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Case Report

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The Fourth Ventricle Epidermoid Cyst: About Two Cases

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Abstract

Epidermoid cysts are rare benign tumors developed from ectodermic inclusions. They usually sit at the ponto-cerebellar angle, the para-sellar region and the temporal fossa. Their seat at the fourth ventricle is unusual. We report two cases of squamous cell cyst in two young patients aged 21 and 33 admitted for intracranial hypertension syndrome associated with walking disorders. The diagnosis of epidermoid cyst of the V4 was evoked on the data of the MRI then confirmed in peroperative and in histology. The surgical excision was subtotal due to adhesion of the capsule to the upper part of the V4 floor. After a 12-month follow-up, the first patient showed no signs of tumor re-evolution. The second patient benefited from a ventriculo-peritonial derivation 45 days after the cyst was removed. After a 7-month follow-up, the patient showed no clinical signs suggestive of tumor re-evolution.

Keywords: Epidermoid cyst, Fourth ventricle.

INTRODUCTION

Epidermoid cysts, also called primitive cholesteatomas or Cruveilhier pearl tumors, are rare benign tumors (about 2% of primary intracranial tumors), developed from ectodermic inclusions. They usually sit at the ponto-cerebellar angle, the para-sellar region and the temporal fossa. Their location at the fourth ventricle (V4) is very rare [1]. We report two cases of epidermal cyst of the V4 and discuss the clinico-radiological, therapeutic and evolutionary features of this unusual localization.

CASE REPORT

Clinical Case: 1

Patient aged 21 years without significant pathological history, who was admitted in an emergency in a table of intracranial hypertension. On admission, the clinical examination found a sleepy patient with walking disorder. Ophthalmologic examination showed decreased visual acuity and fundus eye revealed bilateral stage II papillary edema. Cerebral tomodensitometry showed a spontaneously hypodense lesion, with scalloped contours and a density close to that of CSF, which did not take contrast, developed in the lumen of the V4 responsible for a tri-ventricular active hydrocephalus (Figure 1). It benefited from a ventriculoperitoneal bypass (DVP). The immediate postoperative follow-up was marked by the return of normal consciousness but also its visual acuity. Magnetic resonance imaging showed a heterogeneous hypointense lesion in T1 and hyperintense T2, with irregular contours, completely filling the V4, driving the cerebellar parenchyma upwards and compressing the spinal bulb (Figure 2). The diagnosis of an intraventricular epidermoid cyst was discussed and the decision to address the lesion was made. The patient was operated on a median sub-occipital route with peroperative discovery of a whitish tumor with a pearl-like appearance reminiscent of the characteristic appearance of a squamous cyst without cisternal extension. The surgical excision was subtotal with respect to a portion of the capsule that was very adherent to the upper part of the V4 floor. Immediate surgical follow-up was simple and after a 12-month follow-up, she was not relapsed.

Clinical Case: 2

Elderly patient of Miss B. N is a 33-year-old patient with no significant pathological history who was admitted to the Neurosurgery Department for cerebellar stato-kinetic syndrome for 3 months. At admission, the clinical examination found a conscious, cooperating patient with a stato-kinetic cerebellar

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syndrome. Ophthalmologic examination, in particular the fundus of the eye, revealed bilateral papillary edema stage I. Cerebral computed tomography showed a spontaneously hypodense lesion with a density close to that of CSF, not taking the contrast, developed in the lumen of the eye. V4 associated with associated overlying hydrocephalus (Figure 3). Magnetic resonance imaging showed a heterogeneous hypointense lesion in T1 and in the diffusion sequence; hyperintense in T2, with irregular contours, completely filling the V4, driving the cerebellar parenchyma upwards (Figure 4). The diagnosis of an intraventricular epidermoid cyst was discussed and the decision to address the lesion was made. The patient did not benefit from a derivation of her hydrocephalus.

It was operated in the ventral decubitus by a median sub-occipital route with peroperative discovery of a whitish tumor with a pearl-like appearance recalling the characteristic appearance of a squamous cell cyst (Figure 3). The surgical excision was subtotal with a portion of the capsule that was very adherent to the V4 floor.

Immediate surgical suites were marked by an aseptic meningitis which was taken care of before its release.

The patient was re-admitted 45 days after discharge from hospital, in a table of consciousness disorder, intracranial hypertension. CT performed urgently showed active tetraventricular hydrocephalus. It thus benefited from a ventriculo-peritoneal derivation (DVP).

After a 7-month follow-up, the patient showed no clinical signs suggestive of tumor re-evolution.

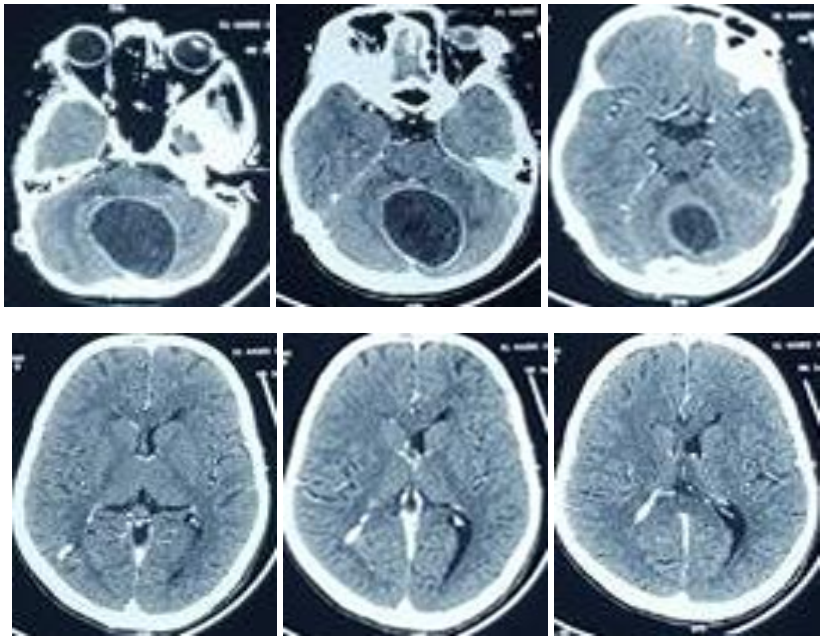

Figure 1: Cerebral computed tomography with the catheter of Ventriculoperitoneal Derivation (arrow)

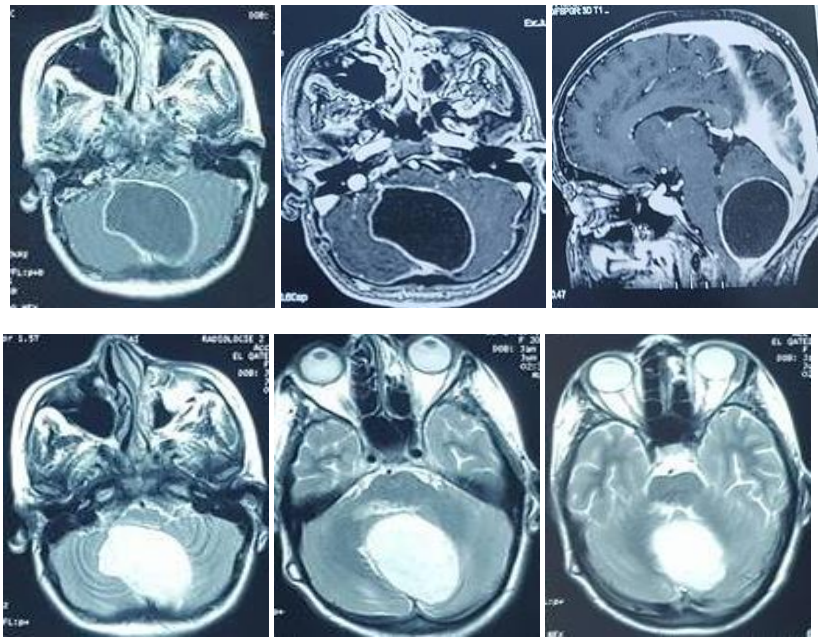

Figure 2: Magnetic resonance imaging (MRI)

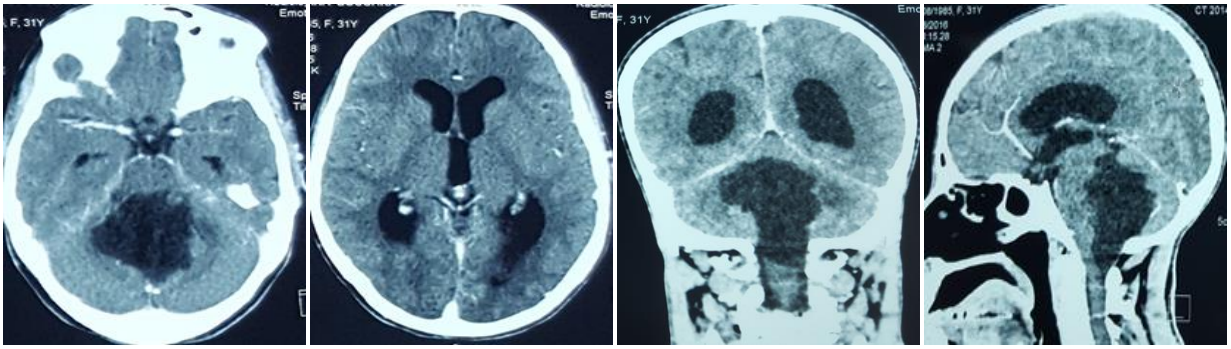

Figure 3: Cerebral computed tomography

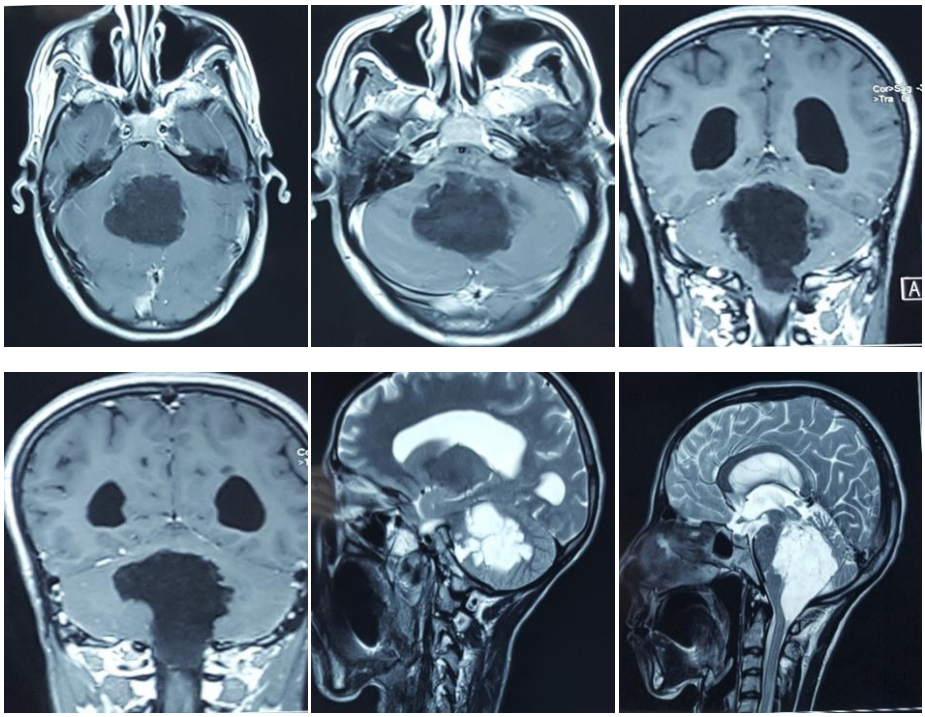

Figure 4: Magnetic resonance imaging (MRI)

DISCUSSION

The epidermoid cyst is a rare tumor, less than 2% of the intracranial tumors [2-4], formerly called cholestéatome or Cruveilhier pearl tumor. The frequent laterality of epidermoid cysts is related to the concomitant development of otic and optical vesicles [5]. The usual localization is found in the ponto-cerebellar angle (40 to 50% of the cases according to the series) [6-8] as well as in the para-sellar and temporal regions. Its location at the fourth ventricle is very rare [9].

This is a benign tumor secondary to either an ectopic inclusion of ectodermal elements at the time of closure of the neural tube between the 3rd and 5th week of gestation [10] or, less often, post penetration - traumatic [11,12] or iatrogenic [13] epidermis in the sub-arachnoid spaces. Despite its genesis during intrauterine life, the discovery of the epidermoid cyst is late between the 3rd and 5th decades [14], as is the case with our 2 patients.

Clinically, cerebellar syndrome is the most frequent manifestation, whereas intracranial hypertension syndrome is less frequent, since hydrocephalus is a late onset and is seen only in less than 50% of cases [14], Similarly, the very slow growth of the tumor and the probable persistence of CSF flow space between the capsule and the walls of the ventricle explains the absence of correlation between the size of the tumor volume and the presence of hydrocephalus at the time of discovery of the tumor [15]. Extension to the ponto-cerebellar cistern through the Luschka holes results in damage to the cranial nerves (mixed nerves, acoustico-facial bundle, trigeminal nerve).

The MRI aspect of epidermoid cysts is identical whatever their location [16]. They are isointenses in T1 and hyperintenses in T2, with clear but irregular limits, without perilional edema or contrast enhancement. Indeed, the signal is often inhomogeneous; it may be variable in intensity depending on the protein content of the tumor. Atypical forms have been reported, with spontaneously hyperintense mass in T1 and hypointense in T2, probably due to the presence of calcifications and high protein content [17]. Differential diagnosis problems with arachnoid cysts and tumor cysts are circumvented by the heterogeneous aspect in the Flair sequence, the increase in the signal in the diffusion sequence and especially the hyperintense and heterogeneous appearance in the CISS-3D sequence [18,19]. Histological analysis of epidermoid cysts is the same, regardless of intracerebral localization. From the therapeutic point of view, the total excision of the cyst and its capsule remains the only guarantee of a definitive cure. However, and as in both cases, the intimate adhesion of the capsule to the V4 floor limits this option considering the neurological and vital risks incurred. Thus, in a review of the literature by Tancredi A. *et al* [13], of 66 patients operated for a V4 epidermoid cyst between 1974 and 2003, total excision was performed in only 30% of cases. The postoperative course is usually simple; however, chemical meningitis

can occur and lead to a communicating hydrocephalus, the prevention of which requires complete removal as much as possible, the avoidance of the dispersion of the cyst content in peroperative operation, as well as the irrigation of the operative focus by of hydrocortisone and even postoperative administration of dexamethasone [1,18]. In postoperative surveillance, diffusion imaging is used to determine whether or not the excision is complete. In case of residual tumor, an annual MRI monitoring allows to evaluate the evolutive potential of the residue [12].

CONCLUSION

The V4 KE are rare, slowly evolving benign congenital tumors. They are most often seen in an ataxocerebellar array associated with intracranial hypertension. Their diagnosis appears relatively characteristic in imaging. The only treatment is surgical.

Conflict of interests:

We have no conflict of interest.

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