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Case Report

A case of a rosette-forming glioneuronal tumor arising from the pons with disappearance of contrast enhancement

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ABSTRACT

A rosette-forming glioneuronal tumor (RGNT) is a rare and slow-growing central nervous system tumor. This tumor is usually assessed by MRI during the follow-up period. RGNT can show alteration of contrast enhancement regardless of tumor growth. Here, we report a case of RGNT arising from pons which shows partial enhancement on initial MRI, smaller enhancement on follow-up MRI at 10 months, and totally disappeared at 18 months without any therapy.

Introduction

The rosette-forming glioneuronal tumor (RGNT) was first described by Komori et al. [1]. RGNT is characterized by both neurocytic and/or perivascular rosettes and astrocytic components [1,2]. This tumor mostly affects young people, with a male to female ratio of 1:1.75 [1]. RGNT of the fourth ventricle was also included in the World Health Organization Classification of Central Nervous System Tumors in 2007 with a grade of I [3]. RGNT is rarely found in the pons, thalamus, spinal cord, optic chiasm, or cerebellar vermis [4]. Past reports of RGNT have featured favorable outcomes [4]. In this report, we present a case of RGNT of the pons, in which spontaneous disappearance of contrast enhancement occurred on follow-up MR examinations and remained thereafter.

Case report

An 18-year-old female presented with headache and vomiting. Initial MRI revealed a mass (10 mm × 12 mm) in the pons which showed low signal intensity on T1-weighted image

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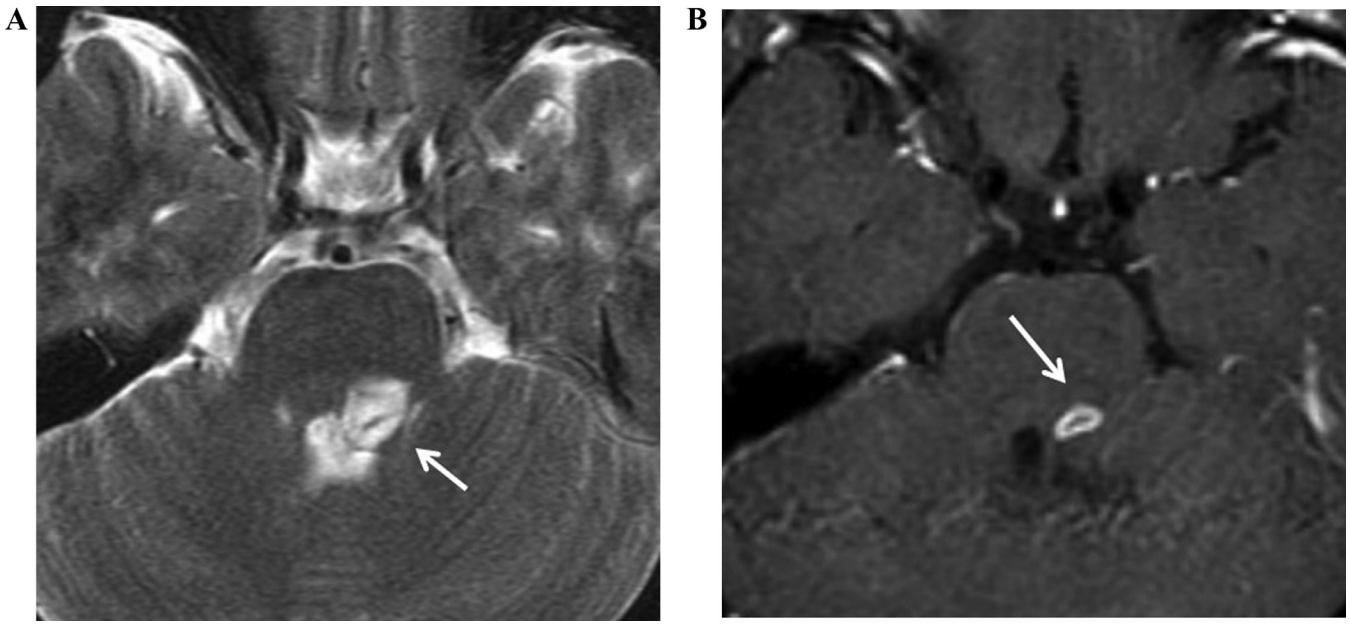

Fig. 1 – (A) T2-weighted image on the first visit to the hospital reveals a hyperintense mass in the pons (arrow). (B) The mass shows partial rim enhancement on postcontrast T1-weighted image (arrow).

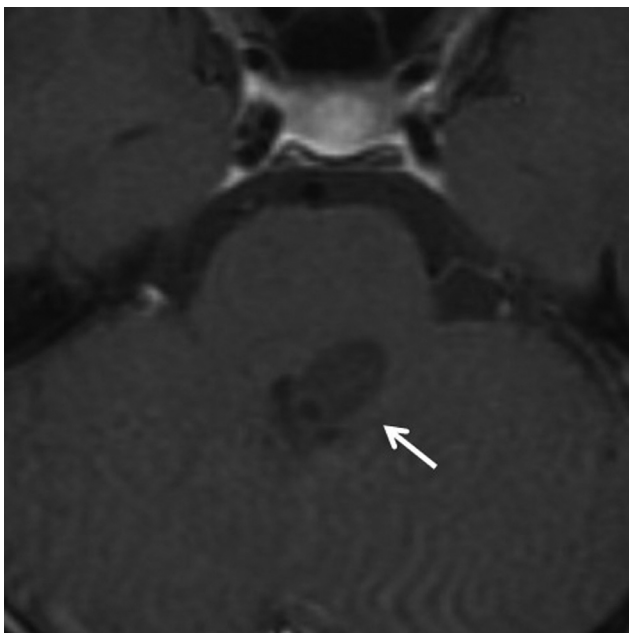

Fig. 2 – Eighteen months later, postcontrast T1-weighted image shows spontaneous disappearance of contrast enhancement (arrow).

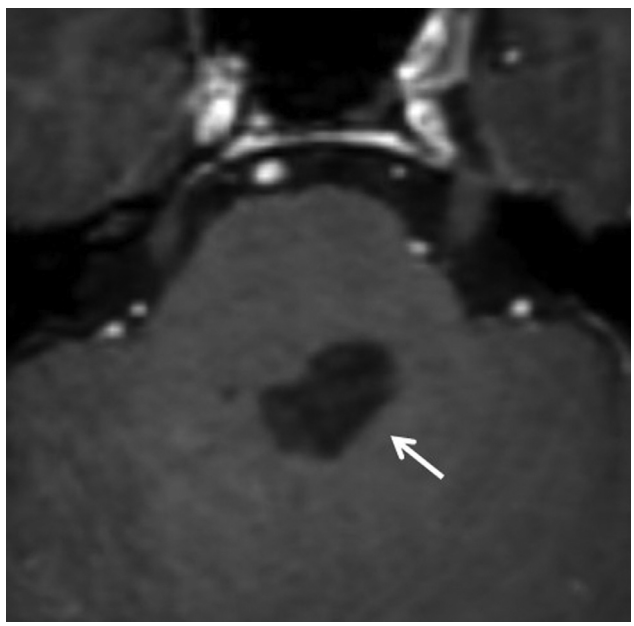

Fig. 3 – Seventeen years after the first MRI, the tumor exhibits no contrast enhancement (arrow).

(T1WI), high signal intensity on T2-weighted image (T2WI) (Fig. 1A) and fluid-attenuated inversion recovery, and partial enhancement of the mass on postcontrast T1WI (Fig. 1B). There was no sign of hydrocephalus. Contrast enhancement was smaller on follow-up MRI at 10 months (19 years old) and totally disappeared at 18 months (20 years old) without tumor growth (Fig. 2). Notably, her symptoms were relieved without any therapeutic interventions. Seventeen years after her first MRI, when she was 35 years old, we observed slight tumor growth, but the signal intensities of the tumor on T1WI and T2WI didn't change. There was no bleed on susceptibility-weighted imaging. Increased diffusion was observed on diffusion-weighted imaging and apparent diffusion coefficient mapping. No contrast enhancement remained (Fig. 3). MR spectroscopy showed that the N-acetyl- L-aspartate /

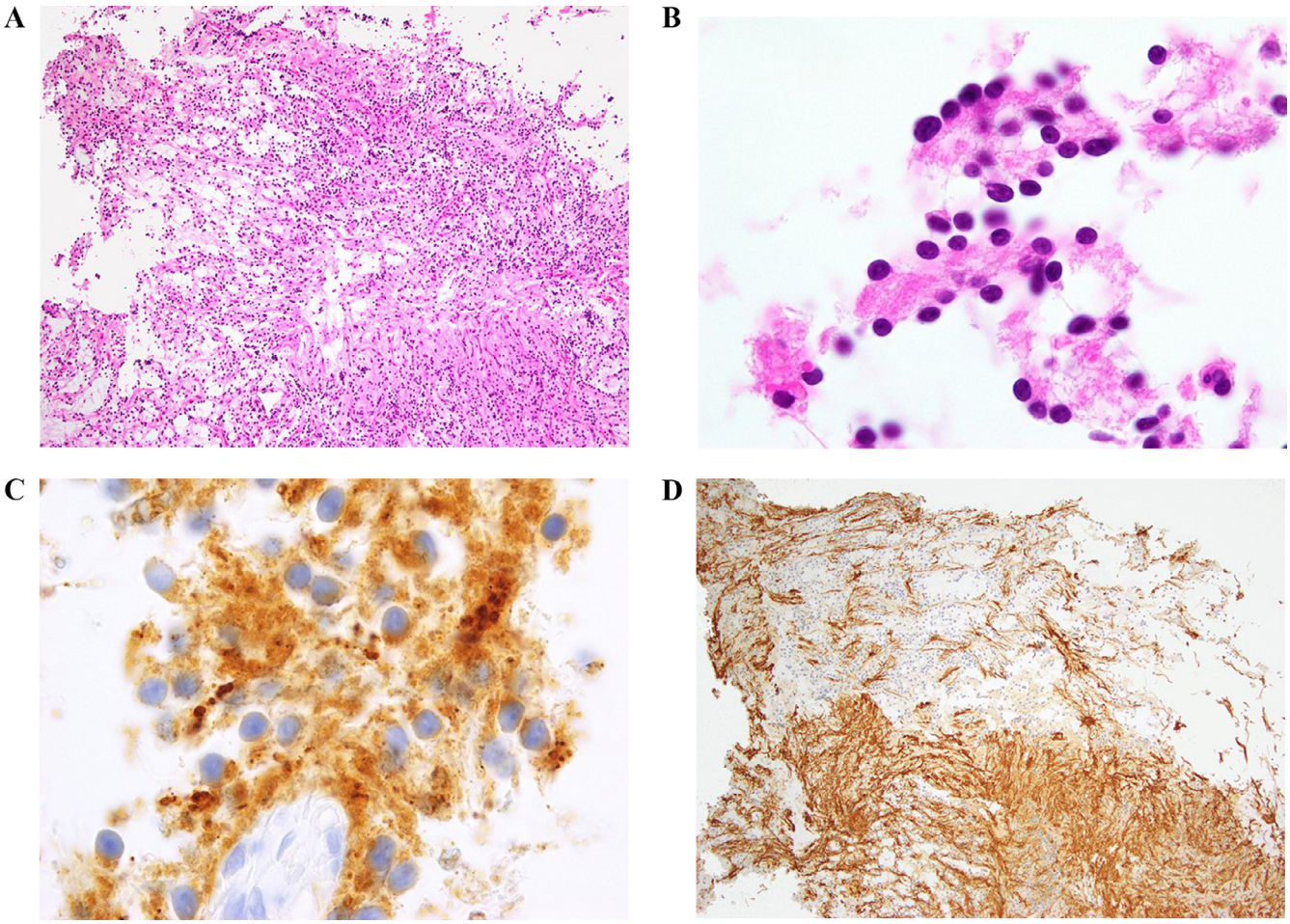

Fig. 4 – Histopathological findings. (A) Hematoxylin and eosin (H-E) staining shows that the tumor consists of a neurocytic component (upper half) and an astrocytic component (bottom half). Original magnification ×100. (B) H-E staining also shows a neurocytic rosette: the neurocytic tumor cells are arranged around a neuropil core. Original magnification ×400. (C) The neurocytic tumor cells and neuropil core are positive for synaptophysin. Original magnification ×400. (D) The astrocytic component is positive for glial fibrillary acidic protein. It is the same field as (A). Original magnification ×100.

chorine ratio was 1.62. No lipid or lactate peaks were present, indicating benign biological behaviors. ^{18}F-FDG PET/CT showed lower uptake than the brain parenchyma. There was no calcification on CT. Postcontrast CT was not used for follow-up to avoid radiation exposure because she was a young female and pregnant at 29 years old. When she was 35 years old, she underwent a tumor biopsy in the fourth ventricle via transcerebellomedullary fissure without splitting the vermis by midline suboccipital craniotomy. Histopathologic examination revealed 2 components of this tumor; the neurocytic and astrocytic components (Fig. 4A). The neurocytic component consisted of neural rosettes and perivascular pseudorosettes with neuropil core positivity for synaptophysin (Fig. 4B and C). The neurocytic tumor cells had slightly hyperchromatic small nuclei with inconspicuous nucleoli and minimal cytoplasm. The astrocytic component tumor cells had oval nuclei with slightly dense chromatin and fine cytoplasmic processes. The astrocytic tumor cells were positive for oligodendrocyte transcription factor 2 and glial fibrillary acidic protein (Fig. 4D). Necrosis, microvascular proliferation, and mitotic figures were absent. The MIB-1 labeling index was lower than 1%. Thus, we diagnosed her with RGNT. She underwent no additional resection, chemotherapy, or radiation therapy.

Discussion

RGNT can manifest as solid, cystic-solid, or multicystic tumors. On CT, the solid region of the RGNT appears hypodense and the cystic region shows lower density. On MRI, the solid part of the RGNT appears hypointense on T1WI and hyperintense on T2WI. Calcification is seen occasionally [4,5]. On diffusion-weighted imaging, RGNT shows no evidence of restricted diffusion unless intratumoral hemorrhage occurs [6]. Yang et al. reported on contrast enhancement patterns in 38 patients with RGNT [7]. A quarter of RGNTs demonstrated no enhancement, and the others showed heterogeneous (44.7%), rim (23.7%), or focal (7.9%) enhancements, which were associated with the cystic/solid nature of the

tumors [7]. RGNT sometimes shows ring enhancement with a central hypointensity, surrounded by thin or no enhancement, resembling a sectioned green bell pepper [5]. Haryu et al. reported that the RGNT of the tectum showed spontaneous disappearance of contrast enhancement [8]. However, there has been no report on RGNT arising from the pons with disappearance of contrast enhancement. On the other hand, Fujiwara et al. reported that contrast enhancement of the RGNT in the vermis was not found initially but emerged by the 6-month follow-up examination [9]. Histopathologically, RGNT consists of 2 components: a neurocytic component that forms rosettes and/or perivascular pseudorosettes, and an astrocytic component that resembles a pilocytic astrocytoma. Gaudino et al. reported serial changes in contrast enhancement on follow-up MRI in 30.8% patients with pilocytic astrocytoma [10]. Contrast enhancement fluctuations were not related to progression of pilocytic astrocytoma [10]. Degenerative and inflammatory intratumoral changes affect blood-brain barrier permeability, resulting in contrast enhancement variability [10]. Moreover, it is also reported that dysembryoplastic neuroepithelial tumors shows enhancing part in the tumor becoming nonenhancing during follow-up and vice versa [11]. According to another case report of dysembryoplastic neuroepithelial tumors which shows focal enhancement in the lesion for the first time 12 years after the initial examination, histopathologic study demonstrates pilocytic astroglial tissue in this tumor [12]. Therefore, the astrocytic component of RGNT and pilocytic astroglial tissue of dysembryoplastic neuroepithelial tumors may be strongly associated with contrast enhancement fluctuations. This knowledge may help us better understand RGNT or other brain tumors with pilocytic astrocytoma component.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2019.05.011.

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