Clinical case

Thalamic rosette-forming a 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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A B S T R A C T

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,
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). Intraleisonal 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.

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 neurooncology meeting, consensus for watchful observation without

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.


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 tumoraire dans le troisième ventricule.
additional treatment was recommended. At 6 months and 1 year follow-up, the patient had no new complaints and no tumor progression was observed on serial postoperative MRIs.

3. Discussion

Our case was characterized by a thalamic localization with tumor dissemination into the third ventricle occurring in a 75-year-old female. The morbidity risk of the microsurgical resection and the age of the patient did justify the treatment of the hydrocephalus and performing a biopsy without adjuvant therapy.

3.1. Pathology and classification

A cerebellar RGNT was first described in 1995 as DNET with rosette-like feature. It was recognized as a separate clinical-pathological entity with unique morphology by Komori et al. in 2002 (11 cases) [1]. The original description was that of a tumor exclusively invading the fourth ventricle and its adjacent region.

The WHO included this rare tumor in 2007 in the category of mixed glial-neuronal tumors and listed it as grade I stating that it also contains both a neurocytic and astrocytic components [1]. These tumors are thought to originate from the progenitor pluripotential cells of the subependymal plate as they display both glial and neurocytic features [12]. After the first description in the fourth ventricle, other locations have been described such as cerebellum, brainstem, third ventricle and pineal region [2–10].

These tumors display distinctive morphology as they are constituted by two histopathological components: uniform neurocytes, with small, round cells forming rosettes and/or perivascular pseudorosettes and astrocytic tumor cells, with spindle cells resembling pilocytic astrocytomas [13]. The majority of RGNT do not reveal histological features of anaplasia. Cellular atypia, mitotic activity and necrosis are usually absent. The proliferation index assessed by MIB-1 labeling antigen and Ki67 antigen were relatively low, usually less than 1–2.2%.

These tumors correspond histologically to a WHO grade I neoplasm like the pilocytic astrocytoma. It is characterized by relatively favorable prognosis.

Long-term follow-up studies are needed to better assess the biological behavior of RGNT and to adapt the therapeutic strategies.

3.2. Clinical description

Generally, these neoplasms affect young adults (in their third or fourth decade) [14] with female predominance (predilection of 2:1). The mean age for the cerebellar localization is 31.5 years.

However, it is interesting to point out that, like in our case, two previous cases have been described, in a 70- and 79-year-old patient, respectively in the third and fourth ventricles [3]. Only these two cases (one in the autopsy finding) were described in elderly patients. These observations confirm that RGNT are benign tumors with a slow-growing behavior.

The majority of cases are located in the fourth ventricle, aqueduct, brain stem, cerebellar vermis or pineal body. RGNT may occur in different locations outside the fourth ventricle such as the spinal cord, the cerebellopontine angle, the optic chiasm, the pineal and tectal regions (Table 1). Local recurrences have also been reported [14,15]. One case of intraventricular dissemination was described in a 16-year-old woman [15].

MRI appearance of RGNTs is consistent with a solid, mixed cystic and solid or cystic tumors. These tumors are generally well circumscribed. The solid areas are iso/hypointense on T1-weighted images and hyperintense on T2-weighted images. A minimal peritumoral edema is usually detected. In most cases, contrast enhancement is seen. Calcification is occasionally detected on CT and satellite lesions could be detected. Li et al. described a case with intratumoral hemorrhage [16]. The differential radiological diagnoses include pilocytic astrocytoma, cystic hemangioblastoma, medulloblastoma, ependymoma, choroidal papilloma [10].

Our case was characterized by an intratumoral hemorrhage with suspicion of third ventricle dissemination. This diagnosis should be considered in the elderly as a differential diagnosis.

3.3. Prognosis and therapeutic propositions

According to the recent literature, it appears that partial tumor resection is beneficial and compatible with prolonged progression-free-survival because they are considered as low-grade tumors [17]. However, if a patient is symptomatic due to mass effect, then aggressive resection may be necessary. It is intuitive that the risk of postoperative morbidity increases with amount of resection owing to intimate proximity of key structures such as the cerebellum or brain stem. In these cases, a less aggressive attitude is recommended.

In contrast, Ellezam et al. [14] described two cases of late recurrence despite gross total resection. Cabezás et al. used chemotherapy (temozolomide) and radiotherapy in an aggressive form of RGNT [6] and Zhang et al. described three patients who received radiotherapy following radiological progression [18]. There are very few references in the literature of high-grade RGNT tumors.

Generally, no adjuvant treatment is performed after surgery. Most of cases described in literature have good prognosis despite frequent subtotal resection.
Furthermore, Zhang et al. could not show any difference in survival when comparing patients with gross total resection versus subtotal resection [18].

Recently, Schlamann et al. reported in an excellent meta-analysis all specific characteristics and outcome of patients harboring RGNT [19]. In the former series, these authors suggest that surgery still remains the first-line therapy for such a tumor. On the other hand, Zhan et al. [18] reported, as in the present case, that selected case of RGNT could be managed conservatively, considering, the slow-growing behavior of such a tumor. Clinical outcomes of patients harboring RGNTs need to be documented by future long-term follow-up studies, in order to estimate the long-term prognosis, factors and recurrence patterns and confirm the presumed indolent course.

In our case, complete resection was impossible due to infiltration/dissemination of third ventricle and the follow-up confirms that there is no tumor progression at 1 year suggesting that RGNT is a certainly benign tumor with benign evolution.

We need more cases and a long-term evolution to confirm our impression but we think that in rare case on RGNT in elderly, biopsy without adjuvant treatment is sufficient.

4. Conclusion

The RGNT are considered like a low-grade tumor. They occur mostly in young adults although they rarely concern the elderly patient. Adjuvant treatment is not required due to the low-grade histological features. In elderly patient, the biopsy without resection could be proposed as an effective option. Further studies and long-term follow-up are advisable to identify strategies for optimal management.

Disclosure of interest

The authors declare that they have no competing interest.

References


