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Cavernous Hemangioma Of The Mastoid: A Case Presentation Of An Exceptional Localization

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Abstract:
Background: *Intraosseous cavernous hemangioma is exceptionally located in the mastoid. This benign tumor, which arises from the intrinsic vascularization of the bone, develops slowly over many years and remains generally asymptomatic.*
Case presentation: *A 53-years-old female patient presented to the out-patient clinic of ENT-HNS department of Avicenna Military hospital of Marrakesh, with 10 years history of slow growing retro-auricular mass with no other complains. Clinical examination revealed a hard and painless mass located on the right mastoid process , fixed to deep mastoid fascia. Temporal bone CT scan shows a Honeycomb-shaped osteolytic lesion with surrounding needle-shape irregularities and microcalcifications in the temporal bone , extending to the mastoid and homolateral occipital bone and disrupting the medial and lateral cortex.*
Skull base MRI shows a hypointense T1, hyperintense T2 retroauricular expansive lesion that moderately and heterogeneously enhanced after injection of contrast medium, measuring 4.3 X 2.6 cm in anteroposterior and transverse diameter respectively.Biopsy reveals a cavernous hemangioma.
Radiological diagnosis, based on CT and MRI scans, is difficult and sometimes misleading.
Anatomopathological examination confirms the diagnosis, showing vascular congestions surrounded by fibrous tissue.
Discussion and conclusion:
Cavernous hemangioma is a rare condition which mainly occurs in nasal fossa.Diagnosis is suspected on imaging and confirmed by biopsy.Treatment is indicated only for symptomatic forms, and is based on mastoidectomy or even petrosectomy. Radiotherapy is an alternative treatment for non-operable forms. It can halt the progression of the tumor without causing a regression of the tumour volume.
Keywords: *intraosseous cavernous hemangioma, mastoid, mastoidectomy, external radiotherapy.*

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I. Introduction:

Bone hemangiomas are benign hamartomatous malformations . They represent 0.7 to 1% of primary bone tumors and 2% of benign bone tumors.They mainly affect the spine and craniofacial region and are mainly found in adults during the fourth and fifth decades of life [11].

Hemangiomas can be histologically divided into 3 types: cavernous, capillary and mixed. Most cranial hemangiomas are cavernous . Hemangiomas of the spine, on the other hand, are generally of the capillary type [11].

Thus far, around 100 cases of cavernous cranial hemangioma (CCH)of the skull have been reported. Most of them occur in the frontal bone [11]. CCH in the temporal bone is less common.
To our knowledge, this is the first case of the cavernous hemangioma in the mastoid bone to date

II. Case Presentation

A 53-year-old female patient ,whose father died of a hematologic malignancy and whose mother died of an undocumented bone cancer, presented to the ENT out-patient clinic of Avicenna Military Hospital of Marrakech, with a 10 years history of a painless slow growing retroauricular mass and no other associated symptoms.The patient's general condition remained preserved .

The clinical examination revealed a hard and painless mass located on the right mastoid process , fixed to deep mastoid fascia, measuring 4cm in long axis [Figure 1].Otoscopy was unremarkable, and there was no facial palsy.

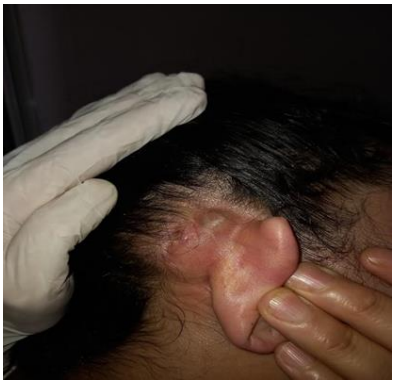

Figure 1 : right retro auricular mass

Temporal Bone CT scan shows Honeycomb-shaped osteolytic lesion with surrounding needle-shape irregularities and microcalcifications in the temporal bone , extending to the mastoid and homolateral occipital bone and eroding the medial and lateral cortex [Figure 2].

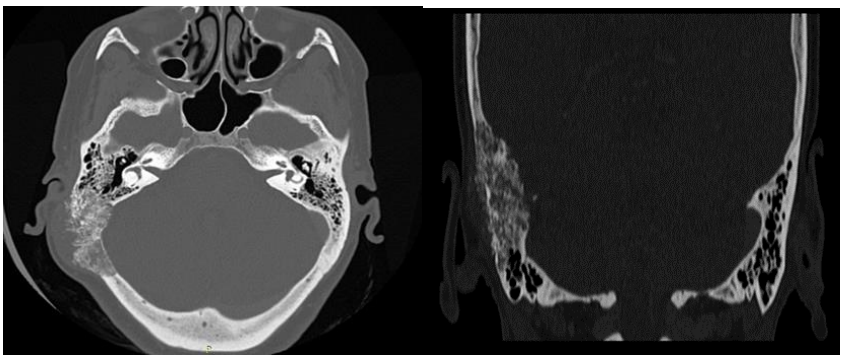

figure 2 :Bone window CT scan of the rock in axial (A) and coronal (B) objective sections Honeycomb-shaped osteolytic lesion with surrounding needle-shape irregularities and micro-calcifications on the temporal bone scale, extending to the mastoid and homolateral occipital scale and disrupting the internal and external cortex.

Skull base MRI shows a right retroauricular expansive lesion centered on the mastoid and occipital bone, lobulated, poorly limited, with irregular contours, hypointense on T1-weighted images and markedly hyperintense on T2-weighted, with diffusion restriction in the periphery, moderated and heterogeneous contrast enhancement , measuring 4.3 X 2.6 cm in anteroposterior and transverse diameter respectively [Figure 3-5].It infiltrates the occipital diploe posteriorly,the sigmoid sinus posteriorly and displaces the transverse sinus.It remains at a distance from the labyrinth and internal auditory canal.

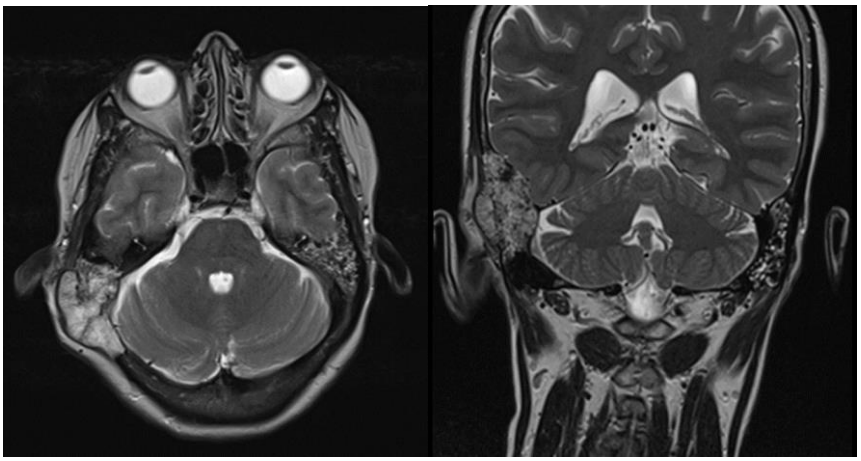

Figure 3: Skull base MRI in axial (A) and coronal (B) view. Presence of a right retro auricular expansive lesion process, centered on the mastoid and occipital bones, lobulated, poorly limited, with irregular contours heterogeneous T2 hyper signal

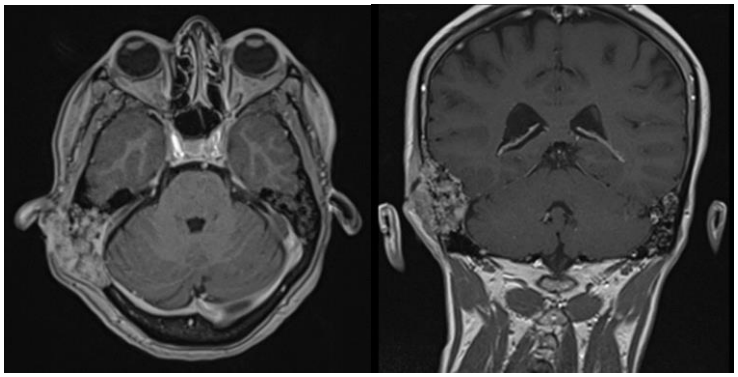

Figure 4: shows moderate and heterogeneous enhancement after contrast injection

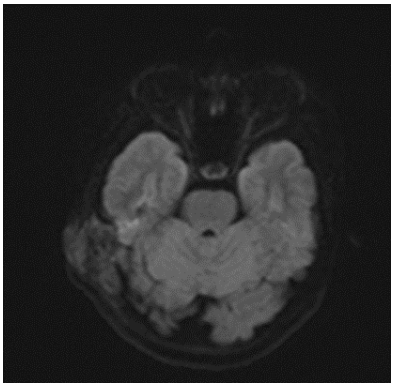

Figure 3 : Diffusion restriction in the periphery of the tumor

A bone biopsy was performed under general anesthesia.During the biopsy, severe and unexpected hemorrhage occurred immediately after the incision of the tumor that resolved with bone wax.

A histopathologic examination of the biopsy shows a bone tissue with trabeculae dissociated by fibrous remodeling associated to vascular dilation, suitable with an intraosseous cavernous hemangioma [Figure 6].

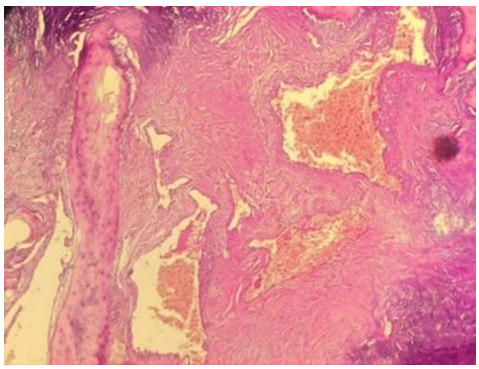

figure 4 : microscopic examination reveals bone tissue with trabeculae dissociated by fibrous remodelling, associated with large vascular structures and red cell-filled lumens (HESx100)

Given the fact that the patient was asymptomatic, the tumor board therapeutic decision was favourable to a clinic and radiologic monitoring every 6 months. Radiation therapy will be used in case of rapidely growing tumor, bleeding or impact symptoms.

III. Discussion:

Cavernous hemangioma, also known as "cavernous angioma" or "cavernoma", is a benign vascular tumor [1,2] with a venous component rarely found in an intraosseous location. Histologically, it isn't different from intra-parenchymal cavernous hemangioma. No link between these two entities has been described in the litterature, and there are no reported cases of dual localization (intra-parenchymal and intra-osseous) in the same patient. Intraosseous cavernous hemangiomas represent 0.7% to 1% of general bone tumors [1-3], and are

preferentially located in the spine [1,2]. Cranial cavernous bone hemangiomas are even rarer, representing just 0.2% of benign skull tumors [4]. They affect most frequently the parietal and frontal bones [1-3]. Mastoid localization is in fact exceptional.

This condition is most common in women, and almost always in young adults [1,2].

It is most frequently discovered accidentaly on radiological examination, or during a clinical examination, with the appearance of a hard, painless swelling covered by healthy, non-adherent skin.

It may be totally asymptomatic, and can sometimes occur with otological manifestations such as hypoacusis, otalgia, ear fullness or peripheral facial palsy.

On otoscopy, the External Auditory canal (EAC) may be normal, and can sometimes have a prominent bulge on it's posterior wall [11].

On imaging, a temporal bone CT scan confirms the localization of the lesion (cortical, medullary, juxta-cortical) in the form of a honeycomb osteolytic lesion with surrounding needle-shape irregularities and micro-calcifications on the mastoid.Temporal bone CT scan shows also the presence of thick and calcic density trabeculations with contrast enhancement , differentiating it from fibrous dysplasia, which does not enhance after contrast [11-14].

Skull base MRI usually shows an isointense lesion on T1-weighted imaging and a hyperintense lesion on T2-weighted imaging [3], which enhances after injection of gadolinium (Fig.) [1]. The tumor's classically heterogeneous appearance is due to the presence of hemosiderin, methemoglobin and oxyhemoglobin secondary to intra-tumoral hemorrhage. Blood flow is very slow [8].

Imaging features are not always specific.Differential diagnoses are numerous such as: osteosarcoma, chondrosarcoma, metastases, multiple myeloma, lymphoma, meningioma, osteoma, Paget's disease, cholesteatoma, eosinophilic granuloma and epidermoid cyst [1,3]. The diagnosis can only be made by anatomopathology which shows vessels with a highly dilated venous component, thin-walled and surrounded by fibrous connective tissue [4]. The pathology appearance of cavernous hemangiomas is identical despite the site of its development (intra- or extraosseous) [5].The tumor usually evovles very slowly **.**

Possible complications include peripheral facial palsy, conductive hearing loss due to invasion of the EAC [11] or the tympanic ossicular chain, peripheral vestibular syndrome due to damage of the inner ear, and damage of the sigmoid sinus .Episodes of painful intra-tumoral haemorrhage may also occur.

Therapeutically, asymptomatic forms usually require clinical and scannographic monitoring every 6 months. For symptomatic forms, the main treatment is surgical excision whenever it is feasible .This includes mastoidectomy , subtotal or total petrosectomy, depending on the volume of the tumour[11]**..**

Preoperative embolization can help prevent heavy bleeding in the case of surgery on large tumors [2].

Post-operative follow-up is generally uneventful, although some articles report cases of recurrence of cavernous hemangiomas in the sinus or orbit after surgery. [15,16]

Radiotherapy is a suitable alternative treatment . it halts tumor progression without reducing the tumor size [2]. It is therefore reserved for inoperable tumours with a high risk of facial palsy.

IV. Conclusion:

We believe this is the first case of a mastoid localization of cavernous hemangioma . This condition is difficult to diagnose. Imaging shows a non-specific osteolytic lesion that may mistakenly appear to be malignant neoplasia.Pathological examination confirms the diagnosis, showing vascular congestions surrounded by fibrous tissue.This tumor exhibits a very slow growing rate over many years.Several therapeutic choices can be choosen: Follow up, surgery after embolization or radiation therapy.

Abreviations:

CT: Computerized tomography
MRI: Magnetic Resonance Imaging
EAC: External Auditory canal
CCH: Cavernous cranial hemangioma

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