Rosette-forming Glioneuronal Tumors of Infratentorial Region: About 3 Cases with Similar Clinical Presentation and Radiological Findings

Anne Laure Bernat; Antoine Guillonnet; Emmanuel Mandonnet; Homa Adle-Biassette; Sébastien Froelich

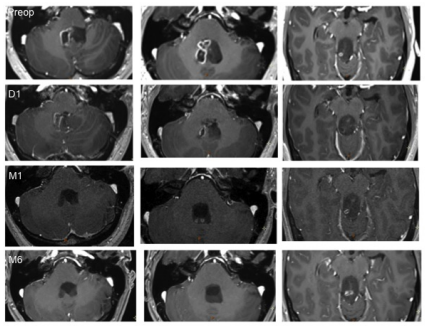
[Lariboisière Hospital, Paris Diderot University

Assistance Publique-Hôpitaux de Paris]

Introduction

RGNT is a rare tumor of the central nervous system, typically arising in the III and IV ventricles. First Described as dysembryoplastic neuroepithelial tumor (DNT) of the cerebellum by Kuchelmeister et al. in 1995 , Komori et al. defined RGNT as a specific disease in 2002. Mixed glial-neuronal tumors of the posterior fossa still rare and poorly described in cases reports only. No consensus exists about its management.

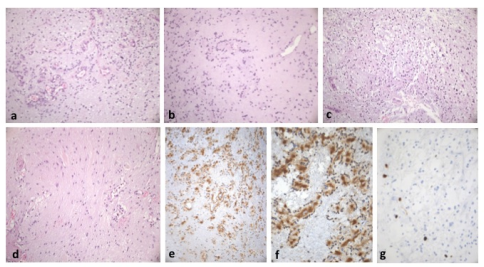
Case A



Clinical presentation

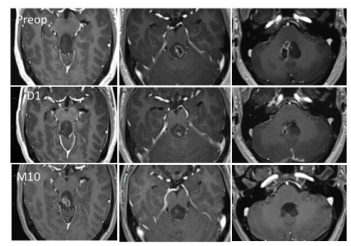
We report 3 cases of young women (mean age of 29 yo) admitted in emergency over the same year. MRI showed infratentorial lesions with hydrocephalus: two of them (**A&B)** located in the fourth ventricle and the last one (**C)** was in the cerebellum. All the tumors had both solid and cystic components showing focal and irregular ring shaped enhancement postcontrast imaging. Treatment of these patients has consisted of subtotal (**C)** or partial resection **(A&B)** without adjuvant therapy. Histology revealed RGNT in the three cases **(Fig.1)**. Except age and sex, any other common factor was found. The mean follow-up was of 13 months. Volume of residual tumor still be stable on the radiological follow up so far in the three cases. We observed however new contrast enhancing points appearing and disappearing alternately in the two cases with partial resection (**A&B)**. The residual enhancing in the case **C** disappeared at 3 months and so far.

Fig.1: Cytologie

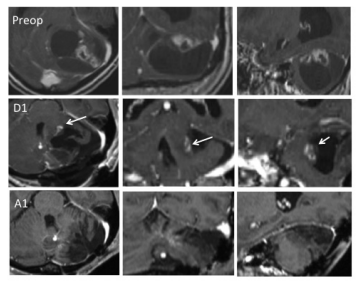


a-b : cellules tumorales formant des pseudorosettes périvasculaires (a) et des rosettes neurocytiques (b).
c-d : Il existait en outre des secteurs microkystiques renfermant des cellules d'aspect oligodendroglial en nid d'abeille (c) et un contingent pilocytique (d).
e,f : les rosettes étaient marquées avec l'anticorps antisynaptophysine
g : immunomarquage anti-Ki67 comportant ici 6% de cellules en cycle.

Case B



Case C



Conclusion

We should think about this rare disease in front of theses typical image findings. Given the behavior of this tumor is still unknown, we recommend a total or subtotal resection as much as possible. Although we observed atypical time-varying intratumoral enhancement on postoperative MRIs without volume change, we decided to follow our patients.

References

1. Komori T, Scheithauer BW, Hirose T. A rosette-forming glioneuronal tumor of the fourth ventricle: infratentorial form of dysembryoplastic neuroepithelial tumor? Am J Surg Pathol. 2002;26(5):582–91.

2. Schlamann A, von Bueren AO, Hagel C, Zwiener I, Seidel C, Kortmann R-D, et al. An Individual Patient Data Meta-Analysis on Characteristics and Outcome of Patients with Papillary Glioneuronal Tumor, Rosette Glioneuronal Tumor with Neuropil-Like Islands and Rosette Forming Glioneuronal Tumor of the Fourth Ventricle. Hills RK, editor. PLoS ONE. 2014 Jul 3;9(7):e101211.

3. Marhold F, Preusser M, Dietrich W, Prayer D, Czech T. Clinicoradiological features of rosette-forming glioneuronal tumor (RGNT) of the fourth ventricle: report of four cases and literature review. J Neurooncol. 2008 Dec;90(3):301–8.

4. Jacques TS, Eldridge C, Patel A, Saleem NM, Powell M, Kitchen ND, et al. Mixed glioneuronal tumour of the fourth ventricle with prominent rosette formation. Neuropathol Appl Neurobiol. 2006 Apr;32(2):217–20.

5. Thurston B, Gunny R, Anderson G, Paine S, Thompson D, Jacques T, et al. Fourth ventricle rosette-forming glioneuronal tumour in children: an unusual presentation in an 8-year-old patient, discussion and review of the literature. Childs Nerv Syst. 2013 May;29(5):839–47.

6. Preusser M, Dietrich W, Czech T, Prayer D, Budka H, Hainfellner JA. Rosette-forming glioneuronal tumor of the fourth ventricle. Acta Neuropathol (Berl). 2003 Nov 1;106(5):506–8.

7. Zhang J, Babu R, McLendon RE, Friedman AH, Adamson C. A comprehensive analysis of 41 patients with rosette-forming glioneuronal tumors of the fourth ventricle. J Clin Neurosci. 2013 Mar;20(3):335–41.