Rosette-forming glioneuronal tumor of the fourth ventricle

Tumeur glioneuronale à rosettes du quatrième ventricule

A 21-year-old woman presented at the hospital with a one-week history of severe headache, vertigo and vomiting. Cranial computed tomography (CT) scans revealed a fourth ventricle tumor with a "cyst and mural nodule" configuration. Magnetic resonance imaging (MRI) of the brain showed a mixed cystic–solid mass occupying the fourth ventricle that measured 3.0 cm in diameter. The mass lesion was hypointense on T1-weighted images, and hyperintense on fluid-attenuated inversion recovery (FLAIR) images. Patchy contrast enhancement was observed on post-contrast T1-weighted images (Fig. 1A–F).

On histopathology, the lesion consisted of two distinct cell lines. The first component showed uniform neurocytes forming neurocytic rosettes—neurocytic tumor cells featuring small round nuclei (Fig. 2). The second tumor component resembled, in most areas, pilocytic astrocytoma with spindle/stellate tumor cells and elongated-oval-shaped tumor cell nuclei. Rosenthal fibers were also observed. These findings pointed to a rosette-forming glioneuronal tumor (RGNT) of the fourth ventricle.

RGNT is a rare neoplasm. The first series of 11 cases was reported by Komori et al. [1], and the entity was accepted as a World Health Organization (WHO) grade I tumor in the 2007 revision of the WHO classification of central nervous system tumors [2]. RGNT has a slight female predominance, and most patients are in their third or fourth decade of life. Nearly all the recently described RGNT were found in the posterior fossa, typically in a midline location, in the region of the fourth ventricle and aqueduct of Sylvius; they may directly extend to involve the brain stem, cerebellum vermis, pineal region, cerebellopontine angle and/or thalamus [3]. One unusual case was reported in the optic chiasma [4].

In previously published reports, the structures of the tumor on MRI were described as solid, mixed cystic and solid, or cystic. Also, the tumors had relatively well-circumscribed margins. After administration of contrast medium, contrast enhancement is seen in most cases, with the enhanced lesion showing focal or patchy contrast enhancement. Calcification is also occasionally detected. Satellite lesions (secondary distant lesions) have been detected in a few published cases [1, 3], but no satellite lesions were found in our present case.

Figure 1 A. Sagittal T1-weighted image (T1WI) shows a mass occupying the fourth ventricle, causing its expansion, and the aqueduct of Sylvius. B. Axial T1WI reveals a mixed cystic–solid mass, with hypo-intense cystic components and iso-intense solid areas. C. Axial fluid-attenuated inversion recovery (FLAIR) shows a mixed cystic and solid mass with a hyperintense solid component. D, E. The tumor presented with patchy contrast enhancement on post-contrast images, and (F) there was no evidence of restricted diffusion on diffusion-weighted images.
Meningeal leukemia in acute lymphoblastic leukemia revealed by an intracranial mass

Leucémie méningée au cours d’une leucémie lymphoblastique aiguë révélée par une masse intracrânienne

Case report

We report here on the case of a 13-year-old girl, with a history of two relapses of precursor B-cell acute lymphoblastic leukemia (B-ALL) post-stem-cell transplantation, who presented with paresthesia and uncontrolled movements in the right leg. Cerebrospinal fluid cytology was negative for leukemic cells. Magnetic resonance imaging (MRI) of the brain revealed a large parafalcine mass, with a dural tail, and leptomeningeal enhancement of the adjacent left frontal sulci. The mass showed isosignal intensity to grey matter on both T1- and T2-weighted images, diffusion restriction and homogeneous enhancement after contrast administration (Figs. 1–3). The mass was found underlying the dura during surgical biopsy. Histological examination revealed diffuse proliferation of lymphoblasts. Immunohistochemical staining demonstrated positivity of the tumor cells to terminal deoxynucleotidyl transferase (TdT) and cluster of differentiation (CD) 79a, while myeloperoxidase, CD3 and CD20 were negative (Fig. 4).

References


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Figure 1 T2-weighted MR image demonstrates a left parafalcine mass with isointensity to grey matter.

Figure 2 Histopathology shows rosettes of small neuronal cells surrounded by neuropil (hematoxylin and eosin stain, original magnification × 40).

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.