ODONTOGENIC MYXOMA OF THE MAXILLARY SINUS IN A NIGERIAN CHILD – REPORT OF A CASE

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ABSTRACT

A case of grossly disfiguring swelling of the right maxilla in a Nigerian girl with delayed presentation due to socio-economic factors is presented.

A 15 year old girl presented with a right facial upper jaw swelling of six years duration. It was painless and progressive, with expansion into the right maxilla, zygoma and filling the maxillary antrum with an intra-oral extension and nasal obstruction. The CT scan revealed an expansive radiolucent mass of the right maxilla and the lesion was surgically excised completely via a right maxillectomy.

The histolopathological features of the excised mass are consistent with fibrous dysplasia of the right maxilla which has undergone myxomatous degeneration.

This case highlights the clinical, radiological and histological characteristics of this rare locally aggressive tumour that underwent myxomatous degeneration as a result of late presentation due to socio-economic factors .More concerted efforts should be made to make the national health insurance scheme fully operational. This will make presentations at the hospitals not to be delayed thereby avoiding complications.

Key words: *Odontogenic, Maxilla; Myxomatous degeneration; Socio-economic factors.*

INTRODUCTION

Myxofibroma (odontogenic myxoma) is a rare benign tumour probably derived from ectomesenchyme[1]. It is a slow growing, locally invasive jaw tumour constituting approximately 8% of all odontogenic tumours[2]. The earliest manifestation is a painless, expansile mass. The teeth in the affected area may separate, migrate and may exhibit root resorption. Radiologically, it occurs as expansive radiolucent unilocular or multilocular lesions with irregular or scalloped margin, found often in tooth bearing regions with thinning and /or perforation of the cortex with extension into the soft tissue[3, 4].

Histologically, the lesion is composed of small stellate or spindle shaped cells with a mucoid rich matrix. Few collagen fibrils are typically present[5, 6].

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The treatment is by surgical excision with long term follow-up, as recurrence rate is high (reportedly 25%) [7]. This high rate may be related more to difficulties encountered with complete surgical excision rather than the inherent biological aggressiveness of the tumour[7]. This case highlights the clinical, radiological and histological characteristics of this rare tumour that underwent myxomatous degeneration as a result of late presentation due largely to socio-economic factors.

CASE PRESENTATION

A 15 year old Nigerian girl presented with a six - year history of a painless progressive swelling of the right upper jaw, extending into the oral cavity. There was no anorexia, weight loss, fever or neck swellings. There was nasal blockage with nasal speech and loss of upper right quadrant on the same side associated with bleeding on brushing of the teeth. She had visited traditional practitioners prior to presentation without improvement.

Examination revealed a bony hard grotesque swelling of the right maxilla measuring 12x12cm, non tender, extending from the right zygoma and encroaching on the right infraorbital ridge displacing the nose to the left (Fig. 1). Intraorally, there is a bony hard swelling filling the whole of the oral cavity with loss in the upper right quadrant (Fig. 2).

The nose was patent but with slight obliteration of right nasal cavity. A clinical diagnosis of right maxillary fibrous dysplasia was made, histopathologic examination of tissue showed fibrous dysplasia with myxomatous degeneration.

Scanogram of the maxillary sinus showed an expansile mass involving the maxilla with mixed density and widespread irregular hyperdense area (bone) interspersed with hypodense areas resembling teeth. Haematologic and biochemical investigations were within normal limits. Operative procedure under a cuffed oral endotacheal intubations, a right Weber- Ferguson incision was made, lesion exposed and right maxillectomy was done. Surgical defect was obliterated with a gutta parcha for 7 days. Patient was discharged home postoperatively after 10 uneventful days. A dental obtrurator was fixed and the out patient

Fig. 1: At presentation (Intraoral extension)

Fig. 2: Right maxillary swelling at presentation

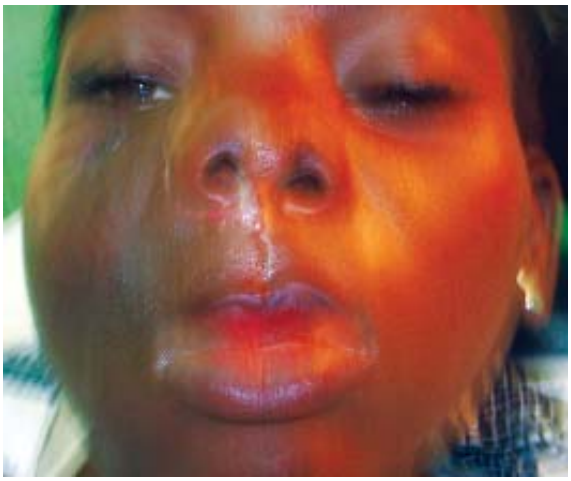

Fig. 3: 12-months post Excision of Tumour

clinic visits have been uneventful nearly two years after surgery.

The excised specimen weighed 150grams and about 10x10x2cm in its widest dimension. The globular tumour mass was attached to the central aspect of the maxilla and is 4x2x1cm in its widest dimension and was firm to hard in consistency. The microscopic features showed fragments of tissue composed of proliferating stellate cells within myxoid stroma and fibroblasts and trabeculae of bone tissue infiltrated by inflammatory cells which mainly comprised of lymphocytes and plasma cells. The overall histopathological features are consistent with myxofibroma.

DISCUSSION

This rare tumour of the jaw is locally aggressive occurring most commonly in the second to third decades of life and the incidence vary widely world wide. It occurred in a female child in her second decade of life in our case as in previous series [2]. However, the presentation here is delayed leading to wide intra-oral extension as a result of poor socio-economic factors. It usually presents as a painless facial swelling and it tends to enlarge and often fill the maxillary sinus before presenting as a facial swelling[1]. The destructive nature of the tumour can cause nasal obstruction, ocular changes or even palatal swellings, all these features were present in our case and it affected the maxilla even though the mandible is usually more affected[1]. On CT scan, images of the odontogenic myxoma shows osteolytic expansile lesions with mild enhancement of the solid portion of the mass with expansion and thinning of the surrounding bony boundaries, as in the case reported [8]. Histologically, the tumour cells are small spindle shaped or stellate cells embedded in a myxoid background which distinguishes it from chondromyxoid fibromas, dentigerous cyst, giant cell granuloma, sarcoma, ameloblastoma and inflammatory polyp[5,6].

However, the tumour shows local infiltration as seen in this case and this explains the local recurrence in situations where incomplete excision is done.

The treatment has traditionally been surgery because myxoma is radio-resistant and adjuvant radiotherapy is generally not recommended in the treatment but some authors advocate pre-operative radiation to achieve shrinkage of the tumour, though, benefit from such therapy is in contention [9] .

Although, most authors recommend conservative enucleation and curettage approach for small lesions and resection for large tumours followed by regular follow up, in our case a more radical approach of maxillectomy was adopted because of the extensive nature of the tumour and to avoid recurrence in an environment with poor follow-up records. In this patient, follow-up has been for two years with no recurrence.

In conclusion, this tumour underwent myxomatous degeneration due to delayed presentation as a result of socio-economic factors. More concerted efforts should be made to make the national health insurance scheme fully operational which will make presentations at the hospitals not to be delayed thereby avoiding these complications.

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