Ossifying fibroma of the occipital bone—A case report and literature review

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Abstract

Ossifying fibroma (OF) is one of the benign fibro-osseous lesions (BFOLs) affecting the craniofacial bones. Involvement of the skull bones specifically the occipital bone is especially rare. We report a case of a 49-year-old adult male with incidental finding of an ossifying fibroma arising from the right occipital bone with transverse sinus involvement.

Keywords: Ossifying fibroma; Occipital bone; Benign fibro-osseous lesions; Computed tomography; Magnetic resonance imaging; Bone scan

1. Introduction

Ossifying fibroma (OF) is one of the benign fibro-osseous lesions (BFOLs) included in a heterogeneous group of bone disorders affecting the craniofacial bones [1]. In adults, it occurs most commonly in the mandible, followed by the maxilla and rarely, the other sinonasal bones, orbit and skull bones.

A small number of case reports had been published on OF involving the calvarium. Based on Medline search, there were 19 cases of calvarial OF that had been reported to date, of which only two of these cases described OF arising in the occipital bone which occurred in young patients.

Here, we present an incidental rare case of an adult with ossifying fibroma arising from the right occipital bone. Literature review of this condition was also discussed.

2. Case report

A 49-year-old Chinese man with no prior medical illness, presented with 2-year history of progressive weakness of both hands and numbness of both fifth fingers. There were no central neurological or visual symptoms.

His neurological examination revealed weakness in gripping, wasting of hypothenar and thenar muscles and peripheral neuropathy bilaterally. The nerve conduction tests demonstrated bilateral ulnar nerve palsies.

MRI (Siemens Magnetom Vision 1.5 T) of the brain and cervical spine were initially done to exclude central cord syndrome, and a posterior fossa lesion was incidentally found. MRI of the brain showed a well-circumscribed extra-axial mass measuring 4.7 cm × 3.1 cm × 3.6 cm which appeared to be arising from the diploic space of the right occipital bone with destruction of the inner and outer tables. On the spin-echo T1-weighted (TR = 735 ms, TE = 14 ms) image, a thin hypointense rim is seen, mainly at the outer aspect of this mass, probably representing the periosteum (Fig. 1A). This lesion appears hypointense on T1-weighted images with clustered areas of signal void on T1- and T2-weighted images representing areas of calcifications or ossifications. On spin-echo T2-weighted (TR = 3800 ms, TE = 90 ms) images, hyperintensity is seen around the area of calcifications and there is loss of signal at the anterior part of the mass (Fig. 1B). This mass showed enhancement on the post-gadolinium (Magnevist) scan (Fig. 1C). The lesion appears hypointense on T1-weighted images with clustered areas of signal void on T1- and T2-weighted images representing areas of calcifications or ossifications. On spin-echo T2-weighted (TR = 3800 ms, TE = 90 ms) images, hyperintensity is seen around the area of calcifications and there is loss of signal at the anterior part of the mass (Fig. 1B). This mass showed enhancement on the post-gadolinium (Magnevist) scan (Fig. 1C). The MRI of the cervical spine showed a linear focal area of low signal intensity on T1-weighted image, high signal intensity on T2-weighted image which did not enhance post-gadolinium in the spinal cord at C6 level, representing syringomyelia.
Fig. 1. MRI brain (axial T1-weighted (A), T2-weighted (B) and T1-weighted post-gadolinium (C) images). T1-weighted image showed a well-circumscribed extra-axial mass arising from the diploic space of the right occipital bone with destruction of the inner and outer tables. A thin hypointense rim (arrow) is seen, mainly at the outer aspect of this mass, probably representing the periosteum. The clustered areas of signal void seen on T1- and T2-weighted images represent areas of calcifications or ossifications. Post-gadolinium image showed heterogeneous enhancement of this mass.

A radionuclide scintigraphy bone scan demonstrated a well-circumscribed region of homogenously intense radionuclide uptake at the right occipital bone (Fig. 2). No other areas of increased radionuclide uptake noted in the skull.

A CT venography was done to assess transverse sinus involvement. The lesion appears to be located in the diploe with expansion and enhancing soft tissue component extending into the posterior fossa region. There was destruction of the inner and outer tables with a thin rim of cortex seen. There were also areas of ground glass appearance with ossifications seen within (Fig. 3). There appeared to be compression of the right transverse sinus but the involvement of the sinus could not be ascertained.

Cerebral and right external carotid angiography showed a tumour blush at the right occipital region with arterial supply from the right occipital artery and the meningeal branch of the right occipital artery. On the venous phase, there was attachment of the tumour to the right transverse sinus. The provisional diagnosis of intraosseous meningioma was made by the neurosurgical team, hence, pre-operative cerebral angiogram was arranged and embolization of the tumour was done to reduce the risk of catastrophic hemorrhage during the surgery.

He subsequently underwent right occipital craniectomy and excision of the tumour. This was followed by cranioplasty with titanium mesh to cover the cranial defect. Intra-operatively, the overlying periosteum was still intact. The tumour appeared vascular, soft, friable and yellowish in colour with no evidence of pseudo-capsule formation. The tumour was found to adhere densely to the underlying transverse sinus. Excessive bleeding was encountered during the surgery in attempting to remove this tumour. Thus, thin layer of the tumour on the transverse sinus surface was left behind. Post-operatively, he still has weakness of both hands but no other post-operative complications encountered. He was discharged well on post-operative day 3.

Histopathological examination of the bone fragments showed trabeculae of calcified osteoid with varying width. The intervening stroma consists of densely packed plump spindle-shaped cells displaying elongated basophilic nuclei. No cytological atypia or mitosis detected and no malignancy is seen (Fig. 4). These histopathological findings were of a benign tumour which was suggestive of ossifying fibroma.
Ossifying fibroma is one of the benign fibro-osseous lesions involving the craniofacial bones. Involvement of the skull bones specifically the occipital bone is especially rare.

Based on literature review, the most common site is the mandible accounting for 75–89% of cases (2), followed by the maxilla, less commonly, the paranasal sinuses and orbit. Rarely, the involvement of the calvarium, i.e. frontal, temporal, parietal and sphenoid bones have been reported [4–13]. Of the calvarial involvement, frontal bone is the commonest site but these were usually associated with frontal, ethmoid or sphenoid sinus involvement or were located supraorbitally [3,6]. To the best of our knowledge, only two cases of OF arising from the occipital bone were reported. Yamashita et al. described a 9-year-old boy with OF in the occipital bone [14] and Binath et al. presented a 13-year-old girl with similar lesion in the occipital bone [15].

OF was first described by Menzel in 1872 [16] and the term ‘ossifying fibroma’ was subsequently coined by Montgomery in 1927 [17]. This lesion was then thought to be a histological variant of fibrous dysplasia, but was later separated into a distinct clinicopathological entity.

The term BFOL was subsequently used in the literature to describe a spectrum of lesions ranging from fibrous dysplasia to ossifying fibroma including cementifying or cemento-ossifying fibroma, psammomatoid ossifying fibroma, psammoosteoid fibroma, juvenile or young ossifying fibroma, and juvenile active ossifying fibroma. Based on the review article of current concepts of BFOL by Brannon and Fowler in 2001 [1], the categorization of these lesions has been suggested, comprises osseous dysplasia, fibro-osseous neoplasms (including the conventional OF and juvenile OF) and fibrous dysplasia. Common to all BFOL is the replacement of normal bone with a tissue composed of collagen fibers and fibroblasts that contain varying amounts of mineralized substance, which may be bony or cementum-like in appearance.

The aetiology and pathogenesis of OF is still unknown. The proposed theories of trauma and developmental abnormality as a cause of OF had been suggested [6], however, in the more recent case reports from year 2000 onwards, the pathogenesis of this tumour had not been discussed.

The peak incidence of OF is in the second and third decades of life with slight preponderance in female [3,13,15,18]. However, no sex predilection has been reported by other authors [15]. Clinical presentation of these tumours is variable, depending on the site and rate of growth. When encountered in the calvarium, the presentation is typically with bone expansion, usually painless. However, several case reports [7–10] described a symptomatic patient with painful mass at the site of the lesion. Midline skull base lesions, may present with symptoms due to mass effect, such as nasal obstruction, headache or anosmia or ocular symptoms include visual loss, diplopia, and exophthalmos. Symptoms of ear discomfort or pain, pulsatile tinnitus, otorrhoea and progressive hearing loss may be the clinical presentation if the lesion involves the temporal bone [12]. Rarer presentations include meningitis or pneumocephalus had also been reported [2]. It may also be asymptomatic and detected incidentally, as in this case.

There have been no reports of malignant transformation of this benign bone tumour, although some case reports describe local aggressiveness with extensive, rapid growth that may mimic malignancy and associated with neurological signs due to an intracranial mass effect [6]. Recurrences have been described in multiple cases [18] and a review of the literature reported a 39% recurrence rate in 61 cases [3]. Partial excision can lead to recurrence [6,15,19].

Radiographic features of OF vary from case to case. Most authors described the usual radiological characteristics of OF as a well-defined lesion, well-circumscribed by the thin shell of bone that the mass displaces outward [3,6,15,19]. On radiographs, the initial lesion is radiolucent, but it progressively becomes radio-opaque, surrounded by uniformly radiolucent periphery or occasionally by peripheral sclerosis [15]. However, there are few reports that described the lesion as poorly circumscribed or with irregular borders and partially radiolucent (osteolytic) or sclerotic (osteoblastic) [6–9].

On CT, OF are typically seen as a well-circumscribed lesion, similar to other benign osseous lesions. Almost all the cases described the lesion located in the diploic space with moderate expansion. The central areas consist of a non-homogeneous matrix, which includes ground-glass opacification representing diffuse calcifications and low attenuation areas containing fibrous tissue or retained mucus (if situated adjacent to the paranasal sinuses). The walls of the involved sinuses may show remodeling and thickening, sometimes in combination with erosions. The fibrous part may enhance post-contrast. The pro-

Fig. 2. Radionuclide bone scan (lateral view) shows a well-circumscribed region of homogenously intense radionuclide uptake in the right occipital bone.
Fig. 3. CT venogram of the brain (axial (A), sagittal (B) and coronal (C) images in bone window) shows an expansile intradiploic lesion at the right occipital bone with enhancing soft tissue component. Destruction of the inner and outer tables with a thin rim of outer cortex is seen. There are areas of ground glass density and ossifications seen within.

Fig. 4. Photomicrographs of tumour specimen showing bony trabeculae rimmed by osteoblasts surrounded by plump spindle-shaped cells (arrow) [H&E stain, 100× (A), 200× (B) and 400× (C)] (Courtesy from Professor Jayalakshmi Pailoor, M Path, FRC Path, Professor and Consultant Histopathologist, Department of Pathology, University of Malaya).
portion of fibrous soft tissue and calcifications or ossifications is variable. Similarly, there were few reports describing the lesion to be partly osteoblastic and osteolytic. In addition to the mixed type of lesion, destruction of the inner and/or outer tables of the calvarium was also seen suggestive of a more aggressive process [6,15,19].

The typical radiological feature of OF seen in this patient was a well-defined expansile intradiploic mass with ground glass density and enhancing soft tissue component representing the fibrous portion. There was also destruction of the inner and outer tables of the occipital bone, however, evidence of intact periosseum and formation of ossified outer table as seen on CT and MR suggesting that this is a slow growing lesion with remodeling process.

Radionuclide scintigraphic bone scan typically shows well-circumscribed area of homogenously intense radionuclide uptake at the site of the lesion as illustrated in this case, suggestive of a lesion with intense osteoblastic activity. Other cases also revealed similar findings of this lesion on bone scans [6,7,9,19].

Based on four case reports of OF involving the skull bone, angiography revealed tumour blush and the vascular supply to the tumour was derived by extra-cerebral vessels [5–7,14]. In this case, the angiography showed the vascular supply to this tumour mass was derived from the extra-cerebral vessels, i.e. the right occipital artery and the meningeal branch of the right occipital artery. The involvement of the right transverse sinus was also seen on the venous phase of the angiogram which was confirmed intra-operatively.

The main differential diagnoses in this case are ossifying fibroma and fibrous dysplasia. However, the radiographic and CT features of ossifying fibroma, i.e. bone expansion, well-defined margin and ground glass appearance cannot be differentiated from those of fibrous dysplasia. There are no other reliable CT or MR criteria to distinguish between these two entities and thus can only be distinguished by histology.

Intraosseous meningioma was also initially considered as a differential diagnosis as its radiological appearance is of an intradiploic expansile lesion. It can present as an osteolytic lesion with cortical erosion (35%) or mixed picture of hyperostosis and osteolysis (9%), even though the most common radiographic finding is hyperostosis (59%) [20]. Furthermore, osteolytic meningiomas associated with soft tissue component should be considered “malignant until proven otherwise” [20]. However, the tomographic view of the radionuclide bone scan in this patient did not show the appearance of increase uptake with central photopenic area typical of an intraosseous meningioma, based on literature review [21]. The MRI appearance was also not suggestive of intraosseous meningioma as the lesion did not show intense and homogenous enhancement post-gadolinium.

In this case, the diagnosis was only made post-operatively. The treatment of OF is complete excision. Due to the dense adhesion to the underlying dura and transverse sinus involvement, complete excision was impossible and rendering the possibility of recurrence. Long term follow up in partially excised case is mandatory to detect recurrence and prevent long term morbidity as in this patient [6,15,19].

References