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Case Report

Epidermoid cyst of the cerebellar vermis: Case report of a rare medial topography ☆,☆☆

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ABSTRACT

Intracranial epidermoid cysts are very rare benign tumors representing less than 2% of intracranial tumors. They are located preferentially in the cerebellopontine angles, parasellar, and temporal regions. We report here the case of an epidermoid cyst of very uncommon medial location in the cerebellar vermis, in a 61-year-old female patient complicated with tumor protrusion into the foramen magnum and active hydrocephalus.

Introduction

Epidermoid cysts are very rare benign tumors and represent less than 2% of all intracranial tumors [1]. They are slow growing, developed either from ectodermal inclusions at the time of neural tube closure between the third and fifth week of gestation, or less frequently secondary to post-traumatic or iatrogenic penetration of the epidermis at the level of the subarachnoid spaces [2,3]. They are formed by the progressive accumulation of keratin debris from an epithelium of ectodermal origin "trapped" during embryogenesis, which is why they are sometimes compared to "skin in the wrong place" [1,2]. These tumors are ubiquitous and are located in the subarachnoid spaces, which they fill without destroying the nerve structures. They are usually located in the cerebellopontine angles as well as in the parasellar and temporal regions [1,4]. Medial forms are exceptional [3,5]. In this case, we report an epidermoid cyst located in the cerebellar vermis, followed by a brief review of the literature.

Observation

A 61-year-old female patient, without any particular medical history, presenting a left lateralized cerebellar syndrome with

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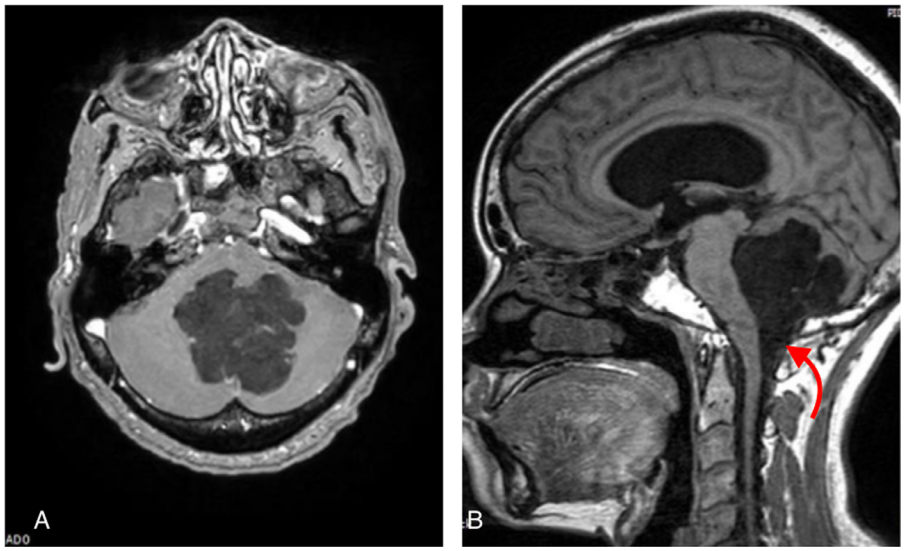

Fig. 1 – T1SE axial sequence with gadolinium (A) and T1SE sagittal sequence without gadolinium (B), showing the cystic mass of the cerebellar vermis in T1 hyposignal, without significant contrast, with obstruction of the V4 and protrusion into the foramen magnum (red arrow).

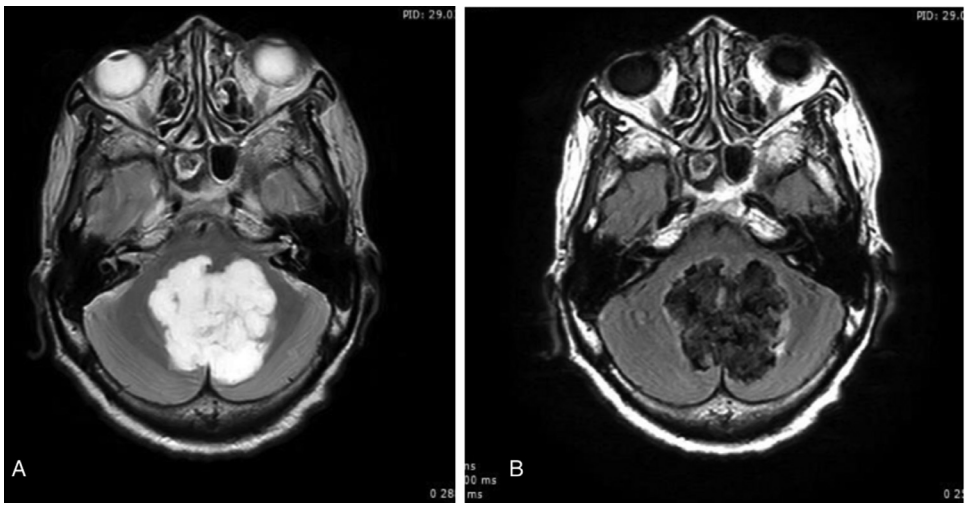

Fig. 2 – Axial T2 SE (A) and FLAIR (B) sequences, showing the cerebellar vermis mass in frank T2 hypersignal and heterogeneous FLAIR hyposignal.

headache and vertigo was referred to us for a brain MRI. The MRI-scan was performed with a 1.5 Tesla high-field MRI according to the following protocol: a 3D T1 SE sagittal sequence, followed by axial sequences in T2, FLAIR, diffusion with ADC mapping and 3D T1 SE after gadolinium injection.

The image analysis revealed a medial infratentorial intra-axial expansive process at the level of the cerebellar vermis, in contact with the fourth-ventricle (V4). This process is well limited, with scalloped contours, and measures axially 5.3 x 5.2 cm for a height of 6 cm. It appears in T1 hyposignal without significant enhancement (Figs. 1A and B); in T2 hypersignal and heterogeneous mild-hypersignal in FLAIR (Figs. 2A and B); in diffusion hypersignal without significant restriction on ADC mapping (Figs. 3A and B). It exerts a mass effect on the 4th ventricle with active hydrocephalus and tumor protrusion into the foramen magnum over a height of 6 mm (Fig. 1B).

The patient died following a short hospitalization and did not benefit from surgical management of her tumor.

Discussion

Intracranial epidermoid cysts are histologically benign, slowly growing, congenital neoplasms, that account for less than 2% of all intracranial tumors and 1%-5% of posterior cerebral fossa tumors [1,4]. Cerebellopontine angle (CPA) is the most common intracranial location for these pathologies constitut-

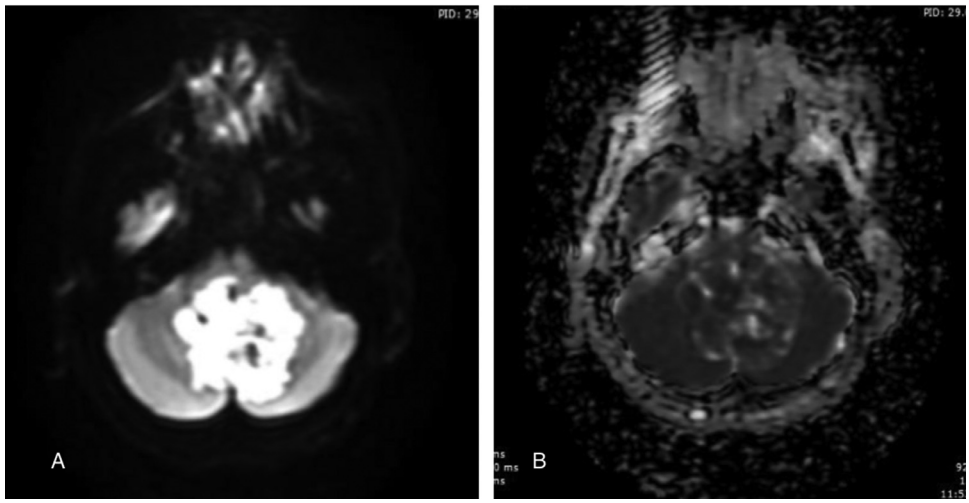

Fig. 3 – Axial diffusion sequence (A) and ADC mapping (B) showing the frank hypersignal in diffusion, without significant restriction on ADC mapping.

ing 40% of all cases [2,6]. The frequent lateralization of epidermoid cysts is due to the concomitant development of otic and optic vesicles [7]. Localization in the cerebellar vermis, as in our patient, is very uncommon with only few cases reported in the literature [8,9].

This lesion, reveals itself at all ages and without preference of sex; however, although occurring during the intra-embryonic life it is diagnosed most often between the third and the fifth decade or beyond, as it is the case in our 61-year-old patient [1,4]. However, exceptional forms have been diagnosed in children [1]. The clinical signs are dominated by the cerebellar syndrome which is most often at the front of the picture. The intracranial hypertension syndrome is less frequently encountered because of the late onset of hydrocephalus [6]. There is no correlation between the volume of the tumor and the presence of hydrocephalus at the time of discovery of the tumor because of its very slow growth and the probable persistence of cerebrospinal fluid flow space between the capsule and the walls of the ventricle [2]. This slow growth also explains the discovery of these tumors at a late stage with large sizes.

MRI is the gold standard for making a positive diagnosis and ruling out differential diagnoses. The epidermoid cyst typically presents as a mass with a signal close to that of the cerebrospinal fluid, in hyposignal T1, hypersignal T2 and a slightly heterogeneous signal in FLAIR with a frank hypersignal in diffusion [10]. The intermediate heterogeneous FLAIR signal allows to distinguish it from the adjacent cerebrospinal fluid. The diffusion hypersignal allows to differentiate it from the subarachnoid cyst which is its main differential diagnosis.

The frank hypersignal observed on the diffusion sequence would be related to the accumulation of protein material (keratin) and on the ADC mapping the restriction is not significant compared to the healthy brain parenchyma [6]. The diffusion hypersignal would then reflect a T2-shine-through effect rather than a real restriction.

Usually epidermoid cysts are well limited, without enhancement or perilesional edema.

In atypical forms, a spontaneously hyperintense T1 and hypointense T2 signal is described, suggesting protein content [4].

The differential diagnosis includes arachnoid cyst, dermoid cyst, cystic metastases, primary cystic neoplasia, and hydatid cyst [1,6].

The best therapeutic option with definitive cure remains total removal of the cyst and its capsule. This surgery is generally easy but this is not always the case in epidermoid cysts of the V4. The intimate adhesion of the capsule to the floor of the V4 limits this option given the neurological and vital risks involved [1]. Thus, incomplete exeresis or dispersion of the cyst contents intraoperatively exposes the patient to complications such as chemical or aseptic meningitis or communicating hydrocephalus [2]. Malignant transformation, though rare, should be suspected if rapid progression of neurological symptoms occur and tumor or residual tumor enhancement is there [11].

Finally, we would say that the location of epidermoid cysts in the cerebellar vermis is a very rare situation and seems to be more negative because of the proximity with the V4, exposing a higher risk of hydrocephalus and also because of the risk of tumor involvement in the foramen magnum.

Patient consent

The patient has signed an informed consent form.

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