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A giant cranial aneurysmal bone cyst associated with fibrous dysplasia

J. Składzień, K. Oleś*, O. Zagólski*, M. Moskała**, M. Sztuka**, P. Stręk*, W. Wierzchowski*** and J. Tomik**

Departments of *Otorhinolaryngology, **Neurotraumatology and ***Pathomorphology, Collegium Medicum, Jagiellonian University, Kraków, Poland

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Abstract. *A giant cranial aneurysmal bone cyst associated with fibrous dysplasia.* An aneurysmal bone cyst (ABC) is a rare, benign fibro-osseous lesion, considered a vascular phenomenon secondary to fibrous dysplasia or a giant-cell tumour, and occurs mainly in long bones and vertebrae. In this case report a 16-year-old male presented with massive epistaxis. He was admitted with a 3-year history of chronic rhinitis, headaches, right ocular pain and recurrent epistaxis. CT scans showed a predominantly cystic, expansive mass obstructing both nasal cavities, extending to all paranasal sinuses and both orbits, with evidence of anterior cranial fossa skull base destruction. The patient underwent a craniofacial resection of the tumour performed with an external approach and an immediate reconstruction of the dural defect. Histology confirmed the lesion was an ABC associated with fibrous dysplasia. The patient's recovery was complete. A large facial aneurysmal bone cyst can damage the facial skeleton and skull base, and requires excision by a combined external approach.

Introduction

An aneurysmal bone cyst (ABC) is an uncommon, solitary, benign fibro-osseous lesion that occurs mainly in long bones and vertebrae.[1-5] ABCs represent approximately 1.4% of all bone tumours, and among those only 3% are located in the cranium,[4,5] where they most commonly affect the mandible[5] in the anterior cranial region.[6] Only a few cases of ABC have been described within the cranium that did not include the mandible; these tumours originated in the sphenoid [2,4,7,8] and maxillary sinus.[9] In addition, there have been a few reports of ABC growths in orbitoethmoid, ethmoid, frontal, and occipital locations.[3,6] Finally, only two reviewed cases have reported giant cranial tumours.[10,11]

The origin of an ABC tumour is considered a vascular phenomenon, although clonal chromosomal changes are sometimes involved.[2,3,12,13] It is currently thought that a pre-existing bone lesion, such as polyostotic fibrous dysplasia, or a giant-cell tumour, initiates an osseous arteriovenous fistula which then creates a secondary reactive lesion of bone - the ABC. However an ABC may also arise as a primary lesion.[2] The tumour typically replaces bone with a spongy fibro-osseous tissue, and then becomes a locally destructive, multicystic lesion filled with blood. The ABC has tendency to destroy and infiltrate bone and can reach a considerable size.[7]

There is no gender predilection in the occurrence of ABC,[14] however it occurs most commonly in patients under 20 years old, and it tends to enlarge rapidly after puberty.[6] The clinical presentation of the ABC lesion is similar to other sinonasal tumours; the symptoms depend on its localisation and extent.[2-4,7,10,15,16] An ABC is diagnosed when multiple cysts containing fluid are detected with computed tomography (CT) and/or magnetic resonance imaging (MRI).[3,7] MRI offers a diagnostic advantage because it is able to detect blood and fluids.[5] The final diagnosis is confirmed by distinct characteristics that are identified in a histological examination of the excised lesion.[3,11] The treatment is a surgical resection using an external approach in most cases.[7,14,17] Some physicians administer arterial embolization before surgery.[6,7] The endoscopic approach has also been used successfully for the treatment of small lesions.[7,14,17]

Case report

A 16-year-old male presented with massive epistaxis. On admission the patient reported a 3-year history of chronic rhinitis, headache, right ocular pain that limited lateral ocular mobility, epiphora and recurrent epistaxis. He had been treated by general practitioners with antiallergic drugs without confirmation of the

diagnosis. Physical examination revealed rhinorrhea, nasal congestion, and 4 mm exophthalmos in the right eye. The findings were consistent with a giant neoplasm. Serum calcium level was normal. The nasal endoscopy detected a tumour of medium consistency within a grey capsule of connective tissue that filled both nasal cavities. CT scans of the sinuses showed a lytic, predominantly cystic, expansive lesion. Fluid and blood-filled vesicles with inner septa were visible within the tumour. The tumour mass filled both nasal cavities, encroaching upon maxillary, ethmoid, frontal and sphenoid sinuses and both orbits, with evidence of skull base destruction in the anterior cranial fossa (Figures 1,2). The biopsy performed on the day of admission revealed an aneurysmal bone cyst.

The patient underwent a craniofacial tumour resection performed with an external approach. No preoperative arterial embolization was performed. A lateral rhinotomy was extended along the upper margin of the orbit and included a coronal incision (Figure 3). The tumour compressed the dura of anterior cranial fossa and was removed together with a 50×50 mm fragment of dura. The excised dura was immediately replaced with artificial dura. The tumour's diameter was approximately 70 mm. It also involved cilia olfactoria.

Histological analysis revealed the cyst walls composed of vascularized, proliferative soft tissue with a network of fibrous septa that contained osteoclast-like multinucleated giant cells and erythrocyte-filled spaces, within a layer of spindle-shaped endothelial cells. The bone surrounding the lesion consisted of immature, undifferentiated fibrous connective tissue and included a small amount of collagen fibres (Figure 4). The analysis was consistent with an ABC associated with polyostotic fibrous dysplasia.

The recovery was nearly complete after a week, though olfactory function was still absent. Ocular mobility rehabilitation was started on the second postoperative day. The patient reported improvements in all preoperative symptoms and was asymptomatic at 9 months follow-up.

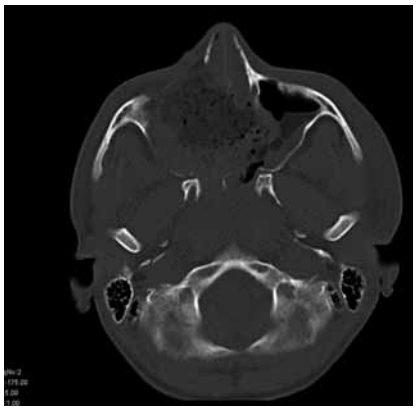

Figure 1
Preoperative computed tomography image of the paranasal sinuses – axial view. The tumour fills the right maxillary sinus, both nasal ducts, and penetrates through the left nasal lateral wall. Fluid and blood are visible in vesicles with inner septa characteristic of the aneurysmal bone cyst.

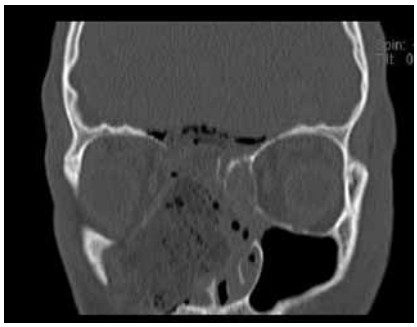

Figure 2
Computed tomography image of the paranasal sinuses – coronal view. The tumour penetrates through the right orbit and through the cribriform plate to the anterior cranial fossa. Both nasal ducts, the ethmoids, and the right maxillary sinus are filled with the mass.

Figure 3
Intraoperative image of the dissected tumour. The mass was removed en bloc through the facial incision.

Discussion

The majority of aneurysmal bone cysts reported to occur in cranial locations were small lesions and were removed by a variety of surgical procedures (Table 1). Here we report the third case of a giant cranial ABC. Saito *et al*.[10] presented a case of an 11-year-old boy with an ABC that extended into

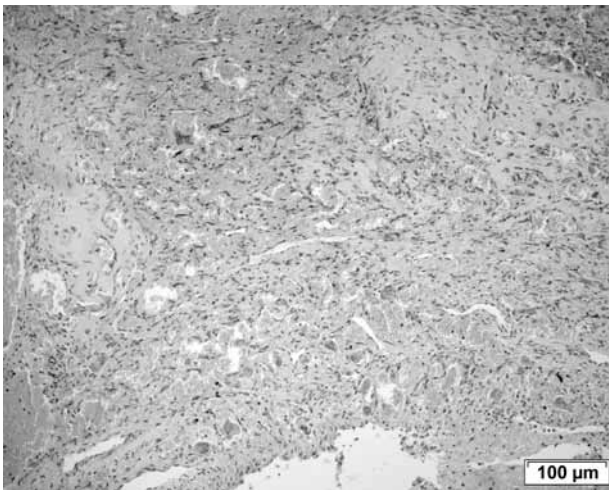

Figure 4
Aneurysmal bone cyst, H&E staining. Proliferative soft tissue is visible with focal osteoclast-like multinuclear giant cells and erythrocyte-filled spaces surrounded by endothelial cells. The bone tissue consists of immature, undifferentiated fibrous connective tissue and includes small amount of collagen fibres, characteristic of polyostotic fibrous dysplasia.

the nasal cavity, the sphenoid and posterior ethmoid sinuses, and the skull base, and involved the planum sphenoidale, the sella turcica, the upper two-thirds of the clivus, and the medial portion of the mid-cranial base. Nadkarni *et al.*[11] presented a 19-year-old male patient with an ABC that involved the anterior cranial fossa floor, occupied and enlarged all paranasal air sinuses, and caused outward displacement of the orbits and superior displacement of the frontal lobes of the brain.

Our patient had been improperly diagnosed and treated, without the necessary imaging required for corroboration in cases involving chronic nasal congestion, recurrent epistaxis, headache, and epiphora.[3,7,11,15] His age predisposed him to rapid growth of the ABC, leading to a highly evolved lesion.[6] The tumour blockade of the ostial region and subsequent chronic sinusitis resulted in chronic rhinitis and headache. In addition, destruction of the skull base could have contributed to the chronic headache.[3,11,15] The extension of the lesion to the orbits resulted in the right ocular pain and restriction of motility.[3,16]

Table 1
The giant cranial aneurysmal bone cyst associated with fibrous dysplasia reported in this case will increase the knowledge base of successful surgeries performed in cases of aneurysmal bone cysts located in the cranium

Authors	**Localisation of the lesion**	**Performed surgery**
Saito *et al.*[10] 1998	Skull base tumour extending to the nasal cavity, the sphenoid and posterior ethmoid sinuses, the planum sphenoidale, sella turcica, upper two-thirds of the clivus, and the medial portion of the mid-cranial base	Anterior craniofacial approach
Liu *et al.*[5] 2001	Frontal sinus	En-bloc excision with a bifrontal craniotomy approach and immediate reconstruction of the defect
de Minteguiaga *et al.*[7] 2001	Sphenoid bone	Endoscopic transnasal procedure
Nadkarni *et al.*[11] 2001	Anterior cranial fossa floor, encroaching upon all paranasal sinuses, with outward displacement of orbits and superior displacement of the frontal lobes	Basal transcranial route
Suzuki *et al.*[9] 2001	Maxillary sinus	Partial maxillectomy
Cansiz *et al.*[4] 2002	Sphenoid bone	Combined subcranial-midfacial degloving approach
Pasquini *et al.*[14] 2002	Maxilla	Endonasal endoscopic approach
Guilemany *et al.*[3] 2004	Orbitoethmoid tumour	Ethmoidectomy with orbitotomy
Guzman *et al.*[1] 2005	Maxillary sinus tumour with extension to the orbit wall and osseous palate	Maxillectomy

Destruction of the right lacrimal duct gave rise to the epiphora.[3] The tumour-induced pressure and destruction of local blood vessels caused epistaxis.

Our initial diagnosis was suggested by CT scans showing a tumour comprised of multiple cysts that contained blood and serous fluids, characteristic of an ABC. This diagnosis was corroborated with an analysis of the tumour biopsy.[5] A radical surgical en-bloc excision of the giant tumour, together with the destroyed bones of the skull base, was necessary to prevent further potentially lethal complications.[4,10,11] The facial skin incision with an extension along the upper margin of the orbit was performed for better access to the upper nasal cavity, the ethmoid sinus and the floor of the frontal sinus.[4,10] In spite of the serious preoperative symptoms and the extensive tumour resection, the patient's recovery was complete with the postoperative facial scar as the only lasting consequence.

In our opinion, a craniofacial resection was the optimal approach due to the size and location the tumour. A lateral rhinotomy would have been insufficient due to the tumour's penetration of the lamina cribrosa. On the other hand, a subfrontal approach would not have allowed visualization of the inferior part of the tumour.

The initial, preoperative diagnosis of ABC was confirmed by postoperative histological analysis of the removed tumour. In particular, an ABC was identified by the presence of cyst walls composed of vascular, proliferative soft tissue and numerous osteoclast-like multinucleated giant cells.[3] The bone surrounding the cysts consisted of immature, undifferentiated fibrous connective tissue, indicating that a pre-existing primary bone lesion characterized by polyostotic fibrous dysplasia preceded the ABC growth.[2,14,17,18]

This case is of interest due to the rarity of an ABC growth in the facial skeleton and the huge mass of the tumour. This report contributes to the database of successful surgical approaches that can be used in the management of these types of lesions.

Conclusion

An aneurysmal bone cyst located in the bony face can cause destruction of the facial skeleton and skull base. When it grows to a highly evolved lesion, we recommend performing an en-bloc excision by a craniofacial resection with an external approach and the immediate reconstruction of dural defects. In this report the patient completely recovered with an absolutely uneventful postoperative course. In addition, it was possible to preserve vision and restore ocular mobility.

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Olaf Zagólski, M.D., Ph.D.
ul. Dunin-Wąsowicza 20/II/9
30-112 Kraków
Poland
E-mail: olafzag@poczta.onet.pl