Pauci-symptomatic large epidermoid cyst of cerebellopontine angle: case report

Maxi cisti epidermoide dell'angolo ponto-cerebellare paucisintomatica: descrizione di un caso

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Cerebellopontine angle tumours • Epidermoid cyst • Diagnosis

Parole chiave

Tumori dell'angolo ponto-cerebellare • Cisti epidermoide • Diagnosi

Summary

It is estimated that 10% of intra-cranial tumours are localized in the cerebellopontine angle and internal auditory canal and early symptoms of the different histological forms are almost identical. Acoustic neuroma account for 90% and meningioma for 5-10% of these tumours, while a small percentage of rare tumours exist, the most frequent being epidermoid cyst, also known as congenital cholesteatoma or keratoma. The case is reported here of a large epidermoid cyst of the right cerebellopontine angle, and the clinical-radiological course is reviewed. The patient, a 35-year-old, male, initially presented an episode of objective rotatory vertigo, and a history of right ear fullness, of a few months' duration, with normal otoscopy. Audiometric test was normal in left ear, while slight pantonal sensori-neural hypoacusia was observed in the right ear. The impedenzometric findings were normal as was the vestibular test. Auditory brainstem evoked response showed an increased latency of fifth wave. Gadolinium-enhanced magnetic resonance imaging of brain revealed the presence of a voluminous epidermoid cyst occupying the extra-axial side of the right cerebellopontine cistern with superior extension into the cistern. Due to the low-grade of symptoms, we had chosen to wait and not perform surgery immediately, with otologic and vestibular test-controls every 6 months, with cerebral magnetic resonance imaging to control extension of the mass, without radiation exposure for the patient. One year after diagnosis, at the last control, otofunctional findings were not modified and repeat magnetic resonance imaging did not demonstrate important variations compared to the first. Thus, the choice not to proceed with surgery was justified since surgery is burdened by the risk of important complications. At magnetic resonance imaging, the epidermoid cyst, unlike the majority of intra-cranial tumours, such as acoustic neuroma and meningioma, does not show gadolinium-enhancement; this again supporting the important role of magnetic resonance imaging in the differential diagnosis of intra-cranial neoformations. It is, therefore, worthwhile stressing the validity of the approach, step by step, in the diagnosis of patients with otologic symptoms, together with the importance of magnetic resonance imaging that, in comparison with computed tomography, allowed us to exactly assess the growth rate of the mass and to "wait and see" without risks and without radiations for the patient.

Riassunto

L'angolo ponto-cerebellare (APC) ed il canale acustico interno (CAI) rappresentano le sedi di localizzazione di circa il 10% dei tumori intracranici, e la sintomatologia d'esordio delle diverse forme istologiche spesso non differisce di molto fra loro. La grande maggioranza di tali tumori è costituita dal neurinoma dell'acustico (90%) e dal meningioma (5-10%), mentre esiste una restante percentuale di tumori considerati rari di cui il più frequente è la cisti epidermoide, anche conosciuta con il nome di colesteatoma congenito o cheratoma. Descriviamo un caso di voluminosa cisti epidermoide dell'APC destro illustrandone le caratteristiche e l'evoluzione clinico-radiologica. Il paziente di 35 anni si è presentato alla nostra osservazione a causa di un episodio di vertigine rotatoria oggettiva accompagnata da fullness auricolare destra da alcuni mesi con otoscopia negativa. L'esame audiometrico risultava normale a sinistra mentre a destra era presente un'ipoacusia pantonale percettiva di lieve entità. I dati impedenzometrici erano nei limiti della norma così come l'esame vestibolare. I potenziali evocati troncoencefalici mostravano una latenza dell'onda V aumentata. La RMN encefalica con gadolinio e.v. ha documentato la presenza di una voluminosa cisti epidermoide extra-assiale occupante l'angolo destro della cisterna ponto-cerebellare con estensione superiore all'interno della cisterna. In virtù della scarsa rilevanza dei sintomi non abbiamo ritenuto opportuno intervenire subito chirurgicamente sulla neoformazione preferendo una vigile attesa, con controlli semestrali della funzionalità uditiva, vestibolare e RMN cerebrale per monitorare l'evoluzione volumetrica della massa evitando l'esposizione del paziente a radiazioni ionizzanti.

Introduction

The cerebellopontine angle (CPA) and the internal auditory canal (IAC) account for 10% of the sites in which intra-cranial tumours develop and symptoms do not depend upon different histological types [1]. The large majority of these tumours are acoustic neuroma (90%) or meningioma (5-10%), while a small percentage of rare tumours exists, the most frequent being the epidermoid cyst (Table I) [1-3]. Accurate histological diagnosis is the first step to start a valid and specific therapeutic approach for each individual patient providing useful information for prognosis according to the different histotypes.

Epidermoid cyst of the CPA (0.2-1% of all intracranial tumours, occupying the third place as far as concerns frequency in CPA tumours) is a congenital slow-growing benign lesion, derived from cranial or bony inclusion of embryonal ectodermic tissue. This cyst, covered by epithelial cells, contains keratin and cholesterol and for this reason, it is also referred to as congenital cholesteatoma or keratoma [4-8].

The average age at clinical onset is usually between 20 and 40 years of age, with a male predominance [7 9]. The otologic symptoms are: sensori-neural hypoacusia, tinnitus and vertigo, with more or less early occurrence and of different degree, being related to the localization of the lesion and its growth rate. Only in an advanced stage of the disease, does it compress the intra-cranial vessels, the cranial nerves (with spasms and facial neuralgias) and the brainstem, obstructing circulation of the liquor; in these cases, it is necessary to perform microneurovascular decompression [10 11]. Radiological features allow the various histological types to be differentiated according to some magnetic resonance imaging (MRI) parameters, even if these are not always pathognomonic and specific [12 13] (Table II).

The case is reported of a large epidermoid cyst of the right CPA and the characteristics and clinical-radiological course are described.

Case report

A 35-year-old male, came to our attention complaining of objective rotator vertigo with the sensation of ear fullness in the right ear which had been present for a few months, with normal otoscopy.

The audiometric test was normal in the left ear, while a slight degree of pantonal sensori-neural hypoacusia was observed in the right ear (Fig. 1). The impedenzometric findings were normal. Vestibular tests showed no spontaneous, or revealed signs of, vestibular dysfunction, with negative spontaneous and positional tests and absence of pathological nystagmus. The labyrinthine reflexes with caloric stimulation (Fitzgerald-Hallpike) were quantitatively and qualitatively normal.

Brainstem auditory evoked response (BAER) showed an increased latency of the fifth wave, with preserved morphology.

Tab. I. Rare tumors of CPA and IAC and their symptomatology (modified from Kohan et al. [13])

Histological type	No. patients (24)	Hypoacusia	Tinnitus	Vertigo	Facial paralysis	Trigeminal neuralgia	Headache	Other symptoms
Epidermoid cyst	4	1	/	/	/	1	1	Epileptic crisis 1
Lipoma	4	2	3	2	1	/	/	/
Facial neuroma	2	1	1	1	1	/	/	/
Arachnoid cyst	2	1	1	/	/	/	1	/
Choroid plexus papilloma	2	/	/	1	/	/	1	Peripheral motor deficit 1
Metastasis of adenocarcinoma	2	2	2	1	1	1	1	/
Metastasis of neuroblastoma	1	/	/	1	/	/	1	/
Ependymoma	1	1	/	1	/	/	/	/
Lymphoma	1	1	1	/	/	/	1	/
Cholesterinic cyst	1	1	1	1	/	/	/	/
Angioleiomyoma	1	1	1	/	/	/	/	/
Venous hemangioma	1	/	/	1	/	/	/	/
Angioma cavernosus	1	1	1	/	1	/	/	/
Pons glioma	1	1	1	1	/	1	/	/

Table II. MRI features of rare tumours of CPA and IAC (modified from Kohan et al. [13]).

Histological type	T1	T1 gadolinium	T2
Epidermoid cyst	Hypointense/heterointense	Isointense	Hyperintense
Lipoma	Hyperintense	No change	Hypointense
Facial neuroma	Hypointense	Hyperintense	Variable
Arachnoid cyst	Hypointense	Hypointense	Hyperintense
Choroid plexus papilloma	Variable	Hyperintense/ Heterointense	Hyperintense/ Heterointense
Metastasis of adenocarcinoma	Hypointense	Hyperintense	Hyperintense/variable
Lymphoma	Isointense/heterointense	Hyperintense	Hyperintense/ Heterointense
Cholesterinic cyst	Hyperintense	Hyperintense	Hyperintense
Angioma cavernosus	Hyperintense	Hyperintense	Hyperintense

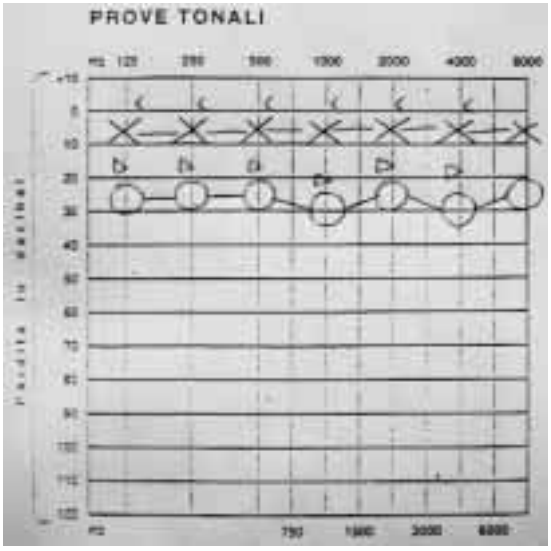


Fig. 1. Audiometric test at first observation.

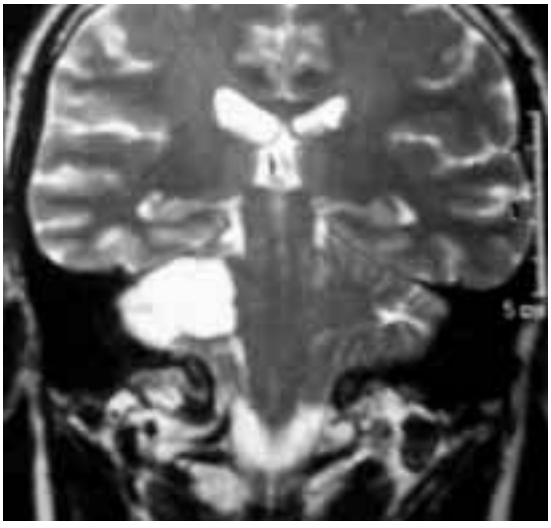


Fig. 2. Coronal MRI: large epidermoid cyst in CPA.

Gadolinium-enhanced MRI of the brain documented the presence of a large epidermoid cyst occupying the extra-axial side of the right cerebellopontine cistern with superior extension into the cistern (Fig. 2). This lesion determined a moderate compression and dislocation to the right side of the pons and the middle cerebellar peduncle, without diffusion in the IAC (Fig. 3). The V cranial nerve (cn) was displaced upwards, the VII and VIII cn lower down. The mass had lobulated and slightly irregular borders. The MRI signal was similar to that of the cerebral liquor in all the sequences, showing an internal septal in T2-weighted MRI images. Ventricle IV appeared slightly distorted, without supratentorial hydrocephalus. The cerebral white substance was normal. The mass was adherent to the sigmoid vein without contact, and it showed no gadolinium enhancement.

The symptoms, the comparative clinical-instrumental data and the neuroradiological images were modest, due to the extreme plasticity of the neurocerebral system that is damaged only when the neoformation is large enough to cause compression on the Central Nervous System.

Due to the low-grade of the symptoms, we had chosen to wait and not to perform surgery on the performing brain MRI to monitor the mass, thus avoiding radiation exposure for the patient.

One year after diagnosis, the otofunctional data have remained unchanged and the MRI images do not show any significant variations compared to the first examination (Fig. 4).

Fig. 3. Coronal MRI: encephalic deformation by the cyst, out from the IAC.

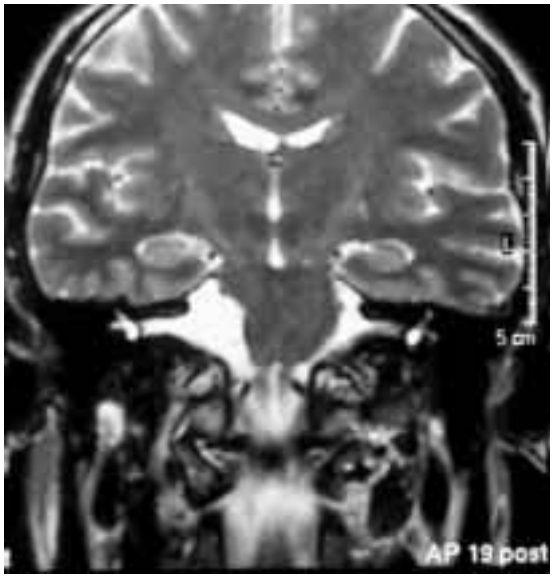
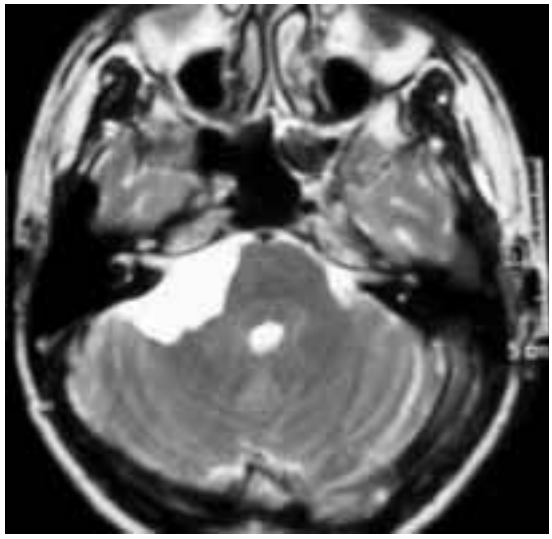


Fig. 4. Axial MRI after 1 year: epidermoid cyst is not modified in dimension.



Discussion

The clinical findings of the epidermoid cyst are not useful in formulating a differential histopathological diagnosis with respect to other intra-cranial neoformations. The otofunctional pattern, unlike MRI, did not lead us to suspect a space-occupying lesion. Furthermore, due to the lack of specificity of the onset symptoms, their persistence might induce the clinician to suspect the presence of an intracranial neoformation.

Congenital cholesteatoma is a benign lesion that may remain with low grade symptoms, or even silent, for a long time, since this is a small, slow-growing lesion. Thus, diagnosis is often made when the mass has already reached large dimensions.

In the absence of serious symptoms, as in our case, we justify the choice not to proceed with surgical treatment since surgery may be burdened by severe complications. Possible post-operative complications following removal of tumours localized in the CPA are: neural paralysis (abducent, facial, trigeminus), dysphagia, deafness, worsening of cerebellar symptoms (ataxia, dysartria, dysmetry, nystagmus), hemiparesis, papilloedema [14 15]. According to the literature, post-operative morbidity and mortality can be due to resection of the cystic capsule, prolonged cerebral retraction, and, frequently, aseptic meningitis and hydrocephalus. To reduce post-operative mortality some Authors have recommended a transparenchymal approach, to be performed in a few selected patients [16].

The large majority of all intracranial tumours, such as acoustic neuromas and meningiomas, show gadolinium enhancement at MRI [17]. The epidermoid cyst does not show any gadolinium enhancement (as observed also in our case), thus supporting the extreme importance of MRI in the differential diagnosis of intra-cranial neoformations [18].

Once again, we wish to underline the validity of the "step by step" approach in the diagnostic workup of patients with otologic symptoms and to stress the importance of MRI that, in our case, allowed us to carefully check the exact growth rate of the mass and to adopt the "wait-and-see" approach without any risk. MRI is extremely important during follow-up, especially in those cases in which surgery is not the first and immediate choice, since MRI is highly specific, sensitive and, unlike CT, avoids radiation for the patient.

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