



RESEARCH ARTICLE

EPIDERMOID CYST OF THE FOURTH VENTRICLE: AN UNUSUAL LOCATION

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Manuscript Info

Manuscript History

Received: 25 October 2020

Final Accepted: 28 November 2020

Published: December 2020

Key words:-

Epidermoid Cyst, Fourth Ventricle, Diffusion MRI

Abstract

Epidermoid cysts are slow-growing congenital tumors developed from ectodermal inclusions. They usually sit at the cerebellopontine angle or basal cistern, their location in the fourth ventricle are exceptional. We report the case of a 44-year-old patient admitted to the Neurosurgery department for chronic daily headaches with visual impairment recently aggravated by cerebellar stato-kinetic syndrome. The diagnosis of epidermoid cyst of the fourth ventricle was suspected on MRI especially in diffusion sequence and then confirmed by the anatomopathological studies. Subtotal surgical excision was performed. The evolution was marked by the disappearance of clinical signs. Radiological and clinical follow up were indicated.

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Introduction:-

Epidermoid cysts (EC), also called primitive cholesteatomas or Cruveilhier pearl tumors are rare benign tumors developed from ectodermic inclusions. Their localizations in the cisterna magna and fourth ventricle are rare, and represent 5-18% of all intracranial EC. [1]

We report a case of epidermal cyst of the fourth ventricle and discuss the clinico-radiological, therapeutic and evolutionary features of this uncommon localization.

Case Report:

A 44 years old female without significant pathological history, presented 2 months before her admission to the Neurosurgery department for chronic daily headaches with visual impairment recently aggravated by cerebellar stato-kinetic syndrome.

On admission, the clinical examination found a conscious patient, well oriented in time and place, with a stato-kinetic cerebellar syndrome.

Ophthalmologic examination showed decreased visual acuity and fundus eye revealed bilateral stage I papillary edema.

Magnetic resonance imaging revealed an extra axial median lesion in the fourth ventricle, compressing the medulla oblongata with irregular but clear limits, without perilesional edema. It appears hypointense in T1, hyperintense in T2, inhomogeneous signal in FLAIR with restricted diffusion and without contrast enhancement. MRI was suggestive of typical intraventricular epidermoid cyst. (Figure 1)

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The therapeutic attitude was a surgical resection of the mass. The patient was operated with subtotal exeresis because of its intimate contact with the posterior side of the medulla oblongata.

The intraoperative appearance of a “beaded tumor” with nodular surface and white pearly color was compatible with an epidermoid cyst. Anatomopathological checkup confirms the diagnosis.

The evolution was marked by the disappearance of clinical signs. A radio clinical follow up was indicated.

Discussion:-

Epidermoid cysts are benign, slowly progressive tumours developed from ectodermic inclusions. They usually sit at the cerebellopontine angle or basal cistern, their location in the fourth ventricle are rare. [1,2]

These are congenital tumors that are revealed at any age without significant sexual predominance, with a peak of frequency between the 3rd and 5th decade.[3]

Clinical symptoms and signs of epidemoid cysts in the posterior fossa are dominated by cerebellar syndrome and headaches, whereas intracranial hypertension syndrome and hydrocephalus are less frequent. [4]

Magnetic resonance imaging (MRI) is the most powerful tool for the early diagnosis of the epidermoid cyst. There MRI aspect is identical whatever their location. They appear as a well limited mass with irregular limits, without perilesional edema. The signal of epidermoid cyst is isointense in T1 and hyperintense in T2, with diffusion restriction and no contrast enhancement. However, the signal of the tumor can be heterogeneous depending on the protein content of the tumor. In the diffusion sequence, the increase in the signal reflects the solid character of the mass, which allows differential diagnosis with arachnoid cysts. It also helps to determine whether or not the excision is complete. [1, 5]

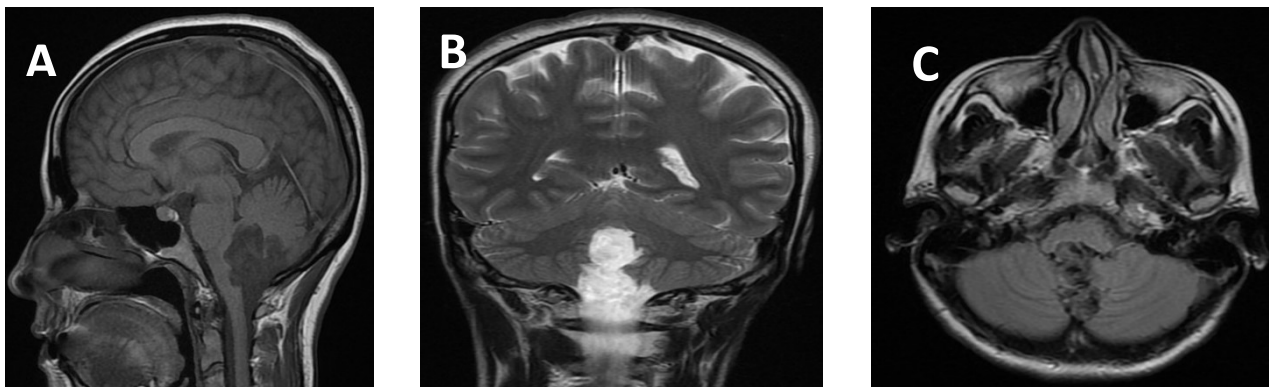
Surgery remains the only effective treatment especially when the resection is total including the tumor capsule. [3] However, complete excision is often difficult in posterior fossa tumors because they are usually adherent to surrounding structures as in our case.

In the case of incomplete excision, the growth of the residue is slower than that of the native tumour, nevertheless it requires annual MRI monitoring to assess its evolutive potential. [4]

Radiotherapy has no place in these non-neoplastic lesions.[4]

Rapid progression of neurological signs and the presence of tumor enhancement should be a wake-up call of malignant transformation.[6]

Figures:-



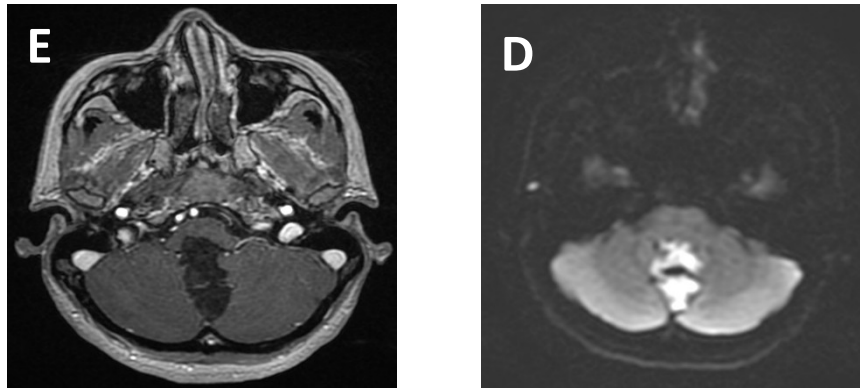


Figure 1:-Magnetic resonance imaging (MRI) revealed an extra axial large cystic lesion located within and expanding the fourth ventricle. The lesion appeared hypointense on T1WI (A), hyperintense on T2WI (B) and heterogenous signal on FLAIR sequence (C). There was restriction on DWI (D) and lesion did not enhance on gadolinium administration (E).

Figure legend:

Figure 1: Magnetic resonance imaging (MRI) revealed an extra axial large cystic lesion located within and expanding the fourth ventricle. The lesion appeared hypointense on T1WI (A), hyperintense on T2WI (B) and heterogenous signal on FLAIR sequence (C). There was restriction on DWI (D) and lesion did not enhance on gadolinium administration (E).

Conclusion:-

Epidermoid cysts of the fourth are a rare benign tumor. They usually reach a considerable size before producing symptoms. Their diagnosis appears relatively characteristic in imaging and diffusion MRI remains the key element for positive diagnosis and post-operative monitoring. Surgery remains the only effective treatment; it should be extirpated totally to avoid recurrence.

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