

Case Report

Neglected Aneurysmal Bone Cyst of the Mandible: A Case Report in a 10 Year Old Girl in Volta Region of Ghana

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Abstract

Cystic bony lesions of the craniofacial region are rare, and if found may be primary or secondary. Clinicians in less resourced health facilities are faced with diagnostic challenges regarding these lesions in the absence of appropriate diagnostic or investigative tools. This is compounded by the lack of specific signs and symptoms and also the influence of herbal medicine. We report a case of a huge cystic bony lesion on the mandible of a 10-years girl from the Volta Region of Ghana. Histopathological and radiological investigations reported the lesion as a primary aneurysmal bone cyst. She was discharged on the 7th operative day and currently doing well.

INTRODUCTION

Aneurysmal bone cyst (ABC) of the jaw is a rare condition in the literature, especially in Ghana [1-3]. This condition was first recognized by Jaffe Lichtenstein as a distinct clinicopathological entity in 1942 [2]. ABCs represent approximately 1% -2% of all primary bone lesions that are sampled for biopsy [1]. The lesion tends to affect the metaphyses of long bones and the dorsal elements of the vertebrae [4-9]. This pathological entity is common in children [8-12] with a slight female preponderance, a male-to-female ratio of 1 - 1.04 [9,12]. The first reported case of the cyst was by Bernier and Bhasker in 1958 [13]. Subsequently, a lot of information on the clinical and histopathological presentation including pathogenesis has been described, however, only few cases of primary ABC are documented [3,13].

We report a case of a primary mandibular aneurysmal bone cyst occurring in the mandible of a 10-years girl from the Volta Region of Ghana.

CASE REPORT

Clinical history

A 10-year old adolescent girl, presented to the maxillofacial unit of the Tamale Teaching Hospital (TTH) with a 3-year history of jaw swelling that had gradually increased in size over the period. She had some initial treatments at the Krachi West Hospital without improvement. The condition was associated with fever, weight loss, dysphagia, headache and fatigue. She had no previous history of any bony lesion and no family history of bony malignancy.

PHYSICAL EXAMINATION

Extra oral

A huge ill-defined firm to hard extra-oral tender mass (10.0 x10.0cm), on the lower right jaw extending from right mandibular ramus to the cervical region. Mass extends from right pre-auricular region to the angle of the mandible.

Intra oral

Lesions extend bucco-lingually more than 2/3 of the oral cavity with displacement (dental anarchy) of involved teeth and distortion of the floor of the mouth, but the oral mucosa was not ulcerated.

There was associated poor oral hygiene.

She was warm to touch and moderately pale, but not in respiratory distress. The cervical lymph nodes were not enlarged.

Working diagnosis

Infected benign mandibular tumour

Differential diagnosis

1. Odontogenic keratocyst
2. Cystic ameloblastoma

MANAGEMENT

Investigation

a. Full blood count (FBC): WBC=10.12x10⁹/L, HB= 8.6g/dl, and PLT=523x10⁹/L

b. Sickling: Negative

c. Plain right lateral x-ray of the mandible: X-ray shows a huge multiloculated osteolytic lesion limited to the mandible, with thinning of the intra-lesional walls and the cortex. There is associated destruction (roots) and displacement of the involved teeth (Figure 1).

d. CT Scan requested but not done.

Drug treatment

The patient was managed pre-operatively with intravenous infusions and tranexamic acid.

Blood transfusion

She was transfused 3 units of packed red blood cells post operatively.

Surgery (Excision biopsy)

Subtotal mandibulectomy with reconstruction plate was done. Intra-operatively, the mass was found to be multicystic with straw- coloured fluid

HISTOPATHOLOGY

Gross

Received a resected 14.0x12.5x7.5cm tan-dark-brown nodular mass with four attached teeth. The cut surface is solid, whitish gray with cystic areas (Figure 2a)

Microscopy

Sections of representative portions of right mandibular bony mass show a tumour composed predominantly of proliferating monomorphic stroma cells and vascular channels. The vessels are filled with blood and have no endothelial lining. No malignancy seen (Figure 2b and 2c)

Histopathological diagnosis

Mandible: Aneurysmal bone cyst of the mandible; solid variant

DISCHARGED AND POST-OPERATIVE PERIOD MANAGEMENT

Patient was discharged on the 7th post-operative day. She was put on the following oral (suspensions) medications: amoksisclav,



Figure 1 Plain X-ray of the mandible of a 10-year old girl with a huge limited multiloculated osteolytic lesion.

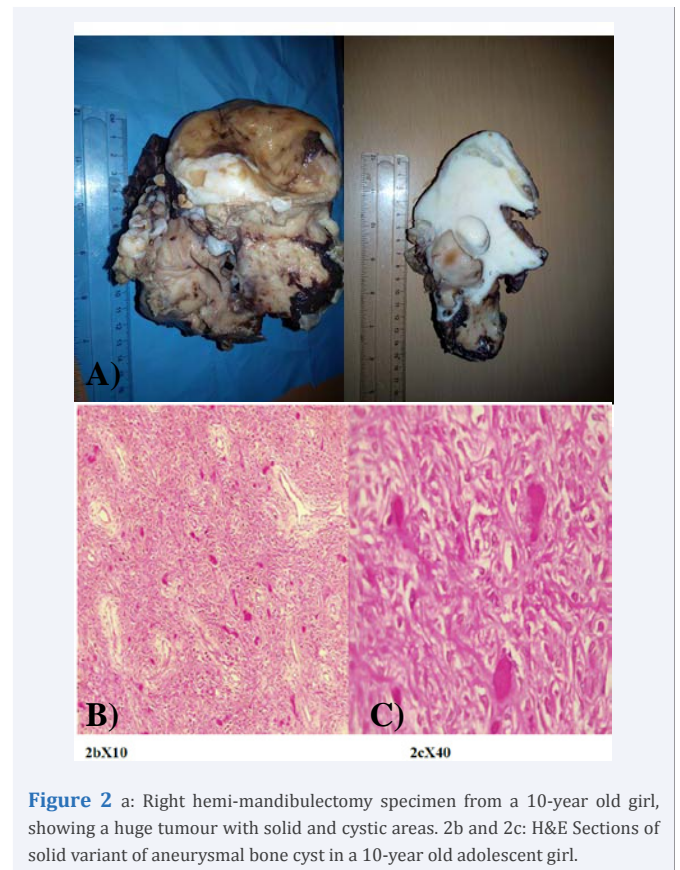


Figure 2 a: Right hemi-mandibulectomy specimen from a 10-year old girl, showing a huge tumour with solid and cystic areas. 2b and 2c: H&E Sections of solid variant of aneurysmal bone cyst in a 10-year old adolescent girl.

metronidazole and paracetamol. She was to have daily open wound dressing with povidone iodine and continue with the oral medications.

DISCUSSION

There is controversy surrounding the aetiology and pathogenesis of aneurysmal bone cyst (ABC). Some authorities are of the opinion that it arises from pre-existing bony lesions [13,14] while others reported it as a primary lesion of the bone [13]. In this current case report, a 10-year female presented with a huge mandibular swelling of 3 years duration that almost occupies the entire oral cavity with displacement of the involved teeth and floor of the mouth. Radiological and histopathological investigations reported the lesion as ABC. Since she had no prior history of bony disease, non-traumatic craniofacial injury, we concluded that her ABC was primary in origin. The anatomic location of the ABC in Ms in this case is in keeping with studies on craniofacial ABC, that reported the lower jaw as the most frequently affected, particularly the body and the mandibular ramus than the maxilla [4-7]. Similarly, the gender and age characteristics of the current case report is in accordance with reported cases of ABC in the literature [8-14].

The typical clinical presentation of ABC lesions is a well-defined swelling of soft tissues due to expansion of the adjacent or underlying bony lesion causing noticeable facial asymmetry [15-17] as described clinically in this current case report. ABCs usually present as a slow progressive growth until cortical plates are eroded at any point and then show a rapid growth.

Malocclusion can be a consequence of facial deformity [18]. These support the clinical presentation of the adolescent female in this current case report, where it was found that the lesion has occupied about two-thirds of the oral cavity with displacement of the involved teeth, mimicking oral presentation of Burkitt's lymphoma in African children.

Pain is an infrequent symptom of ABC, except of rapidly growing lesion [19] as in the present case. Other less common clinical presentations could be root resorptions of teeth, disesthesias, proptosis, diplopia and progressive nasal obstruction in maxillary lesions [19,20]. Solid aneurysmal bone cyst is usually asymptomatic whereas vascular form usually presents an invasive rapid growing evolution with extension to overlying tissues [16,21,22]. The ABC in this case report was a solid variant, less painful and this may explain the late stage at presentation with the associated intra-oral findings.

Moving forward, the clinicopathological characteristics present in this current case report reflect a neglected condition, and may thus just been the tip of an iceberg regarding the clinical stage at which patients in Ghana present with bony lesions to our health facilities. The clinical picture painted here may potentially be attributed to the fact that, most bony lesion present with non-specific clinical feature and may be painless, unless complicated by erosion into vascular spaces, nerve compression or ulceration with superimposed infection. In this advanced stage patients may then seek medical care in a health facility as in this case report. In certain instances, even at the health facility, the initial diagnosis or treatment may be missed and this may further compound the problem. Also, the proliferation and easy accessibility as well as low cost herbal medicine that treat all manner of illnesses in our country Ghana is a major contributory factor with these advanced clinical stages of presentation of most tumours. Simple and benign bony lesion may unduly be delayed by these herbalists, or have the presentation of the primary pathology, and by the time these cases find their way to a health facility clinical judgment is hampered by whatever treatment or procedure that was earlier performed by the herbalist.

Thus benign bony lesion without the availability of the appropriate investigative or diagnostic will remain a diagnostic challenge in most developing countries such as Ghana.

CONCLUSION

ABCs are uncommon, but may presents very late with advanced clinical features suggestive of a neglected disease as in this case. The advanced stage of presentation of ABC may be attributed to lack of information on the disease, low perception of its existence by clinicians and the availability and accessibility of alternative medicine that prevent seeking orthodox medicine. Clinicians in less resourced hospitals have a diagnostic challenge regarding bony lesions, and this calls for health authorities to re-think about the in equality in distributions of health equipments in Ghana.

RECOMMENDATIONS

1. Herbalists should be trained to identify and refer bone diseases that require specialist care and also not to delay patients at their camps.

2. There is the need to re-look at the efficiency and the coverage of the national health insurance scheme, to include most major surgical operations.
3. District and regional hospitals across the country should be well resourced and equipped to recognize and manage such maxillofacial conditions early.
4. Patients and patient relatives should be educated on the need for early reporting bone abnormalities.
5. Hospital patient referral systems should be strengthened.

CONSENT TO PUBLISH THIS CASE REPORT

We obtained verbally permission from the parents of the patients.

AUTHOR'S CONTRIBUTIONS

EDM drafted the case report. MY provided the clinical history and performed the surgery. EDM (Pathologist) prepared the tissue and reported the slides. EDM, and MY read through the case report, edited and approve it for publication

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