesion even in the absence of any visible connection with the ventricular system.

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REFERENCES


