Case report/Kazuistyka

Aneurysmal bone cyst of maxilla with ectopic molar tooth – A case report

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ABSTRACT

Aneurysmal cysts of bone are rare non-neoplastic, locally aggressive lesion of bone with propensity for rapid growth, affecting mainly the long bones and spine. It rarely occurs in the head and neck region and within the head and neck mandible (especially the molar areas) is common. Aneurysmal cyst of bone involving the maxilla in the first decade of life is even rarer. We report a case of giant aneurysmal bone cyst of maxilla in an eight-year-old male with ectopic molar tooth within the cyst. Is endoscopic excision possible? Endoscopic excision of the cyst was done successfully and no recurrence was noted even after 1 year of follow up.

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Introduction

The term aneurysmal bone cyst was first coined by Jaffe and Lichtenstein in 1942 [1]. The term aneurysmal bone cyst is misleading as it is not lined by epithelium and it is not a true cyst. The World Health Organization defines an aneurysmal bone cyst (ABC) as “a benign tumor” like lesion with an expanding osteolytic nature, consisting of blood filled spaces of variable sizes separated by connective tissue septae containing trabeculae or osteoid tissue and osteoclastic giant cells [2]. An appropriate terminology would be central giant cell tumor or central giant cell lesion.

Aneurysmal bone cyst most commonly occurs in aphyseis of long bones (>50%) followed by spine (12–30%) [3]. Head and neck is not the preferred site and within the head and neck mandible (especially the molar areas) is more common than maxilla. Between 2% and 12% of ABCs occur in the maxillofacial skeleton [3]. The average age of presentation is 13 years, 80% of patients are less than 20-years-old and there is no sex predilection [3]. A diagnosis of dentigerous cyst is usually entertained when maxillary cyst is associated with ectopic tooth within cyst. However, we report a rare case of aneurysmal bone cyst of maxilla with ectopic molar tooth within the cyst causing diagnostic dilemma. Endoscopic removal of the aneurysmal bone cyst was done.

Case report

Eight-year-old male presented with history of right sided cheek swelling for two and half months with complaints of...
**Fig. 1** – Preoperative photograph of the child showing cheek swelling and proptosis

**Fig. 2** – Computed tomographic scan of nose and PNS showing multicystic expansile cystic mass in the maxilla with ectopic molar tooth
dull pain over the swelling. He also complained of protrusion of right eye for one and half months. He had no other visual complaints. There was history of swelling over the right side of palate for one and half months. There was no history of nasal obstruction; nasal discharge or nasal bleed. He denied any history of facial trauma in the past.

Physical examination showed 2 cm × 2 cm diffuse swelling of right side face. The swelling was firm to hard in consistency, non-tender and non-fluctuant. Anterior rhinoscopy showed medial bulge of the right side of nasal cavity. Oral cavity showed 3 cm × 3 cm bulge in the right side of hard palate. There was absence of second molar tooth. Eye examination showed non-axial proptosis and eye was pushed forward, outward and upward. Visual acuity was 6/6 in both eyes and extra ocular movements were full (Fig. 1).

Contrast enhanced computed tomography of nose and PNS showed expansile, multi loculated, homogenous, soft tissue mass within the right maxillary sinus with thinning of the walls of maxillary sinus. The mass was extending superiorly into the ethmoid sinus and inferiorly involving the hard palate and medially the mass was extending into nasal cavity. A radio opaque mass was seen in the ethmoid sinus? Ectopic tooth suspected (Fig. 2). MRI showed hypo intense on T1, hyper intense on T2 weighted, expansive multi loculated cystic lesion with fibrous septae filling right maxillary sinus, and hypointense shadow on T1 weighted in ethmoid sinus Ectopic tooth? (Fig. 3).

Based on clinical and radiological findings dentigerous cyst, radicular cyst, fibrous dysplasia, ameloblastoma were suspected. Fine needle aspiration was done; altered blood was aspirated and was reported as cyst.

Endoscopic excision of lesion was done. Inferior and middle turbinate was excised and right maxilla was opened. Bluish colored cystic lesion was seen. The cyst was excised completely with help of micro debrider. Molar tooth was removed from the right ethmoid sinus. The wall of cyst was sent for histopathology (Fig. 4).

Histopathology showed multiple blood filled spaces with stroma consisting of spindle shaped cells and multinucleate giant cells and at places osteoid with prominent osteoblastic rimming was seen. The features were consistent with aneurysmal bone cyst (Fig. 5).

The child is under our regular follow up. Postoperative CT scan of nose and PNS was done after 3 month and there is no evidence of recurrence (Fig. 6).

Discussion

Only 2% of aneurysmal bone cysts occur in the head and neck region with lower jaw being the most common site of involvement [3]. Aneurysmal bone cysts of maxilla are rare and are usually associated with other bony lesions such as fibrous dysplasia [4, 5]. In our review of literature we encountered 120 cases of aneurysmal bone cyst of maxilla. However, only seven cases of primary aneurysmal bone cyst of maxilla have been reported till date. Our case is unique because it was primary aneurysmal bone cyst of maxilla with ectopic molar tooth within the cyst. The average age of presentation is 13 years, 80% of patients are less than 20-year-old and there is no sex predilection [3]. The present case was 8-year-old male child.

Aetiopathogenesis of aneurysmal bone cyst is still controversial and various theories have been put forward to explain its pathogenesis. Popular theory classifies it as primary or congenital or secondary (arising within the
Fig. 4 – Endoscopic excision of the bluish cystic lesion with ectopic molar tooth within the cyst

Fig. 5 – Histopathology showing multiple blood filled spaces with multinucleate giant cells and osteoblastic rimming and osteoid elements consistent with aneurismal bone cyst
preexisting bone lesion such as fibrous dysplasia) [6, 7]. A traumatic etiology i.e. bone trauma has also been favored by some authors [1]. Few authors propose intra medullary hematoma to be cause for aneurysmal bone cyst and this explains why aneurysmal bone cysts are more common in the long bones [8]. Some have suggested vascular origin for aneurysmal bone cyst [9]. They proposed that local hemodynamic disturbances, like arterio-venous shunts or malformations, increase intraosseous venous pressure with expansion of the vascular tissue bed, leading to bone resorption and cyst formation and cystic appearance on radiograph [9]. Recently, chromosomal alterations of segments 17p and 16q have been described suggesting a neoplastic origin of the lesion [10].

Aneurysmal bone cysts are variable in their mode of presentation. It may vary from asymptomatic slowly progressive swelling to rapidly expanding swelling giving rise to pain, deformity, and pressure symptoms. Aneurysmal bone cyst of maxilla usually presents with facial pain, facial swelling, nasal obstruction, proptosis, loosening of teeth and malocclusion. Common differential diagnosis includes fibrous dysplasia, dentigerous cyst, radicular cyst, giant cell reparative granuloma, ameloblastoma, simple maxillary cyst.

Radiology is suggestive but not diagnostic for aneurysmal bone cyst [11]. Radiographically it may appear as expansive, osteolytic, unicocular or multilocular radiolucent lesion, with expansion and thinning of the surrounding cortical bone. This variegated appearance in imaging could be due to the various types of aneurismal bone cyst (solid, vascular and mixed) [12]. The radiographic appearance of maxillary aneurysmal bone cysts is even less characteristic than that of mandibular lesion. MRI is mandatory in complex case as in this case. MRI findings of fluid-fluid levels within the lesion are specific for aneurysmal bone cyst [13]. On CT scan ABC may appear unicystic, multilocular, or moth eaten, causing expansion, perforation, or extensive destruction of the bony cortices. CT scan accurately identifies tissue septae [11].

Histologically of aneurysmal bone cyst consists of numerous blood-filled caverns or sinusoids surrounded by variable amount of fibrous connective tissue stroma. The connective tissue stroma contains multinucleated osteoclast like giant cells adjacent to sinusoidal spaces, inflammatory cells, extravasated erythrocytes and hemosiderin. Osteoblasts and woven osteoid bone may be seen within the connective tissue stroma [14]. Three types of ABC have been described based on histopathological features [14]. Solid type (5% of the cases) is characterized by a dense stroma, scanty sinusoids, few blood vessels and caverns, bone expansion (instead of perforation), and without severe bleeding during surgery. Vascular variant (95% of cases) is characterized by a loose scanty stroma, numerous engorged blood filled sinusoids and caverns. Brisk bleeding during surgery and extensive bony destruction with spread in the soft tissues are also obvious. The mixed type lies between the 2 previous variants [14]. Our case was of solid variant type as it was not associated with severe bleeding during surgery.

The treatment of choice for aneurysmal bone cyst of maxilla is conservative surgical resection [15]. Conservative
surgical methods as curettage must be preferred for young patients. Angiography with preoperative embolization of feeder vessels has been advocated to reduce bleeding during surgery. Various approaches used for maxillary aneurismal bone cyst include open maxillectomy, Caldwell luc and transcancele approach and endoscopic approach. Endoscopic approach is preferred and can be performed in select cases. We have used endoscopic approach to excise tumor in the reported case.

Recurrence is major issue in the management of aneurysmal bone cyst. Simple curettage is associated with high recurrence rates, varying from 21% to 50% [16]. For lesions that are resected with more radical methods the recurrence rates vary from 11% to 25%. Most cases recur in the first year of initial treatment. It is most commonly due to inadequate or incomplete removal of tumor. The treatment for the recurrent lesions must be radical (mandibulectomy or maxillectomy), especially when recurrences are multiple. Irradiation of the ABC is not advisable due to the high risk of malignant change into a sarcoma [17].

Conclusion

Primary aneurismal bone cysts of maxilla are rare. Ectopic molar tooth within aneurysmal bone cyst is even rarer and can lead to diagnostic dilemma. MRI and CT scans are crucial for diagnosis. Endoscopic excision of the ABC of maxilla can be done successfully. Preoperative embolization of feeder vessels may reduce bleeding during surgery. Recurrent case would require more radical approach.

Authors' contributions/Wkład autorów

RKV, RK – prepared manuscript, AB – reported pathology, NKP – edited manuscript.

Conflict of interest/Konflikt interesu

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Ethics/Etyka

The work described in this article has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans; EU Directive 2010/63/EU for animal experiments; Uniform Requirements for manuscripts submitted to Biomedical journals.

REFERENCES/PIŚMIENNICTWO