

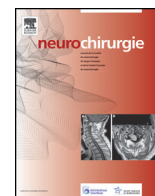


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Clinical case

Thalamic rosette-forming glioneuronal tumor in an elderly patient: Case report and literature review



Tumeur glioneuronale à rosette du thalamus chez une femme âgée : cas clinique et revue de la littérature

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ABSTRACT

The rosette-forming glioneuronal tumor (RGNT) is a novel type of brain tumor recently listed in the WHO 2007 classification of central nervous system (CNS) tumors. We report the case of a 75-year-old woman harboring a thalamic RGNT with third ventricle dissemination. Age and location make the present case exceptional and which has never previously been reported. A review of the clinical, pathological and radiological features is presented along with the relevant literature.

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R É S U M É

Les tumeurs glioneuronales à rosette sont une nouvelle entité de tumeurs cérébrales décrites dans la classification WHO en 2007. Nous rapportons le cas d'une femme âgée de 75 ans présentant une tumeur glioneuronale à rosette du thalamus avec dissémination ventriculaire. L'âge et la localisation font de ce cas un cas rare dans la littérature. Nous présentons une revue de la littérature avec analyse des particularités cliniques, radiologiques et pathologiques.

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1. Introduction

The rosette-forming glioneuronal tumor (RGNT) is characterized by its biphasic architecture with uniform neurocytes forming rosettes and/or perivascular pseudorosettes and astrocytic tumor cells, is a new tumor entity described since 2007 in the WHO CNS tumor classification. Initially classified as a dysembryoplastic neuroepithelial tumor (DNET) described in the fourth ventricle, the RGNT is considered as a rare, slow-growing tumor affecting young adults [1]. RGNT has previously been reported in various brain

locations such as fourth ventricle, cerebellum, brainstem or pineal gland [2,3–10], although its location into the thalamus with dissemination into the third ventricle, to the best of our knowledge, has never been reported before. We report the case of a left pulvinar RGNT that occurred in a 75-year-old woman with 3rd ventricle dissemination and intratumoral hemorrhage diagnosed and managed by a straightforward endoscopic approach as well as discuss the current therapeutic strategy.

2. Case report

A 75-year-old woman presented at our outpatient clinic with a 2-month history of progressive unsteady gait, headache,

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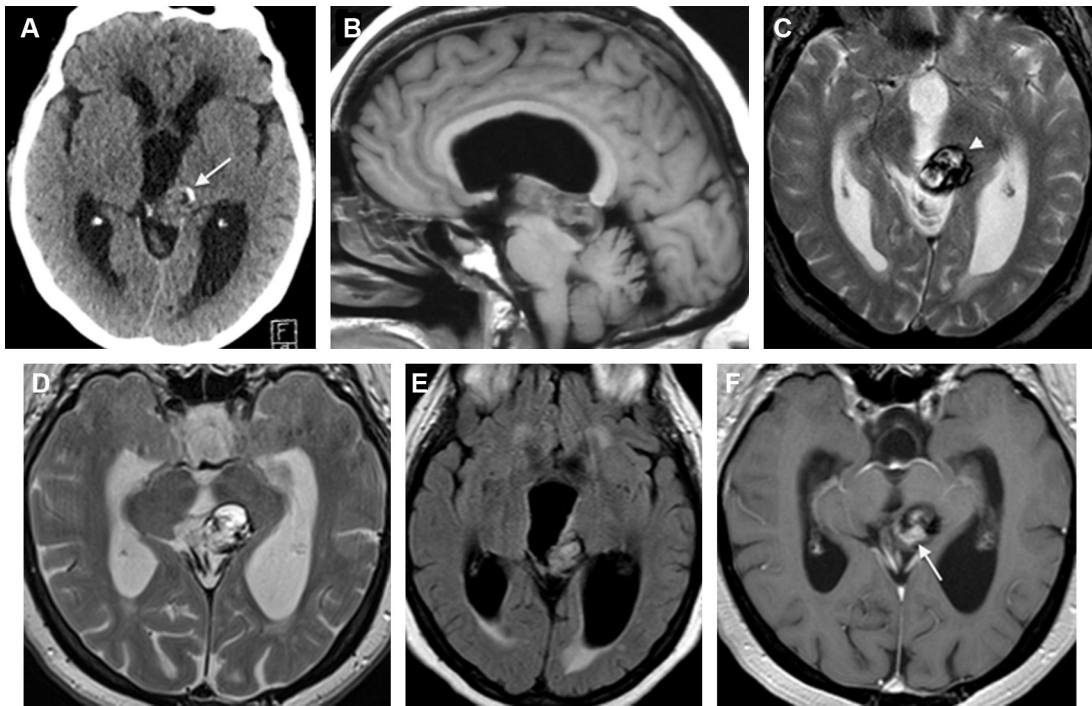


Fig. 1. A. Axial CT scan showed a deformation of the posterior part of the third ventricle with intralesional hemorrhage (arrow head). B. MRI sagittal T1: left posterior thalamic exophytic lesion (into the third ventricle) with quadrigeminal mass effect. C. Axial gradient echo T2 MRI: the lesion was multilocular with calcifications (arrow). D. Axial T2 MRI: left posterior thalamic exophytic lesion with supratentorial ventricular dilatation. E. Axial flair MRI: the hypersignal along the wall of the 3rd ventricle did suspect the intraventricular dissemination. F. The lesion presented heterogeneous contrast enhancement (arrow head) after gadolinium injection in the axial T1 enhanced.

A. Scanner en coupe axiale montrant une déformation de la paroi postérieure du troisième ventricule avec la présence d'une hémorragie intratumorale (flèche). B. IRM sagittale T1 : lésion de la partie postérieure du thalamus exophytique dans le troisième ventricule. C. IRM en coupe axiale gradient écho T2 montrant la lésion du thalamus avec des calcifications (flèche). D. IRM axiale T2 : lésion exophytique de la partie postérieure du thalamus avec dilatation ventriculaire supratentorielle. E. IMR en axial flair : l'hypersignal le long de la paroi du troisième ventricule fait suspecter une dissémination intraventriculaire. F. La lésion présente une prise de contraste hétérogène (flèche) après l'injection de produit de contraste en coupe axiale T1.

drowsiness and urinary incontinence. No history of neoplasm was observed.

Head CT scan and magnetic resonance imaging (MRI) revealed a left posterior thalamic lesion compressing the quadrigeminal plate responsible for tri-ventricular hydrocephalus (Fig. 1A–F). Intralesional bleeding (Fig. 1A), calcifications (Fig. 1C) and heterogeneous enhancement were also detected (Fig. 1F).

Due to the age and the suspicion of tumor subependymal dissemination (Fig. 1E), we planned an endoscopic approach with the double objective to treat the obstructive hydrocephalus and perform a biopsy of the lesion. Under general anesthesia, the patient was in supine position with the head stabilized with Mayfield Kerr headrest. A single frontal right-sided burr hole was performed, according to the neuronavigation system (Vector Vision BrainLab®). Following placement of the burr hole, dural coagulation, and pial piercing, the endoscope (Hopkins II optical system, Endoscopy-America, Charlton, MA. and operative channel by Karl Storz®) was advanced toward the lateral ventricle, and CSF was taken for cytological and biochemical analyses. After entering the foramen of Monro, the endoscope direction was readjusted, following the ventricular landmarks as well as the indications of the neuronavigational system, aimed toward the tumor. The tumor was then identified (Fig. 2), and after visual inspection of the lesion, careful coagulation of its surface was performed, followed by biopsy. A total of eight samples were taken for histopathology investigation including frozen section. A third ventricular dissemination was observed (Fig. 2). Hemostasis was achieved by a gentle Ringer irrigation. Following tumor biopsy, a standard endoscopic third ventriculostomy was completed via a second standard right frontal coronal burr hole.

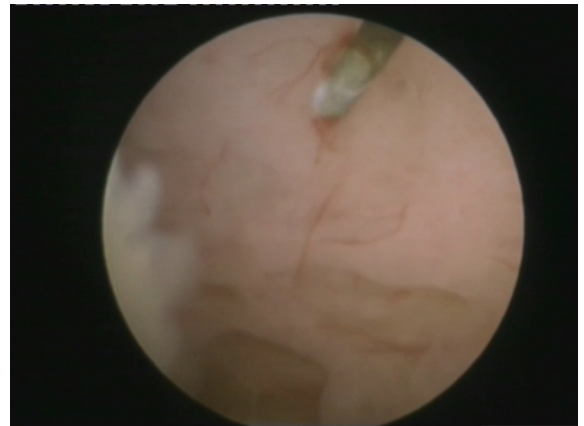


Fig. 2. Endoscopic observation with the visualization of the exophytic lesion and the third ventricle dissemination.

Vue endoscopique avec visualisation de la lésion exophytique et d'une dissémination tumorale dans le troisième ventricule.

The endoscopic biopsy option was chosen with regard to the patient's age as it was considered to be a quite safe and minimally invasive procedure. In case of negative biopsy specimen, the option of open surgery could have been discussed and proposed to the patient.

Histological examination demonstrated a RGNT constituted by a neurocytic and glial architecture (Fig. 3A and B).

The case was discussed during our interdisciplinary neuro-oncology meeting, consensus for watchful observation without

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