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Case Report & Case Series

Papillary glioneuronal tumour: Case report

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ABSTRACT

Papillary Glioneuronal tumour is a new entity (WHO Grade I) with only 97 reported cases in the literature. These typically well demarcated tumours are histologically and radiologically distinct, however, sometimes there is divergence from the typical characteristic findings as seen in our case. In this case report, we describe the clinical, radiological and histological findings of this entity and further discuss the management and prognosis of this tumour.

1. Introduction

Amongst the mixed glioneuronal tumours, 'Papillary Glioneuronal Tumour' (PGNT) is a new variant and has been described recently as a distinct neurooncological entity (WHO Grade I) in the 2007 classification guidelines [1]. To date, approximately 97 cases have been reported in the literature [1–3]. These typically well demarcated tumours are histologically and radiologically distinct. Our case report shows previously described characteristic histological features such as biphasic architecture, pseudopapillary pattern, lack of anaplastic features associated with compact areas composed of neuronal elements in different maturation states [2]. The distinct finding in our case was absence of cystic change on radiology and histology. In addition there was lack of proximity to ventricles.

2. Case report

We present a case report of a PGNT in a 40 year old right handed woman who presented with a six week history of increasingly severe headache, dizziness, falls and lethargy. She was neurologically normal on examination and without papilloedema, hemiparesis or visual field defect. CT and MRI of brain showed a well rounded moderately enhancing intra axial solid lesion in left parietal lobe with disproportionately extensive vasogenic oedema. The lesion measured 21 × 21 × 24 mm and demonstrated some calcific foci within (Fig. 1). There was demonstration of significant mass effect with effacement of left occipital horn and generalised sulcal and cisternal effacement. There was rightwards subfalcine herniation and uncal herniation. No destructive bony lesions were seen in the skull.

On MRI, the tumour demonstrated decreased ADC (Apparent Diffusion Coefficient), and low T2 signal consistent with hypercellularity. On perfusion MRI, there was markedly elevated CBV (Cerebral Blood Volume). Differential diagnoses on imaging were glial tumour or metastasis. CT scan of chest and abdomen was unremarkable.

Left parietal craniotomy with stereotactic debulking of tumour was performed. Intraoperatively, the tumour was greyish-brown and vascular in appearance with extensive surrounding white matter oedema. A complete macroscopic excision was performed and the specimen was sent for histological examination.

Histological examination showed a biphasic tumour which had both glial and neuronal components. The tumour had pseudopapillary structures which were composed of hyalinised blood vessels. Immediately adjacent to the fibrovascular core were spindled to cuboidal cells. Between the pseudopapillary structures, were small to intermediate sized cells which had uniform round nuclei, salt and pepper chromatin and small nucleoli (Fig. 2). Some of these cells showed perinuclear halo. On immunohistochemistry, the cytoplasm and processes of spindled and cuboidal cells stained positive for GFAP (Fig. 3) indicative of glial cell origin, and the second population of cells stained strongly with synaptophysin (Fig. 3) confirming their neuronal lineage. Mitosis was rare. Vascular proliferation and necrosis were not identified. Cortical dysplasia was not observed in peritumoral brain tissue. The tumour had a very low Ki67 proliferative index, about 1%. Although imaging showed calcification, it was not identified on histology. Based on the histological findings and immunoprofile, a diagnosis of PGNT (WHO grade 1) was made.

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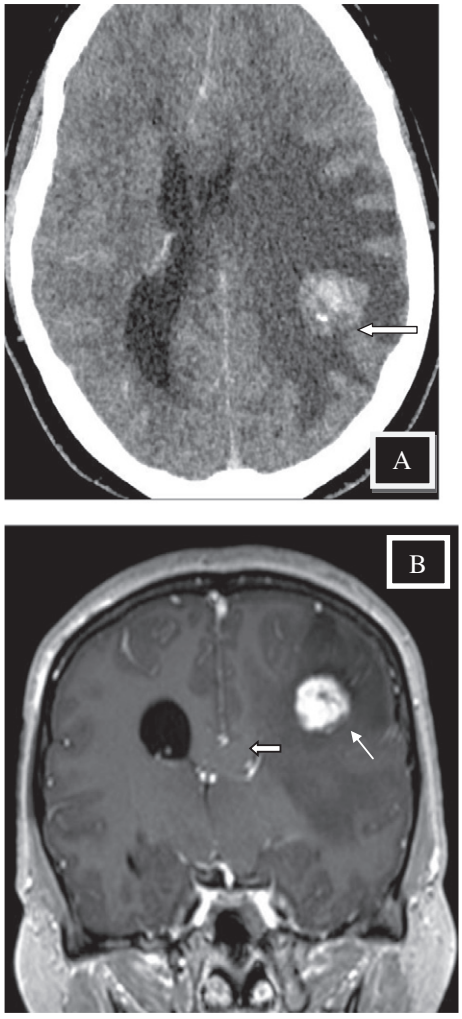

Fig. 1. (A) Post contrast CT shows heterogenous lesion with intralesional calcification (arrows) and intense enhancement. (B) Coronal MRI post contrast image demonstrating avidly enhancing left parietal tumour (line arrow), with marked perilesional edema, mass effect, compressed left ventricle and rightward subfalcine herniation (block arrow).

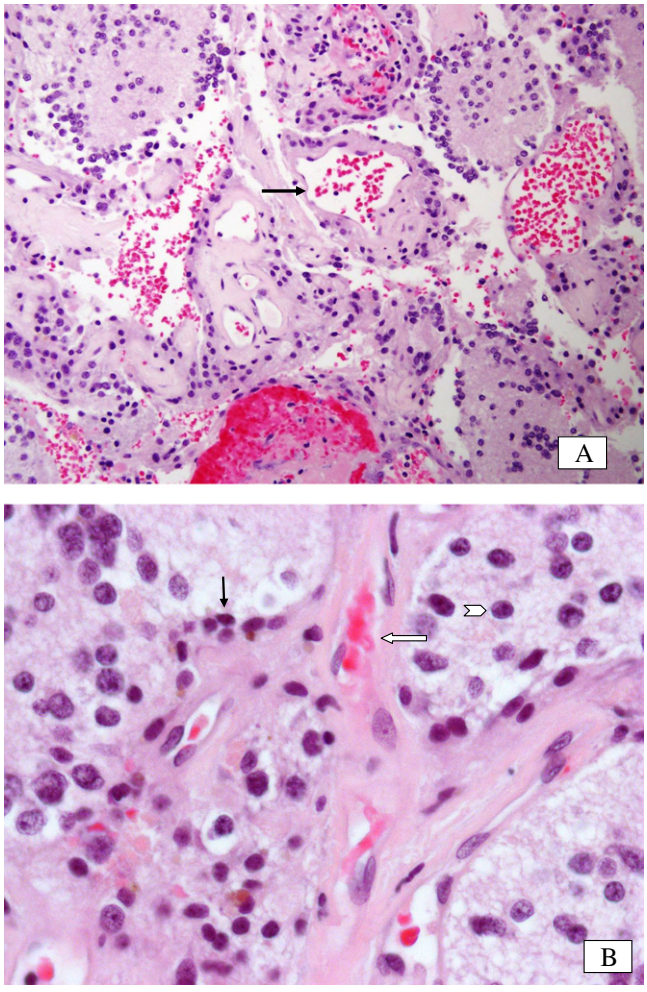

Fig. 2. (A) H&E showing characteristic pseudopapillary (arrow) architecture of the tumour. (B) Hyalinised fibrovascular cores (block arrow) lined by cuboidal cells (line arrow) and cells in interpapillary stroma show salt and pepper chromatin (arrow head).

3. Discussion

PGNT was first described by Komori et al. [4] in 1998. The tumour affects a wide age range with predilection to young adults. No sex predilection has been observed. All lobes have been reported to be involved, however, predilection to temporal lobe has also been reported [1]. Commonly, lesions lie in proximity to ventricles [1].

Radiologically, many lesions present with solid and cystic components. Cyst with mural nodule architecture is thought to be typical, which leads the differential diagnosis to pilocytic astrocytoma, pleomorphic xanthoastrocytoma and ganglioglioma [1,2]. In our case, the important difference was lack of distinct cystic formation. Other important observation, was lack of proximity to ventricles. The tumour was located in the parietal lobe with mass effect on left occipital lobe. Radiologically it was thought to be metastases or glial tumour and there was low suspicion for PGNT, which emphasize the need for better radiologic characterization of this entity.

The histogenesis of these tumours is not exactly known [5]. The proximity of the majority of PGNT lesions to the ventricle implies that the presumptive origins are the multipotent subependymal stem cells; this hypothesis was partially confirmed by the biphenotypic differentiation and expression of stem cell markers [6]. Recently, immunohistochemical expression of Olig2 in a fraction of tumour cells has been reported, suggesting an additional oligodendroglial or at least oligodendroglia-like tumour cell component [5].

On histology, the major differential diagnoses considered were extraventricular neurocytoma, ependymoma, and ganglioglioma. All these tumours lack pseudopapillary patterns. Ganglioglioma are frequently accompanied by malformation of adjacent cortical development or cortical dysplasia [7]. There was no cortical dysplasia in our case. When compared to PGNT, pilocytic astrocytomas typically have a biphasic appearance and contain astrocytes of spindled morphology and more rounded cells, but no evidence of neural differentiation in rounded cells. The location, histological absence of glial cells, EMA immunostaining and cytokeratin immunostaining are helpful in differentiating PGNT from other papillary tumours including papillary ependymoma, papillary meningioma, choroid plexus papilloma and choroid plexus carcinoma [8,9]. In our case, immunostains for EMA, S-100 and AE1/AE3 were negative.

Most of the currently recognized PGNT have a benign course. There are, however, examples of more aggressive PGNT described in the literature [3]. Up to 11.8% of cases exhibit recurrence, and recurrence rates for complete resection and incomplete resection are 5.1% and 33.3% respectively [1].

In our case, the postoperative course was uneventful and without neurological deficit. Immediate postoperative contrast enhanced

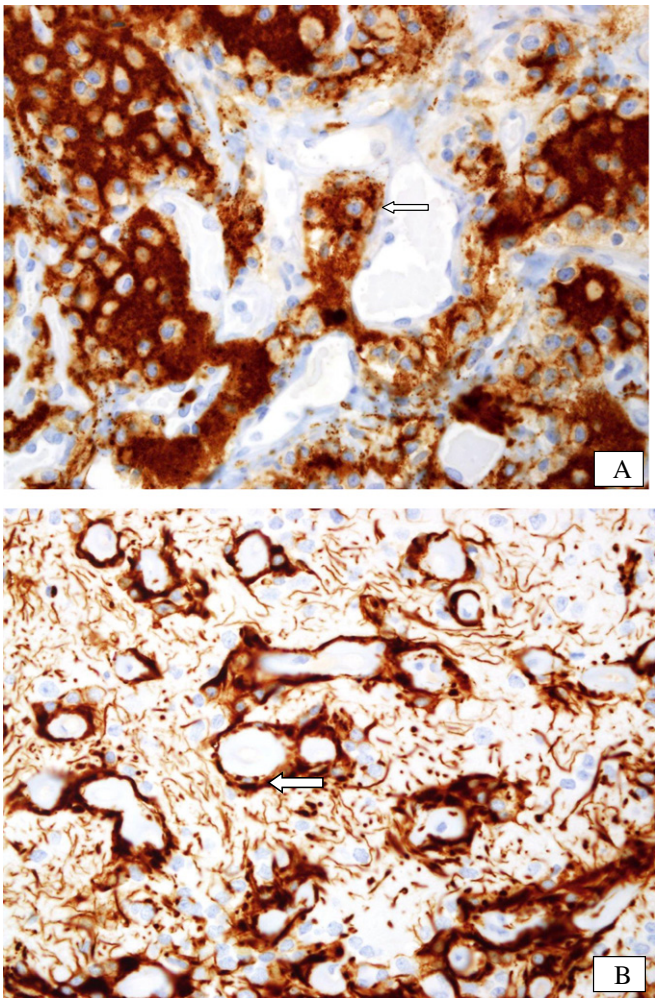

Fig. 3. (A) Cells in the interpapillary zones strongly expressed synaptophysin (arrow) in their cytoplasm, reflecting their neurocytic origin. (B) Tumour cells ensheathing the papillae showed cytoplasmic GFAP positive (arrow).

cerebral CT scans confirmed complete tumour excision but with the expected appearances of unchanged extensive vasogenic oedema. She was discharged home well four days postoperatively.

A follow up cerebral CT scan six weeks postoperatively showed complete resolution of vasogenic oedema in a well and asymptomatic patient. Subsequent outpatient follow up eight weeks postoperatively again showed a well and asymptomatic patient, neurologically normal and seizure free. Further follow up at another six months with a progress cerebral MRI scan was then arranged.

4. Conclusion

In conclusion, PGNT is a rare distinct entity with characteristic radiological and histological findings. In our case typical radiological findings were not identified but histological examination made this rare diagnosis. Further radiologic characterization of the lesion is needed to allow preoperative diagnosis and distinction from other lesions with similar radiologic features. PGNT could not be correctly diagnosed without pathology. PGNT should be included in the differential diagnosis of the biphasic tumours and further confirmed with the specific immunostaining pattern.

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