

Imaging of cerebellopontine angle tumors: case series.

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Abstract: *The tumoral pathology of the cerebellopontine angle is most often benign, dominated by: vestibular schwannoma and meningioma. We report in this article the experience of the radiology department of the Hassan II University Hospital of Fez on 25 patients with a tumor of the PCA, over a period of 2 years, from January 2019 to December 2021. Clinical symptomatology consisted mainly of vestibular syndrome in 15 patients, headache in 8 patients, cerebellar syndrome in 5 patients and more rarely HTIC syndrome in 3 patients. An imaging assessment was performed, including a CT scan in 16 patients and all our patients had an MRI. A clinical-radiological-histological comparison allowed us to retain the diagnosis of certainty. The age of our patients ranged from 27 to 64 years, with a mean age of 45.5 years. Females were predominant (16F/9H) with a sex ratio of 1.7. The tumors identified were as follows: schwannoma (n=16), meningioma (n=6), epidermoid cyst (n=2), arachnoid cyst (n=1). The purpose of this work is to discuss the specific characteristics of CT and MRI imaging of the different tumors of the APC. Therefore, to show the importance of the different imaging methods in the diagnosis, therapeutic decision and monitoring.*

Keywords: cerebellopontine angle, tumors, MRI, CT.

INTRODUCTION :

The tumor pathology of the cerebellopontine angle is most often benign. They are dominated by: vestibular schwannoma, meningioma and epidermoid cyst. Imaging plays a central role in the diagnosis, the definition of therapeutic indications and the follow-up of patients with tumors of the cerebellopontine angle. The contribution of different neurophysiological and imaging techniques, in this case MRI and CT in both positive and etiological diagnosis of expansive processes of the cerebellopontine angle is the subject of this work.

METHODS:

This is a retrospective study of 25 patients with CPA tumor, collected in the Radiology Department of the University Hospital of Fez over a period of 2 years, from January 2019 to December 2021. This work was based on the exploitation of clinical records and the interpretation of the radiological assessment.

RESULTS:

In the light of a descriptive analysis of the different data, the age of our patients varied between 27 and 64 years, with an average of 45.5 years, with a predominance of the age group between 50 and 60 years. A female predominance with a sex ratio M/F: 1.7. The most frequent clinical symptom was the vestibular syndrome, which consisted mainly of sensorineural deafness, vertigo and tinnitus, with a percentage of 48%. Headache was second in line with a percentage of 26%. Cerebellar syndrome was present in 26% of cases, and intracranial hypertension was the least frequent with a percentage of 10%.

Neuroradiological exploration, in particular MRI, was performed for all our patients, whereas CT scans were performed in 16 cases as an emergency measure. Imaging data showed 16 cases of schwannoma (64%); 6 cases of meningioma (24%); 2 cases of epidermoid cyst (8%) and 1 case of arachnoid cyst (4%).

Schwannomas represented the majority in our series, with 13 cases of vestibular schwannomas (Figure 1), two of which had neurofibromatosis type 2 (Figure 2), 2 cases of trigeminal schwannomas (Figure 3), and 1 case of facial schwannoma (Figure 4).

6 cases of meningioma were reported in our series (Figure 5). The tumor had an ovoid shape and well-limited contours in all cases. Peripheral calcifications were noted. The tumor had an obtuse connection angle with the petrous bone.

2 cases were reported of epidermoid cyst, (figure 6) one on the left and the other on the right APC.

Finally, only 1 case of arachnoid cyst was found in the left cerebellopontine angle (Figure 7). The tumor had an ovoid shape with well-limited contours.

DISCUSSION:

Schwannomas:

These are benign tumors of nerve origin that can arise on all cranial pairs [1].

Vestibular schwannoma:

Vestibular schwannoma, represents 80% of the tumors of the CPA [2]. Although the schwannoma develops on the vestibular nerve, the first chronological symptom and in order of frequency is an unilateral sensorineural deafness that is slowly evolving [3]. In patients over 50 years of age, this hearing loss is often overlooked by the patient, which explains the long average delay (more than 3 years) between the onset of symptoms and diagnosis. The other main symptoms are tinnitus and vertigo. Clinical examination often shows nystagmus [4]. More rarely, the discovery of the tumor is made in front of a neurological symptomatology with a cerebellar syndrome and/or signs of intracranial hypertension.

On MRI, the vestibular schwannoma appears as a rounded or oval tumor, with clear and regular contours. Its angles of connection to the posterior surface of the petrous bone are acute. On T1 and T2 weighting, the signal is not specific [5]. After injection of contrast agent, there is always a clear enhancement of the vestibular schwannoma. In diffusion sequence, the tumor appears isointense with a high ADC in case of a benign lesion. In case of malignancy, the tumor appears hyperintense with a low ADC. The enlargement of the internal auditory canal (IAC) is a sign that has long been a mainstay of the diagnosis of vestibular schwannoma and may accompany 50% of cases.

On CT, it is spontaneously isodense and may rarely be hypo- or hyperdense. Rare cases of calcification have been described in the literature [6]. After injection of the contrast agent, the vestibular schwannoma becomes hyperdense, with homogeneous contrast except for large schwannomas or areas of necrosis. The schwannoma is often centered by the IAC. This particular location of the tumor allows it to be differentiated from other tumors of the CPA, especially meningiomas.

Trigeminal Schwannoma:

It can develop at the expense of the nerve's root in the CPA, at the level of Meckel's cavum or from Gasser's ganglion. MRI allows a precise study and shows a localized thickening of the nerve. The lesions are hypo- or isosignal in T1, hypersignal in T2 and intensely enhancing after injection of gadolinium. On CT, it is only visible if it is greater than 1 cm. This spontaneously isodense mass enhances after injection of contrast agent. When the tumor is larger, it may erode or destroy the petrous apex.

Facial nerve schwannoma:

A large facial schwannoma may be difficult to distinguish from schwannoma of the VIII nerve, but in general, the lesion appears eccentric in the IAC relative to the latter. On MRI, it appears in T1 iso or hypointense homogeneous with enhancement after injection of gadolinium. MRI provides an accurate assessment of extension. The CT scan shows enlargement of the geniculate ganglion compartment and the facial canal.

Mixed nerve schwannoma:

Schwannomas of the glossopharyngeal (IX), vagus (X), and spinal (XI) nerves are not common. The schwannoma of the hypoglossal nerve (XII) is even rarer. They are encountered with predilection in neurofibromatosis type 2 [7]. On MRI the lesion is

usually hypointense in T1 and hyperintense in T2. It is intensely enhanced after injection of gadolinium. The extension, which is well appreciated by MRI, may be upwards into the CPA, but rarely into the tympanic cavity. On CT, enlargement of the posterior horn, a normal IAC or erosion of its lower lip should draw attention to this diagnosis. The lesion is rarely calcified but cystic remodeling may be seen.

Meningiomas:

They account for 10-14% of the posterior cerebral fossa tumors [8] and 12% of meningiomas are found in the CPA [9]. They most often develop in the posteromedial part of the petrous bone and are eccentric to the porus. Typically, they appear sessile, with a broad base of dural implantation against the petrous bone, and obtuse connection angles. More rarely, they appear flat, linear. It may be seen as part of a meningiomatosis of the base or be encountered in neurofibromatosis type 2.

The clinical presentation of CPA meningiomas is variable: when they are small and without contact with the cranial nerves, they are usually asymptomatic and then discovered incidentally or accompanied by headache without specific character. When they are larger and/or when they arise in contact with the cranial nerves, they can manifest themselves by a symptomatology linked to their mass effect, in particular by damage of the V nerve with facial hypoesthesia or damage of the VIII nerve with vertigo.

The usually slow growth of these tumors accounts for the fact that the symptoms are often not very intense or progressive, which may lead to late diagnosis.

On T1 weighting, meningiomas are rather iso intense [2]. In T2 weighting, the signal of meningiomas depends on their histological nature. Meningothelial meningiomas (most frequent in the cerebellopontine angle) and angioblastic meningiomas are rather hyperintense in the gray matter [2], whereas transitional and fibroblastic meningiomas are rather iso or hypointense [2]. After injection of contrast agent, contrast enhancement is intense, early, prolonged and homogeneous. This enhancement mode is due to the tumor hypervascularization of the meningioma. [2]

It is important to note the very high frequency of enhancement of the meninges adjacent to the lesion, which most often appears thickened, giving the "comet tail" sign. [2]

Calcifications, if present, give a hypointense signal on the T2* sequence, but their diagnosis in MRI is less obvious than in CT.

On CT, the most suggestive aspect is that of a spontaneously hyperdense lesion, sometimes iso or hypodense, which forms an obtuse connection angle with the petrous bone. Usually the tumor is eccentric to the IAC. After injection of contrast agent, it enhances more prominently and rapidly compared to the schwannoma. The presence of disseminated calcifications in the tumor is an element in favor of meningioma. It may be fully calcified, defining psammomatous meningioma. Meningiomas may be accompanied by osteocondensation of the petrous bone due to the stimulation of osteoblasts by the meningioma cells.

Epidermoid cysts:

This tumor represents 0.2% to 1% of all primary intracranial tumors and less than 5% of CPA tumors [1].

The symptomatology of epidermoid tumors is due to compression. It can be either an auditory or vestibular symptomatology due to irritation of the acoustic-facial bundle or, more frequently, sensory disturbances of a hemifacial region reflecting compression of the trigeminal nerve.

MRI in T1 and T2 weighted sequences typically shows a lesion with a liquid signal, not enhanced after injection of gadolinium, but often heterogeneous, especially in T2, and characterized by a mottled appearance in the Fiesta high resolution T2 sequence. This is noted in 60% of cases [10].

Certain arguments such as the heterogeneity of the signal, the sharp and irregular contours in "geographical map" molding the surrounding structures as well as the extension in the sub-arachnoid spaces plead in favor of the diagnosis. However, this MRI aspect is sometimes inconclusive or atypical, hence the need for other more specific sequences [11].

Diffusion sequences have made the diagnosis of epidermoid cysts easier. There is a marked increase in signal within epidermoid cysts, related to a decrease in the diffusion of water molecules within the lesion due to its relatively thick content compared to pure fluid content. The apparent diffusion coefficient (ADC) is restricted thus allowing differential diagnosis with lesions whose ADC is identical to that of the CSF, notably arachnoid cysts. [12]

CT, usually shows a mass with sharp, irregular contours, without perilesional edema and molding the adjacent brain structures. This mass is hypodense measuring between - 50 + 20 HU, often heterogeneous with a "salt and pepper" appearance and not enhanced after intravenous injection of contrast agent [13,14].

Arachnoid cyst:

This is a benign, congenital cyst occupying the arachnoid space. They are discovered incidentally, without any clinical consequences [15]. The arachnoid cyst represents 1% of intracranial tumors. The CPA is its second most common location after the sylvian valley.

MRI describes a non-lobulated, homogeneous, CSF-isointense mass, which means hypointense in T1 and hyperintense in T2, most often fading on the Flair sequence. After injection of gadolinium, the cyst remains hypointense unless there is peripheral meningeal inflammation, or communication with the CSF that leads to contrast of the cyst wall. Diffusion shows a liquid-like diffusion coefficient, allowing differential diagnosis essentially with epidermoid cysts [15].

On CT, these are hypodense lesions, well limited and with the same density as the CSF. It is responsible for a mass effect on the elements of the posterior cerebral fossa. Hydrocephalus by compression of the 4th ventricle is possible [15].

Occasionally, arachnoid cysts may be responsible for thinning of the bone opposite (scalloping) [15].

Dermoid cyst:

Dermoid cysts represent 0.3% of intracranial tumors. They are congenital tumors, dermoid cysts contain appendages of the skin, such as hair, follicles, sebaceous and sweat glands. These cysts are benign lesions and develop slowly, due to sebaceous secretion and desquamation of the epithelium.

It presents with T1 hypersignal, hyposignalT2 patches, not enhanced by contrast agents in MRI and hypodensity with a thicker wall compared to squamous cyst, it is often calcified [15].

CONCLUSION:

The major third of CPA tumors in adults are vestibular schwannoma, meningioma, and epidermoid cyst. Imaging has undergone a considerable expansion with the new generation of CT and MRI machines. The pathology of the cerebellopontine angle has become perfectly accessible thanks to these modern imaging means.

FIGURE :

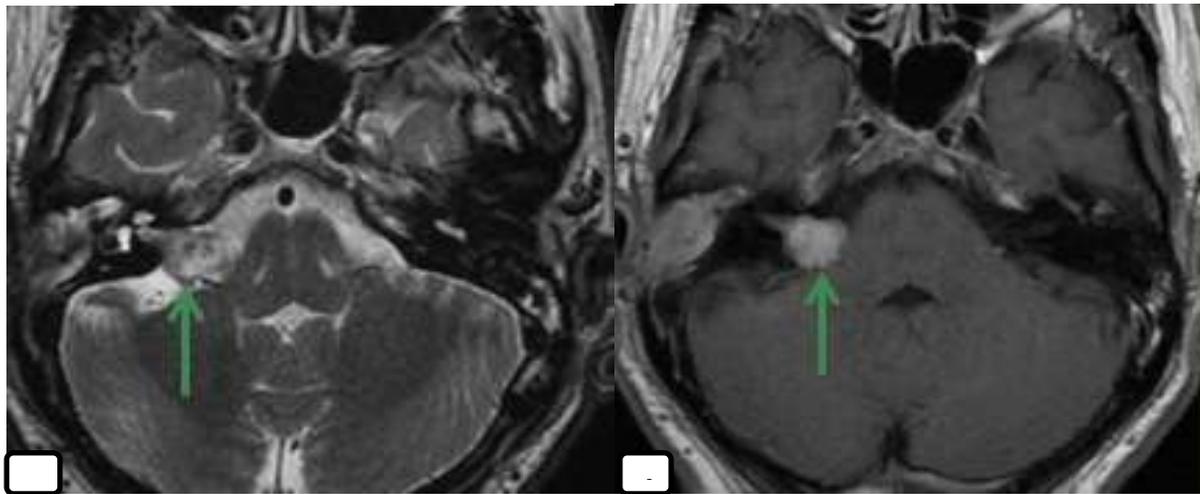


Figure 1: MRI in axial section in T2 sequence (a) and in axial section in injected T1 sequence(b): Extra-axial process at the level of the right internal auditory canal hyperintense in T2 intensely enhanced after contrast. Note the "ice horn" sign (arrow) → Vestibular Schwannoma

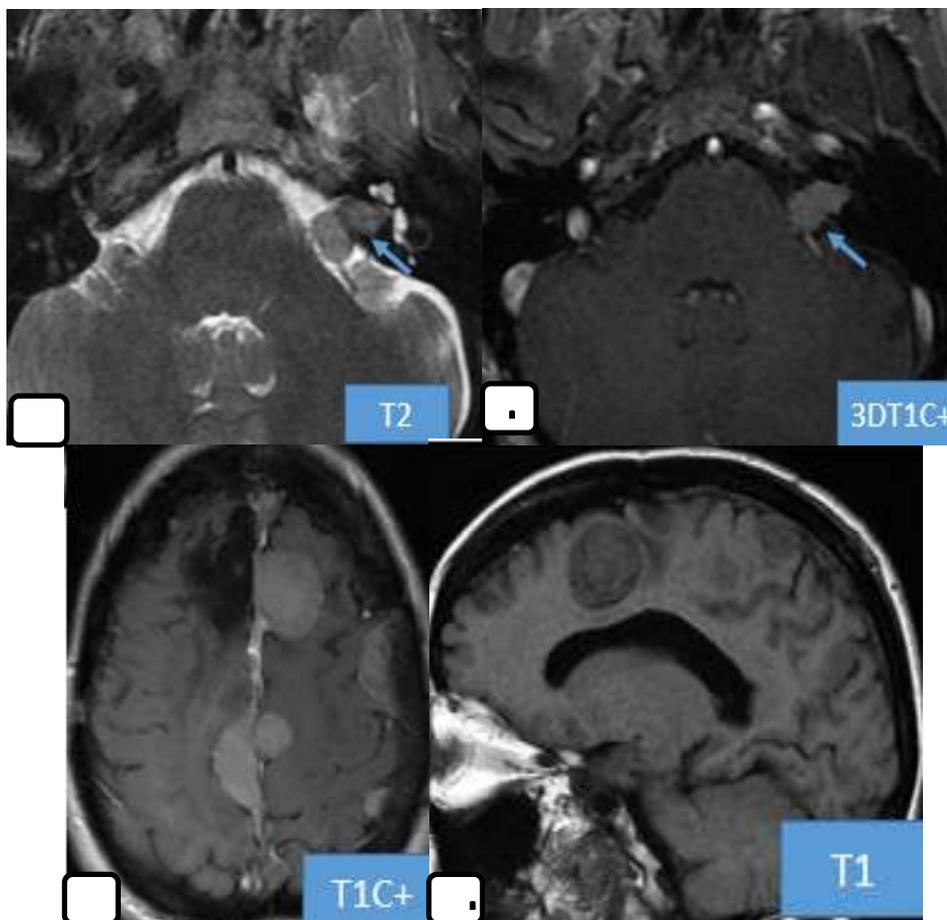


Figure 2 : MRI in axial section in T2 sequence (a); in axial section in injected T1 sequence (b,c) and in sagittal section in T1 sequence: left vestibular schwannoma in discrete T2 hypersignal, enhanced after contrast (blue arrow) associated with multiple meningiomas in the setting of neurofibromatosis type 2

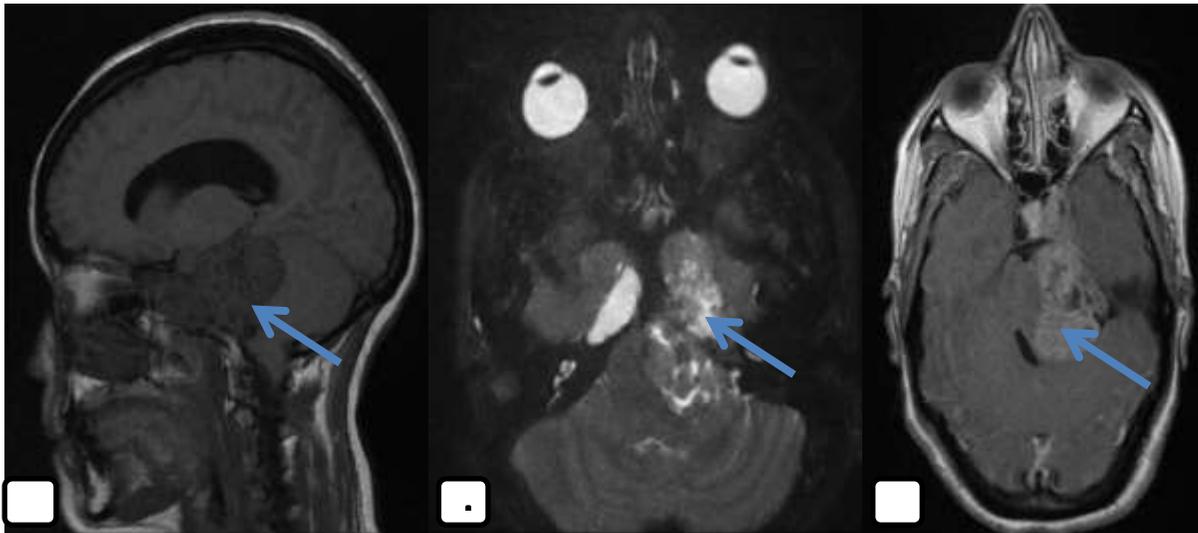


Figure 3 : MRI in sagittal section in T1 sequence (a) in axial section in T2 FS sequence (b); in axial section in injected T1 sequence(c): Extra axial expansive lesional process centered on the APC and in left peri-puncture extended to the left latero-sellar region, poorly limited, delineating areas of central liquefaction, the solid area is described in hyposignal T1, moderate hyposignal T2, enhances intensely and heterogeneously after contrast. It invades the IAC, and the homolateral cavernous sinus, backs up the TC with onset of tonsillar involvement, compresses the V3 → **Trigeminal schwannoma.**

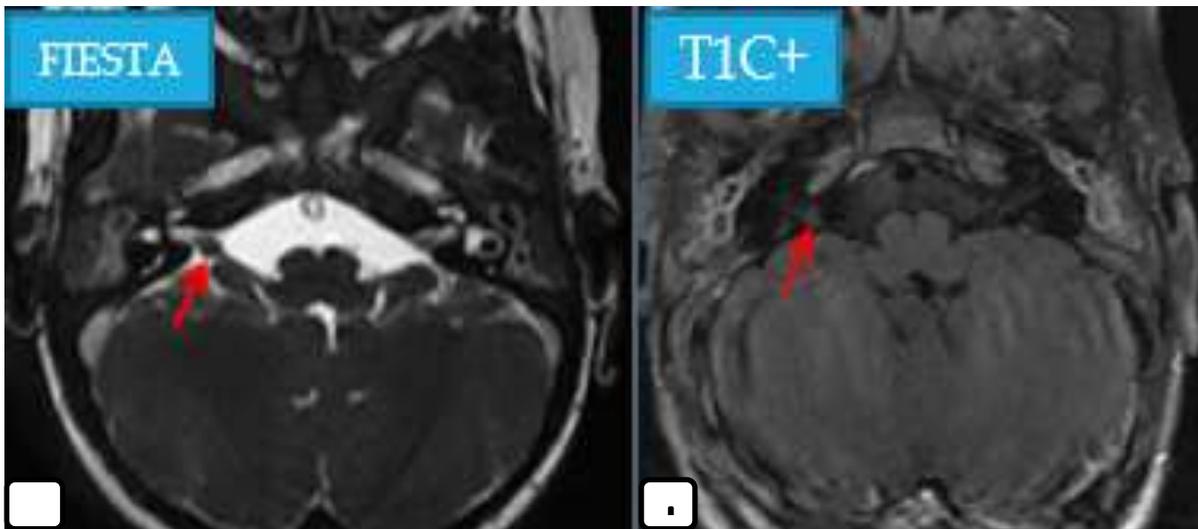


Figure 4 : MRI in axial section in FIESTA sequence (a) and in axial section in injected T1 sequence (b): Fusiform tumefaction of the right facial nerve in its cisternal course (superiorly and anteriorly) iso-signal T2, poorly enhanced after contrast. → **Schwannoma of the facial nerve**

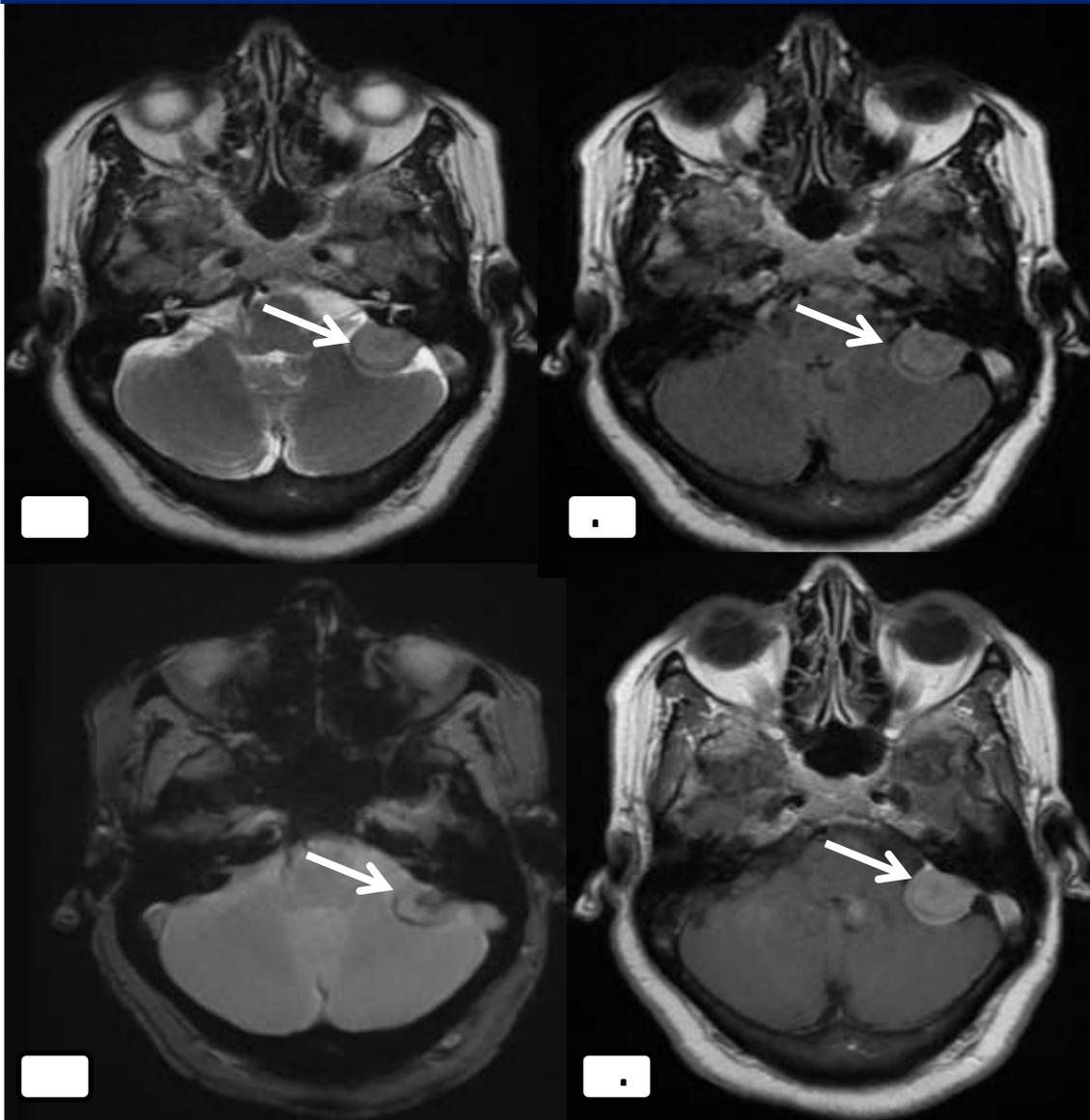


Figure 5: MRI in axial section in T2 sequences (a); in FLAIR sequence (b); in T2* sequence (c) and in injected T1 sequence(d): Extra-axial lesional process flush with the left CPA, broad-based implantation, well circumscribed, discrete hyper T2 and FLAIR, homogeneously enhanced intense and early after contrast, surrounded by a collar in hypo T2* signal related to calcifications, measuring 20x21mm. It exerts a discrete mass effect on the homolateral cerebellar parenchyma and determines a contact with the sigmoid sinus without invasion of the latter →**Meningioma**

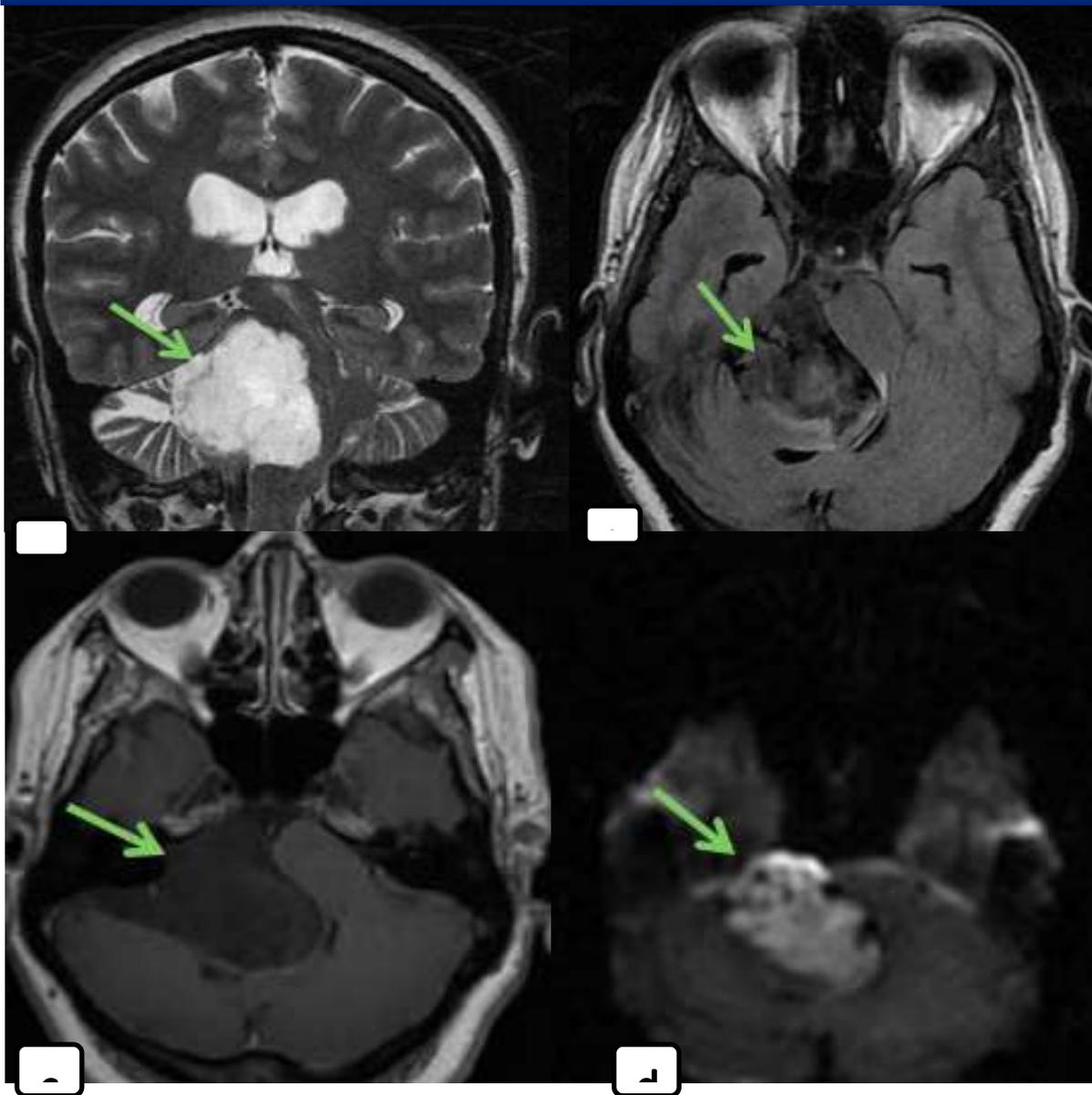


Figure 6 : MRI in coronal section in T2 sequences (a); in axial section in FLAIR sequence (b); in axial section in T1 injected sequence (c) and in Diffusion sequence (d): Lobulated mass, extra axial, occupying the right cerebellar ponto angle, hypointense in T1, hyperintense in T2 with diffusion restriction and not showing enhancement after contrast agent injection. Repression and compression of the brainstem, right cerebellum and fourth ventricle with moderate passive upstream hydrocephalus. →**Epidermoid cyst**

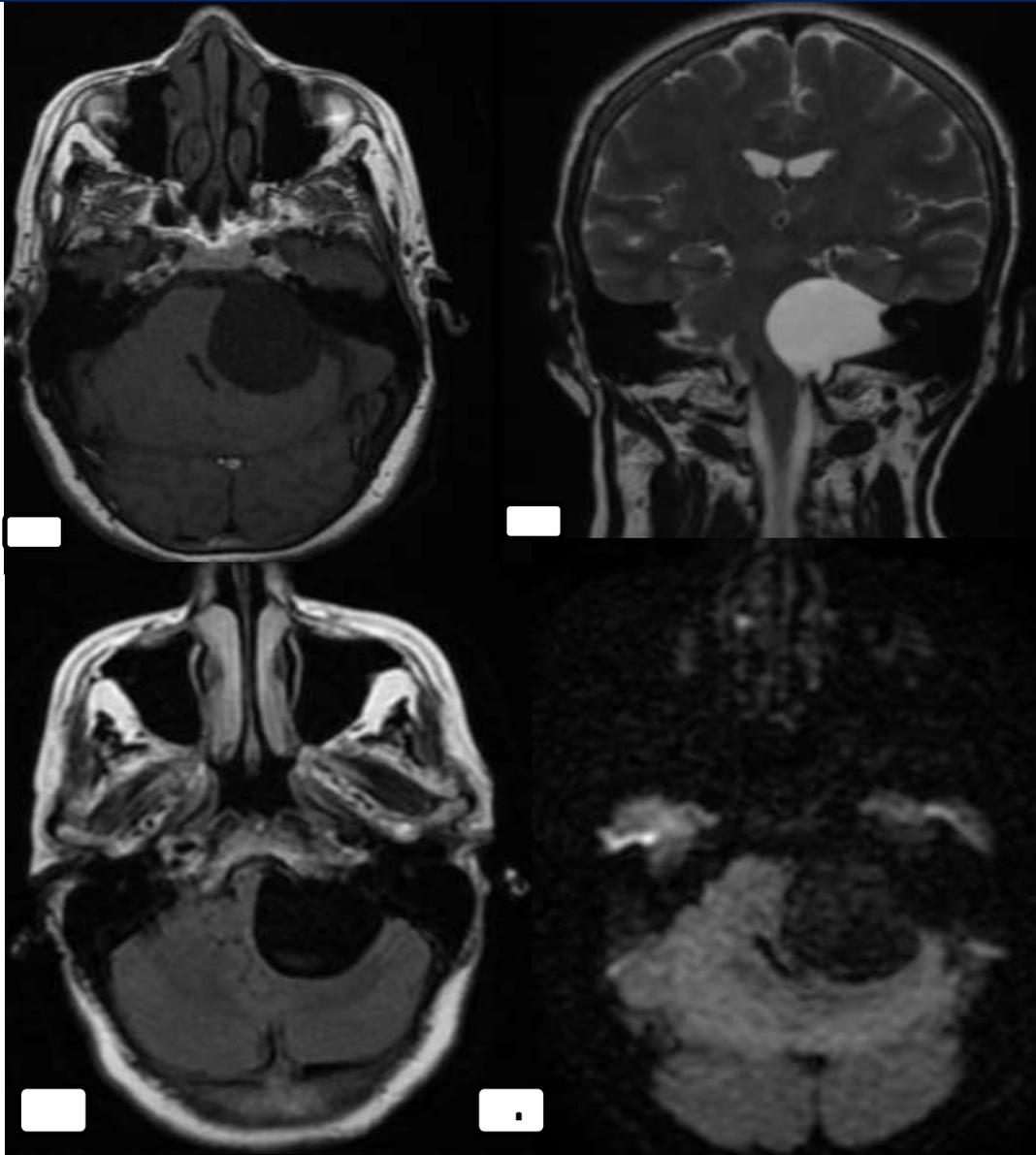


Figure 7 : MRI in axial section in T1 sequences (a); in coronal section in T2 sequence (b); in axial section in FLAIR sequence (c) and in Diffusion sequence(d); Extra-axial cystic formation at the level of the left cerebellopontine angle, with a liquid signal that cancels out on the Flair sequence, not restrictive in diffusion.

This formation pushes the cerebellar parenchyma, the V4, the protuberance and the medulla oblongata towards the right side, responsible for a descent of the cerebellar tonsils. → **Arachnoid cyst**

CONFLICTS OF INTEREST :

The authors declare no conflict of interest

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