CASE REPORT

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Orbitoethmoid aneurysmal bone cyst. Case report and literature review*

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SUMMARY

Aneurysmal bone cyst is a benign, vascular, variable growing and expansive lesion that can occur in any part of the skeletal system, but mainly in long bones and vertebrae. We present a case of orbitoethmoid aneurysmal bone cyst in a 62-year-old female presenting epiphora. Nasal endoscopy was normal. Sinus CT scan revealed an expansive mass in the right ethmoid sinus extending and destroying the right lamina papiracea. The tumor was completely resected through paralateral rhinotomy. Histological analysis showed fibrous septa containing multinucleated giant cells and bone tissue surrounding blood vessel lumens, bordered by endothelial cells. These findings are characteristic of aneurysmal bone cysts. After 30 months of postoperative follow-up the patient remains disease-free and asymptomatic.

Key words: aneurysmal bone cyst, paranasal sinuses, ethmoid sinus

INTRODUCTION

Although aneurysmal bone cyst (ABC) is a non-neoplassic lesion, it expands from the affected bone and destroys the surrounding tissues. Only 2% of all aneurysmal bone cysts are found in the head and neck area, being mandible the most frequent site and usually affecting young patients. ABC was first described in 1942 by Jaffe and Lichtenstein (1942). There have been occasional reports of sphenoid (Cansiz et al., 2002) and maxillary (Suzuki et al., 2001) sinus involvement but primary ethmoid ABC is extremely rare, with only eight reported cases (Odenthal, 1967; Baker et al., 1982; Jordan et al., 1983; Patel et al., 1993; Zielnik et al., 1995; Citardi et al., 1996; Winnepenninckx et al., 2001; Hrischhikesh et al., 2002), four of them with orbital extension (Odenthal, 1967; Jordan et al., 1983; Patel et al., 1993; Citardi et al., 1996), and one in the lachrymal bone (Citardi et al., 1996).

Surgery is the therapy of choice for ABC. Radiation therapy alone or combined with chemotherapy has not been effective in the treatment. Radiation therapy carries the hazard of radiation-induced sarcoma (Bernier and Bhaskar, 1958). Another effective treatment reported is the cryosurgery, especially in ABC of the jaw (El Deeb et al., 1980).

Due to the rarity of these tumors we also report all the published ethmoidal ABC in order to contribute to the characterization of the clinicopathological features and to the management of these neoplasms.

CASE REPORT

A 62-year-old-female with no prior history of trauma or sinus disease consulted for a 2 months history of right-sided nasal pain with rhinorrhea and epiphora without nasal obstruction. She also presented right ocular pain that increased with lateral eye movement but without proptosis or limitation in ocular motility. Pupillary and funduscopic examination were normal. A follicular non-Hodkin lymphoma was diagnosed 2 years earlier by right cervical adenopathy exeresis. Currently, the

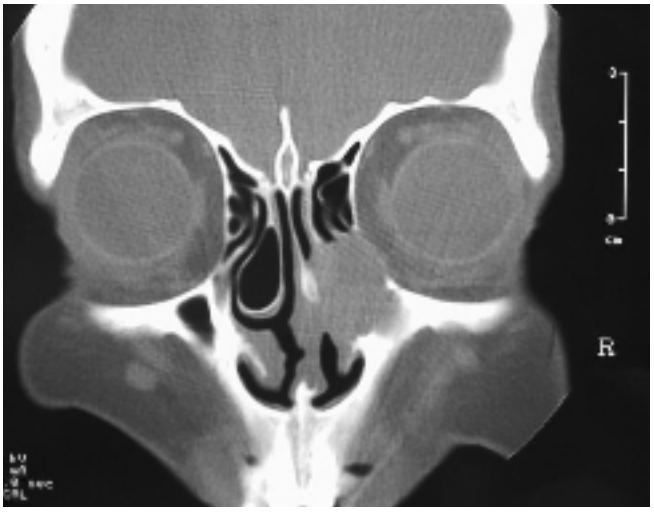

Figure 1. Coronal CT shows the lesion in the right nasal cavity with lytic and expansive characteristics, surrounded by a thin rim of bone.

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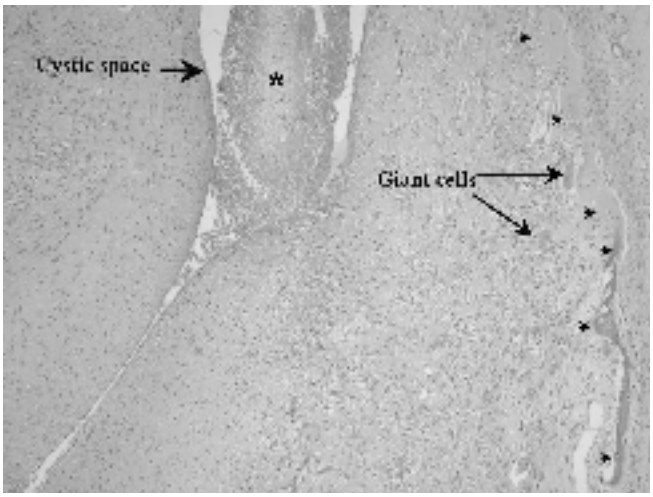

Figure 2. The lesion is constituted of fibrous tissue with presence of cystic spaces filled by blood (*). Focally, osteoid (head arrows) and giant cells are seen (HE X100).

patient presented a complete remission after chemotherapy and radiotherapy treatment. The evolution was clinically and radiologically favorable although he referred a yatrogenic anosmia. Physical examination demonstrated right-sided eye tearing and rhinorrhea. The nasal endoscopy was normal. CT scan showed a solitary, well circumscribed, and expansive ethmoidal mass (2x1,5x2 cm) with signs of bone erosion of ethmoides, right lachrymal bone and lamina papyracea. There were no pathological lymph nodes in cervical CT (Figure 1). A right paralateral rhinotomy with complete excision of the tumour was performed. The intraoperative biopsy was haemorrhagic on incision but with no signs of malignancy. The lesion was predominantly cystic and of medium consistency with a hard grey-pink capsule of connective tissue. The lachrymal sac was opened and marsupialised during the dissection to extract the tumour. To avoid postsurgical stenosis, the lachrymal tube was maintained during one month.

Histological analysis revealed a network of fibrous septa containing multinucleated giant cells surrounding blood-filled channels, bordered by a thin layer of spindle-shaped endothelial cells. The neoplasm was diagnosed as an aneurysmal bone cyst (Figure 2).

The patient had an uneventful post-operative course, remaining asymptomatic with epiphora resolution and no evidence of disease 30 months after surgery.

DISCUSSION

Aneurysmal bone cyst (ABC) is a benign, vascular and expansive lesion which pathogenesis remains still unknown. The term “aneurismal” is due to the similarities in the radiological features of an aneurysm. These tumors typically originate in membranous bones of the thorax and pelvis, long bones metaphysis and vertebrae (El Deeb et al., 1980). All the reported cases in the literature occurred in young patients, before 20 years old (Odenthal, 1967; Baker et al., 1982; Jordan et al., 1983; Patel et al., 1993; Zielnik et al., 1995; Citardi et al., 1996; Winnepenninckx et al., 2001; Hrischhikesh et al., 2002). Only <2% of all aneurysmal bone cyst are found in the head and neck area, being mandible the most frequent site. We report the second case of primary ethmoid ABC affecting lachrymal bone and the first case affecting an elder patient.

The etiology and pathogenesis of ABC remains unknown. Preexisting lesions like giant cell tumor, fibrous dysplasia, fibromyxoma, angioma, osteoblastoma, osteosarcoma, chondroblastoma, chondromyxoid fibroma, unicameral bone cyst, and eosinophilic granuloma are the most frequent lesions associated to the origin of ABC (Martinez et al., 1988). Primary ABC has been associated to hemorrhage secondary to trauma and local alterations such as venous thrombosis or arteriovenous aneurysm (Kransdorf and Sweet, 1995). However the unclear role of chemotherapy and radiotherapy, in the case reported here there was no evidence of previous trauma and pre-existing lesion in CT practised when the patient was diagnosed of follicular non-Hodkin lymphoma treated with chemotherapy and radiotherapy.

To date, only 8 cases of ethmoidal ABC have been reported. In contrast to the age of our patient, ABC has predominated in younger patients, with no differences between gender and race. Female/male ratio of patients is 2:1, while the mean age is 16.4 years (range: 1-62 years). The clinical presentation is similar to the other sinonasal tumors, the most frequent symptom being unilateral nasal congestion present for a mean of 9.2 months (range 1-36 months). Other reported symptoms are hemifacial parestesias and pain, unilateral rhinorrhea, epistaxis, epiphora, and sinusitis symptoms. Highly evolved tumors may produce signs and symptoms deriving from the orbital extension (ophthalmalgia, diplopia, exophthalmia, sensation of retroorbital pressure, and visual disturbances), and invasion of the skull base. The range of the tumor size is 1.5-8 cm, with an average size of 4 cm, and no differences on side presentation. In two cases, a history of chemotherapy and radiotherapy treatment was reported approximately two years before the ethmoidal ABC diagnosis. The presence of chromosomal changes in 17p13 and/or 16q22 could suggest that some ABC may have a neoplastic origin (Panoutsakopoulos et al., 1999).

Radiologically, the ABC may be difficult to differentiate from other tumors of the sinonasal area, although they have some characteristics that may suggest the diagnosis. CT scan shows lytic and expansive characteristics of the lesion surrounded by a thin rim of bone. Fluid and blood levels with inner septa can often be observed, especially when the patient remains quiet for at least ten minutes before CT (Davies et al., 1992). This inner septa may cause an heterogeneous enhancement on CT. MRI is a useful imaging technique that demonstrates a signal heterogeneity, due to its multi-cystic composition and its irregular contrast uptake depending on the cyst contents surrounded by a low-intensity thin rim of bone. Blood or bloody intra-

cyst fluid have a strong signal on T1- and T2-weighted images but clear intracyst fluid has a weak signal on T1-weighted images and a strong signal on T2-weighted images (Beltran et al., 1986).

Surgery is the treatment of choice. Selective arterial embolization in large ABC may improve the results of the surgery. The treatment approach depends on the tumor size and extension, ranging from conservative surgery like curettage, enucleation, and endoscopic surgery to more aggressive interventions such as paralateral rhinotomy and bicraneal craniotomy. However, there is some controversy concerning the most suitable surgical approach due to the high rate of recurrences (26-60%) that these tumors present after a simple curettage and enucleation (Liu et al., 2001). One of the reviewed cases (Zielnik et al., 1995) was treated with a local curettage, presenting recurrence in approximately 8 months after surgery.

In summary, we may conclude that the surgical treatment of choice of these kinds of tumours may be both endoscopic sinus surgery or paralateral rhinotomy and, since most recurrences present during the first year after surgery, more than two years of follow-up may not be necessary.

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