G Chir Vol. 31 - n. 8/9 - pp. 368-370
Agosto-Settembre 2010

casistica clinica

Papillary glioneuronal tumor of the fourth ventricle: case report and review of the literature

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SUMMARY: **Papillary glioneuronal tumor of the fourth ventricle: case report and review of the literature.**

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Rosette-forming glioneuronal tumour (RGNT) of the fourth ventricle is a relatively new entity recently described. Neuronal and mixed neuronal-glial tumours have been incorporated in the same category in the WHO Classification of Nervous System Tumours. This category comprises heterogenous neoplasms composed of neuronal and glial cells with various grades of differentiation.

We present a case of papillary glioneuronal tumor occurring in a 44-year-old man with an unusual hemorrhagic onset. The clinical, morphological and immunohistochemical features are discussed and the published literature is reviewed.

RIASSUNTO: **Tumore papillo-glioneuronale del quarto ventricolo: caso clinico e revisione della letteratura.**

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Il tumore glioneuronale del quarto ventricolo è un'entità antomopatologica di recente definizione e in letteratura sono riportati, ad oggi, solo 17 casi.

Gli Autori presentano la loro esperienza e pongono l'attenzione ai dati morfologici e immunoistochimici della lesione revisionando la letteratura pertinente.

KEY WORDS: Fourth ventricle - Central nervous system - Intracerebral haemorrhage - Glioneuronal tumor.
Quarto ventricolo - Sistema nervoso centrale - Emorragia cerebrale - Tumore glioneuronale.

Introduction

Papillary glioneuronal tumor is a low-grade uncommon neoplasm (1, 2) recently listed as a rare variant of ganglioglioma in the World Health Organization (WHO) Classification of Tumors of the Nervous System (3). It has been named in this way because of its papillary feature (due to the dehiscence of the cells about the vessels) and because of its double astrocytic and neuronal differentiation at either the morphological, immunohistochemical or ultrastructural level.

Papillary glioneuronal tumor was first described by Komori et al. in 1998 as a novel clinico-pathologic entity (1, 5). Nevertheless, morphologically similar tumors have been previously reported under many different names, as pseudopapillary neurocytoma with glial differentiation, atypical extraventricular neurocytic neoplasm and pseudopapillary ganglioneurocytoma (4).

Case report

A 44-year-old man was admitted to our Neurosurgical Service because of a sudden headache followed by vertigo, vomiting and imbalance of gait. Emergent computed tomography (CT) exam showed a cerebellar haemorrhage in the vermis with bleeding in the fourth ventricle without hydrocephalus. Cerebral angiography was normal while T1-T2 weight magnetic imaging supported the diagnosis of cavernous hemangioma (Fig. 1).

The patient underwent median sub-occipital craniotomy with total removal of the lesion, gray and friable, except for a portion infiltrating the lateral recess of Luschka; intraoperatively we have immediately understood that it didn't deal with a vascular lesion with

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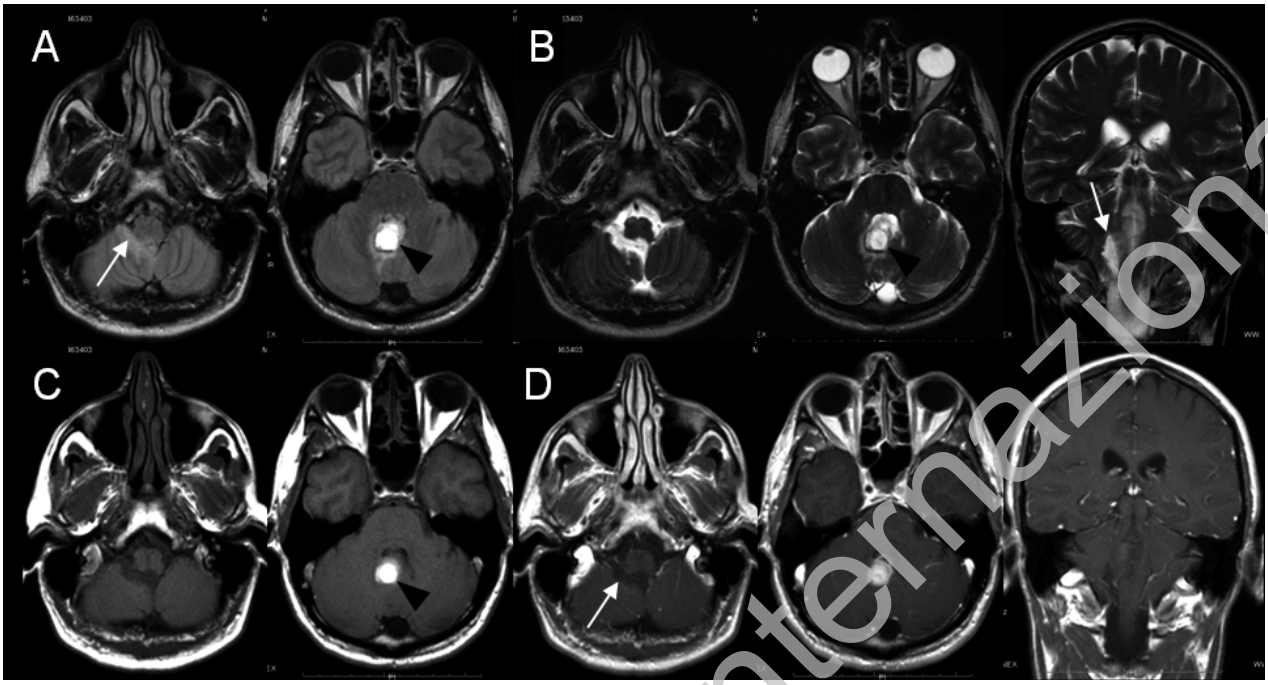

Fig. 1 - Magnetic resonance (MR) imaging of the brain.
Flair (A), T2-weighted (B), unenhanced T1-weighted (C), and gadolinium-enhanced T1-weighted (D) axial and coronal MR images showed a nodule of subacute hemorrhage (arrowheads) in the inferior right vermis, with few associated edema. The hemorrhage was within the context of an expansile and infiltrative lesion filling the right foramen of Luschka (arrows). The lesion showed low signal intensity on T1-weighted images and high signal intensity on Flair and T2-weighted images. There was no hydrocephalus.

confirmation of tumor formulated by the histology. Microscopically, haematoxylin and eosin (H&E) stained sections showed a pseudocystic gliotic cavity lined in some parts by hemosiderin-laden macrophages with a mild lymphocytic inflammatory infiltrate in the subjacent area. Some areas showed a unique complex pseudopapillary architecture that became fairly solid in multiple areas. The vascular walls were thickened, hyalinized, and lined by a single layer of cells with small round nuclei and inconspicuous nucleoli. These cells showed immunoreactivity to glial fibrillary acid protein (GFAP) and S-100 antibodies. There were few areas of mild vascular proliferation associated with piloid gliosis lining the cyst edge, and a single mitotic figure was identified in one of these areas (Fig. 2).

The post-operative course was characterised by VI cranial nerve palsy with diplopia and gait ataxia with an improvement of these troubles at three months follow-up.

Discussion

As reported for other glioneuronal tumours, RGNT probably derives from a common progenitor cell originated from sub-ependymal plate, able to differentiate toward both glial and neuronal phenotype. Rosette-forming glioneuronal tumors of the fourth ventricle contain both glial and neuronal elements (5). It remains unknown whether they arise from a single neoplastic precursor cell giving rise to glial and neuronal cell types, because of divergent differentiation, or from independent glial and neuronal cells within the same tumor. Molecular analysis of their clonal derivation will provide important evidence about the underlying tumor genesis, as it did in clonality assay of gangliogliomas (6). Interestingly, this type of mixed glioneuronal tumor arises preferentially in the fourth ventricle.

Komori et al. reported 11 cases of RGNT. On the basis of the lack of atypia, low Ki-67 labelling index, slow growth and no tendency to recur, the authors defined the RGNT as a low-grade tumour suggesting to categorise this entity as WHO grade I. Komori et al.first reported, in 1998, this entity as papillary glioneuronal tumor (PGNT). A similar histologic case was reported by Kim and Suh (7) in 1997 as pseudopapillary neurocytoma.

Papillary glioneuronal tumor is usually extraventricular with only one case having been reported in the third ventricle and is composed of glial and neuronal histologic components. The glial component is astrocytic, characterized by pseudopapillary formation and featuring conspicuous hyalinized vasculature. Only in one case were hyalinized vessels not identified (8). The vessels are enclosed by a uniform, single, or pseudostratified layer of small cuboidal cells with round reticular nuclei without atypia, and scant cytoplasm. These cells are uniformly immunoreactive for GFAP and S-100 antibodies, and one case showed immunostaining with synaptophysin antibody (9). The interpapillary spaces contain small

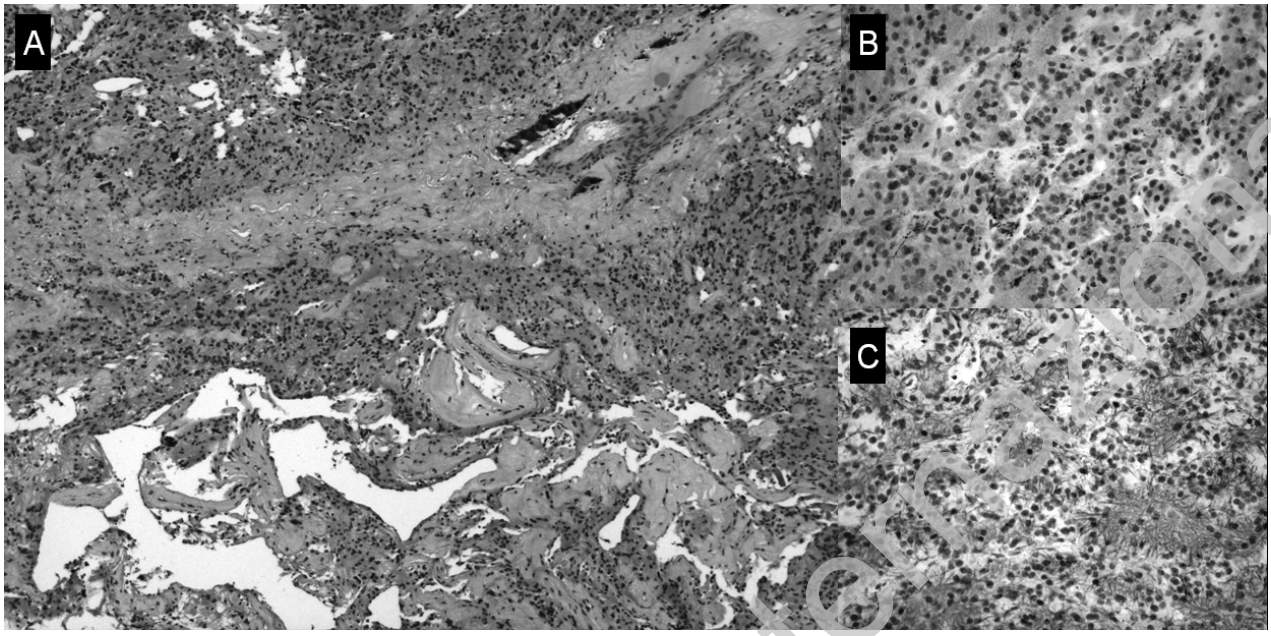

Fig. 2 - Pathologic specimens. Vascular, fibrotic pseudopapillae, solid tumour areas, and large vessels inside fibrotic septae are evident at haematoxylin and eosin, original magnification, x50 (A). Synaptophysin positive (B), and GFAP positive (C) tumour cells, the latter in direct contact with vessels, are evident at immunohistochemistry, diaminobenzydine, original magnification x200.

round neuronal cells with perinuclear halos, resembling oligodendrocytes that have been shown to stain for anti-Olig (10) and synaptophysin antibodies. Other pertinent findings seen in most of the cases include the presence of Rosenthal fibers, foci of dystrophic calcification, and hemosiderin deposition in the surrounding reactive tissue. Proliferative indices as determined by immunostaining with Ki-67 have uniformly been shown to be low.

The main peculiarity of our case is the onset with cerebellar haemorrhage. The frequency of intracranial haemorrhage in patients with intracranial neoplasms varies in the different series from 2% to 5%. Several mechanisms were advocated to explain the pathogenesis of brain tumor related haemorrhages: endothelial proliferation with subsequent obliteration of the lumen; thin walled or poorly formed vessels; perivascular necrosis with subsequent loss of vessel support; presence of intratumoral arteriovenous fistulae (11). The surgical treatment of RGNT is the best solution even not exempt of risk with high probability of permanent and severe neurological deficits.

References

1. Komori T, Scheithauer BW, Hirose T. A rosette-forming glioneuronal tumour of the fourth ventricle: infratentorial form of dysembryoplastic neuroepithelial tumour? Am J Surg Pathol 2002;26: 582–591.
2. Preusser M, Dietrich W, Czech T, Prayer D, Budka H, Hainfellner JA. Rosette-forming glioneuronal tumor of the fourth ventricle. Acta Neuropathol (Berl) 2003;106: 506–508.
3. Kleihues P, Cavenee W. Pathology and Genetics: Tumors of the Nervous System. Lyon, France: IARC Press, Chapter 6, pp 96–114,2000.
4. Shibuya M, Yamazaki H, Sugiyama M. A case of papillary glioneuronal tumor.Brain Tumor Pathol 20 (Suppl): 2003;73.
5. Komori T, Scheithauer BW, Sung J . Rosette-forming mixed neuronalglial tumor in the fourth ventricle. J Neuropathol Exp Neurol 1998 ;57: 520.
6. Zhu JJ, Leon SP, Folkerth RD, et al. Evidence for clonal origin o neoplastic neuronal and glial cells in gangliogliomas. Am J Pathol 1997;151:565 – 71.
7. Kim DH, Suh YL. Pseudopapillary neurocytoma with glial differentiation. Acta Neuropathol (Berl) 1997;94: 167–191.
8. Lamszus K, Makrigeorgi-Butera M, Laas R, Westphal M, Stavrou D. 24-year-old female with a 6-month history of seizures. Brain Pathol 2003;13:115-7.
9. Barnes NP, Pollock JR, Harding B, Hayward RD. Papillary glioneuronal tumor in a 4-year-old. Pediatr Neurosurg 2002;36: 266- 70.
10. McLendon RE, Provenzale J. Glioneuronal tumors of the central nervous system. Brain Tumor Pathol 2002;19:51-8.
11. Lieu AS, Hwang SL, Howng SL, Chai CY. Brain tumors with hemorrhage. J Formos Med Assoc 1999; 98: 365–367.