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CORRESPONDENCE

Hypertrophic Arachnoid Granulation of the Occipital Bone

Neuroradiological Differential Diagnosis

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Background

Nodules of arachnoid cells absorb cerebrospinal fluid (CSF) and drain it into the venous system [1], are microscopically observable and referred to as arachnoid villi. Those macroscopically visible are called pacchionian granulations (PGs) [2]. These usually protrude into venous sinuses, as demonstrated by autopsy studies [3]. The outermost layer is composed of endothelial cells, while the underlying layer is composed of arachnoid cells and fibroblasts [4]. They are evident from the age of 4 years, developing in proximity to venous sinuses or bridging veins near the superior sagittal sinus, within 3 cm of the midline. They may increase in numbers and dimension with age, probably because of CSF pressure [1] and can fill and dilate the dural sinus or expand the inner table of the skull [5, 6]. A differential diagnosis from osteolytic bone diseases is often difficult. While computed tomography (CT) scanning remains the most appropriate neuroradiological method to investigate bone diseases, magnetic resonance imaging (MRI) is a more suitable for evaluating these particular conditions.

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Case Report

A 64-year-old previously asymptomatic male patient, was referred to this department because of progressive bilateral hypoacusia, dizziness and tinnitus associated with a sensation of fullness in both ears and slight left-sided otalgia. The results of the neurological examination were normal but otoscopic examination showed a phlogistic process of the left tympanic membrane. An audiometric examination showed a right-sided perceptive and a left-sided mixed hypoacusia. A skull X-ray examination was performed to rule out chronic otitis media and revealed an osteolytic lesion involving the suboccipital bone in the midline region (Fig. 1). A subsequent CT scan study revealed an osteolytic mass lesion, involving both inner and outer bone layers (Fig. 2a, b), without contrast enhancement (Fig. 2c). The MRI examination showed a hypointense lesion in T1-weighted images (Fig. 3a), hyperintense in T2-weighted image (Fig. 3b) with a subtle capsule around the osteolytic lesion visible after gadolinium contrast enhancement (Fig. 4) which was recognized as a giant PG. No irregularity of the occipital bone profile was observable on local examination. Because of the incidental diagnosis and the absence of related symptoms, surgery was not considered. The patient underwent treatment with antibiotics for the otitis and showed prompt improvement of symptoms, but persistence of the hypoacusia. After a 3-year follow-up the results of the neurological examination were unchanged and no occipital bone profile modifications were perceptible.

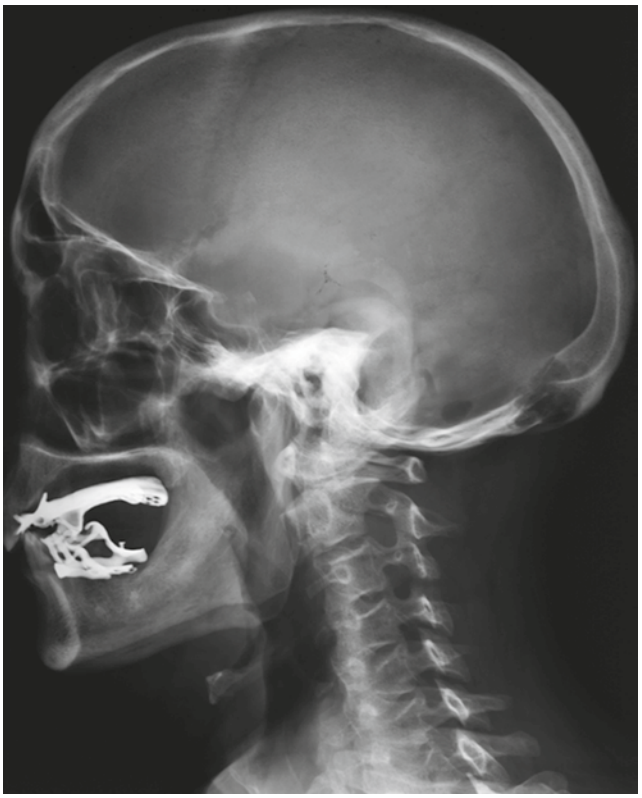

Fig. 1 Skull and cervical X-ray showing an osteolytic lesion of the occipital bone without apparent sclerotic margins

Discussion

Development, Localization and Clinical Findings

Macroscopically evident PGs are considered as anatomic variants of the more common microscopic arachnoid villi [2]. Early twentieth century neurophysiologists already suggested that PGs were exaggerations of the much smaller arachnoid villi found to be prevalent in animals and proposed that arachnoid granulations develop from arachnoid villi [7]**.** A study by Kida et al. [8] on human arachnoid granulations demonstrated that a large part is made up of a central core contiguous with the subarachnoid space and is composed of arachnoid cells and fibroblasts. An arachnoid cell layer of the granulation is continuous with the underlying arachnoid membrane and a fibrous capsule, reflected from the surrounding dura mater, covers the arachnoid cell layer except at the apical portion of the granulation and finally, an arachnoid cap cell layer covers the apical portion of the granulation and directly contacts the venous lumen [7, 8].

The presence of a cell layer which contacts both the CSF and venous blood, along with the location of the arachnoid cap cells at the apical portion of the granulation, suggests the possibility of a specialized functional role for these cells in the outflow of CSF in humans. Perfusion studies suggest that the ultrastructure of AGs can accurately replicate the unidirectional flow of CSF [9] and AGs are defined as giant when they fill the lumen of a dural sinus and cause local dilatation or filling defects [10, 11], with subsequent increase in intracranial pressure [12]. Some authors have suggested that giant AGs might be responsible for pseudotumor cerebri syndrome by obstructing the sinusal venous flow [13, 14]. Moreover, because they are usually located within 3 cm of the midline at the entry of the cortical veins into the sagittal sinus [3, 4] where there is a weakness of the dura mater, they could be regarded as arachnoid herniations secondary to intracranial CSF pulsation through dural defects into the sinus [15]. However, giant AGs have also been reported distant from the midline particularly in relation with the transverse sinus [5, 6, 16]. Greitz et al. proposed that AGs are the principal site of CSF re-absorption, considering them as Starling resistors in order to prevent cortical venous collapse during variations in intracranial pressure [17]. Krish suggested a volume buffering function of the intracranial CSF compartment, where AGs replace the fontanelle as a rapid volume buffering structure after its closure [18]. According to these theories, giant AGs could be the result as well as the cause of intracranial hypertension [15]. In contrast posterior or

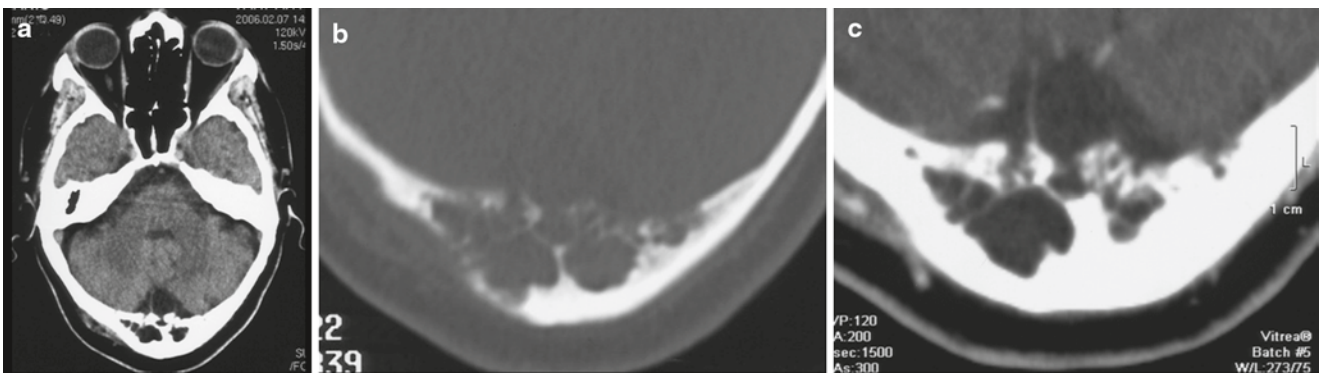

Fig. 2 Axial CT scan shows a hypodense osteolytic lesion of the occipital protuberance (**a**); bone window reconstruction highlights the erosion of both tables of the calvaria due to a hypodense mass (**b**), after contrast administration no enhancement of the lesion in documented (**c**)

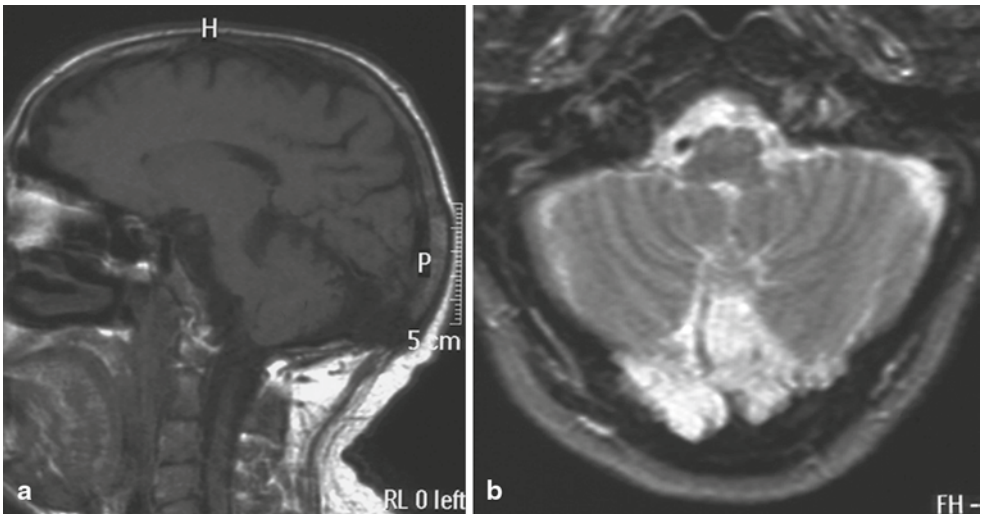

Fig. 3 Sagittal T1-weighted MRI shows the low signal intensity of the occipital lesion (**a**) but appears with high signal intensity, isointense to cerebrospinal fluid on axial T2-weighted MRI (**b**)

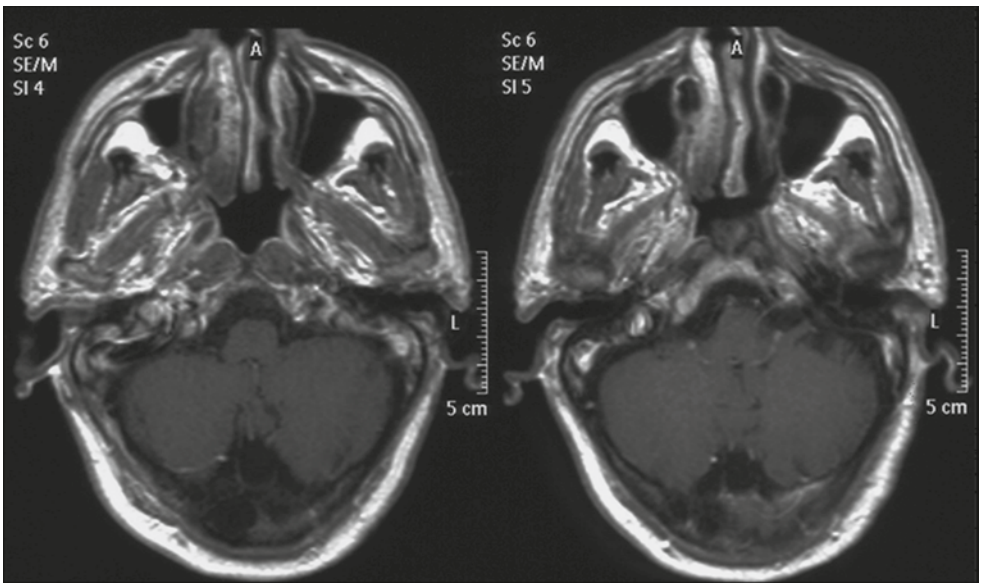

Fig. 4 Axial T1-weighted MR images with gadolinium show only a subtle enhancement of the lesion margins

middle cranial fossa AGs are neither connected with veins nor in relation to CSF absorption and their function remains unclear.

Neuroimaging and Differential Diagnosis

Besides the osteolytic aspect on X-ray imaging, in MRI AGs appear hypointense or isointense relative to the brain on T1-weighted and hyperintense on the T2-weighted images, variable in signal on proton-density weighted images and showing minimal heterogeneous contrast enhancement [16]. Relative to CSF all granulations are isointense on T2-weighted images, while almost all are isointense on T1-weighted images and in fluid attenuated inversion recovery (FLAIR) images 90.3% of the granulations are instead isointense and the remaining 9.7% are between CSF and grey matter [16].

In diffusion-weighted images (DWI) all AGs showed isointensity to normal brain tissue, which was higher than the reported signal intensity of arachnoid cysts and lower than that of epidermoids. [19, 20].

However, it may be difficult to differentiate AGs from dermoids, epidermoids, hemangiomas and other lesions presenting signal intensities similar to CSF, although dermoid and epidermoid cysts frequently involve both the inner and outer tables, hemangiomas and eosinophilic granulomas are located mainly in the intradiploic space and rarely involve the inner table [16]. In contrast AGs determine an impression on the inner table and only in few cases involve the outer table. As AGs can involve dural sinuses differential diagnosis should also include sinus thrombosis and intrasinus tumors [4, 16, 19, 20]. Nevertheless, AGs are not hyperintense on FLAIR images, although lesions usually hyperintense on T2-weighted are thought to be hyperintense

on FLAIR images with the exception of epidermoids. When suspected prominent AGs are noted, FLAIR images should help to differentiate granulations from the dural sinus or skull lesions, mainly from epidermoids [16, 19].

Finally, giant AGs should also be differentiated from circumscribed bony lesions of the skull, such as sinus pericranii and arachnoid herniation; however, osteodural leaks can be found at some specific sites of predilection along the ethmoid, midline sphenoid and lateral sphenoid sinuses [21]. For unknown reasons, arachnoid granulations can be seen on the floor of the anterior and middle cranial fossa and less frequently on the posterior temporal bone wall [22]. High-resolution CT imaging is required to recognize osteodural leaks; however, MR imaging supplements the CT examination with respect to recognition of the presence of arachnoid herniated tissue and differentiation of its contents. These granulations are usually easily visualized with MR angiography and they appear as rounded or elliptical areas with no signal. The elliptical defects may simulate a thrombus, but a review of T2-weighted images usually shows their true nature [20].

Management

Giant PGs are usually asymptomatic and discovered as incidental findings, although they can give rise to otorrhea, rinorrhea or otitis media [5]. The finding of giant PGs has often been associated with pseudotumor cerebri and benign intracranial hypertension syndrome with headache, vertigo and blurred vision associated to papilledema [13]. Giant AGs can sometimes cause osteolytic phenomena and subperiosteal bleeding [21, 22].

Neuroradiological assessment should include a CT scan in order to assess the presence of bone erosion, MRI images (including FLAIR sequences) in order to rule out other similar findings, such as dermoids and epidermoids and MRI angiography to determine the anatomic relationship with venous vascular structures. More invasive studies, including standard venous angiograms should be performed only if a major venous filling defect is suspected by MRI angiography and for preoperative studies if necessary.

Not enough details are known concerning the nature of the related headache and no long-term follow-up studies which can help to discern if symptoms are related to radiological findings have been performed. Dural sinus pressure measurement across the lesion is a novel and valuable approach to determine if they really cause venous obstruction and hypertension: a normal venous pressure without significant differential pressure across the lesion can exclude them as being the cause of symptoms [10]. Surgery should be considered exclusively in cases of symptomatic AGs or when a neoplastic disease cannot be ruled out neuroradiologically. During surgery much CSF can flow out from the subarachnoid space when AGs are excised so that careful dural plasty and/or osteoplasty is therefore necessary [6].

Conclusion

Cases of PGs have been reported in which differentiation from other bone lesions was difficult and most of them involve the midline or the cranial base. However, hypertrophic PGs should be included in the differential diagnosis of all osteolytic cranial lesions whether localized near the midline or not, either in the convexity or cranial base. They are not neoplastic lesions and surgery should be considered only if important related symptoms are present.

Conflict of Interest Statement The authors declare that there is no current or potential conflict of interest in relation to this article.

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