CASE REPORT

Bimaxillary presentation of central ossifying fibroma: a unique aggressive entity

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SUMMARY

Central ossifying fibroma is a benign neoplasm, having slow growing nature. Some rare lesions show very aggressive nature, multifocal appearance and reach up to a very massive size. So, these kinds of cases require special attention for their treatment. A unique case of central ossifying fibroma with aggressive nature, multifocal appearance is reported. This case shows growth both in maxilla and mandible with maxillary lesion massive in size involving maxillary sinus. There have not been any such cases reported so far in the literature showing bimaxillary growth of ossifying fibroma.

BACKGROUND

Ossifying fibroma (OF) is a benign fibro-osseous lesion (FOL), commonly found in the jaws, especially in the mandible. Buccal or lingual cortical bone swelling or expansion, slow growing is the most common clinical feature and an aggressive form causing the facial asymmetry. The radiographic picture of OF is more frequently a well-defined mixed lesion (radiolucent/radiopaque). Most OF can be treated by conservative surgical curettage or excision. Although the recurrence rate of this tumour seems to be extremely low, the patients should be on regular follow-up. Thus, here we present the expansile form of these lesions, which developed first in mandible and later recurrence in maxillae, where the whole lesion was removed and displayed no signs of recurrence. The patient is on continuous follow-up since the past 2 years and shows no sign of recurrence and is happy with her facial aesthetics preservation with Weber Ferguson approach.

CASE PRESENTATION

A 22-year-old female patient reported to the oral and maxillofacial surgery department presenting with giant painless swelling since the last 2 months involving the maxilla. Initially, the swelling was smaller in size and gradually increased in size after her accidental fall from the staircase. The patient had a history of segmental mandibulectomy 40 months previously at our centre for the same kind of growth involving the right side of the mandible, and she had no other relevant medical history. The patient was unmarried and lives with her mother.

Examination revealed a large, non-tender, hard swelling on the right side of the maxilla extending superiorly from lip commisure inferiorly to supra-orbital rim and anteroposteriorly from lateral wall

of nose to 1 cm to the tragus anteriorly. Swelling had pushed the right eye Craniolaterally. Inferior extension showed lateral nasal fold obliteration with the involvement of fronto-nasal suture and marked deviation of nares towards the left side. Swelling measured approximately 7 cm×7 cm in dimension (figure 1).

Overlying skin showed normal colour with stretched contour. The right eye had normal vision and eye movements. Intraorally, maxillary alveolus on the right side showed considerable expansion and obliteration of the buccal sulcus starting from canine to the second molar. Swelling involved the hard palate beyond the midline and extended posteriorly to the soft palate also. The right side mandible teeth were absent, as they were removed at the time of segmental mandibulectomy 40 months previously and restored by prosthetic rehabilitation.

INVESTIGATIONS

Along with routine blood investigations, blood calcium levels were found to be within normal limits, thus no sign of hyperparathyroidism was noted.

CT scan showed large well-defined expansile lesion in maxillary region on the right side measuring about 4.9 cm×4.4 cm×4.6 cm in dimension. The lesion appeared heterogeneously hyper dense and showed multiple internal punctuate calcifications along with mild heterogeneous enhancement on postcontrast scan. The right maxilla, medial, lateral and anterior wall of the right maxillary sinus along with right upper alveolar margin appeared eroded by the lesion. The lesion was seen occupying the right maxillary sinus and right side of nasal cavity (figure 2).

Other accessory findings were left maxillary sinus showed soft tissue lesion measuring about

Figure 1 Preoperative extraoral photograph of the lesion (frontal and lateral views).

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Figure 2 Preoperative CT scan of the maxillary and mandibular lesions.

1.1 cm×1 cm was seen in the floor of left maxillary sinus. Another lytic expansile lesion measuring about 1.5 cm×1.7 cm×1.1 cm was seen in the right ramus of mandible. Right body and ramus of mandible showed discontinuity with thinning of bone due to postoperative changes (figure 3).

Bones showed generalised diffuse osteopenia with prominent trabeculae. Incisional biopsy was performed and histopathological examination revealed that the specimen showed characteristics of central OF.

Histopathological report revealed a well-circumscribed mass of highly cellular connective tissue stroma. The cellular stroma was composed of spindle-shaped cells were arranged in whorl pattern (figure 4). Osteoid areas with osteoblastic lining were also noted with stroma. Periphery of some sections showed parallelly arranged collagen fibres suggesting the lesion to be well circumscribed. This was confirmative of central OF.

DIAGNOSTIC FLOW CHART

The most important point to keep in mind that patient was already operated for OF of right mandible 40 months back—could be case of recurrence/spread.

- ▶ First—clinical (extraoral and intraoral examination; large, harge, non-tender swelling—suggestive of tumour mass);
- ▶ Second—radiographic interpretation (heterogeneously hyperdense—suggestive of any tumour, ameloblastoma, odontogenic cyst, fibrous dysplasias, hyperparathyroidism jaw tumour syndrome);

Figure 3 Preoperative three-dimensional CT scan (both frontal and lateral views).

Figure 4 The microscopic features of central ossifying fibroma.

- ▶ Third—laboratory investigations (routine blood investigations, blood calcium—within normal limits);
- ▶ Fourth—histopathological examination—confirmed it to be OF.

DIFFERENTIAL DIAGNOSIS

Hyperparathyroidism-jaw tumour's syndrome (HPT-JT), osseous dysplasia, osteofibroma.

TREATMENT

The patient was operated under general anaesthesia and site exposed via a Weber Ferguson approach. The incision line was drawn through the vermilion border, along theiltrum of the lip, extending around the base of the nose (or entering the nostril floor for a better aesthetic result) and along the facial nasal groove. It then extends infraorbitally 3–4 mm below the cilium to the lateral canthus extending as subciliary incision below lower eye lid. The tumour was seen to have dissected its way through the lines of least resistance involving the maxilla, zygoma and the maxillary sinus and had extended well into the nasal cavity. Whole tumour was raised and removed in toto (figure 5). Teeth involved were also removed with the lesion. Two small daughter tumours were also removed; one from anteroposteriorly over maxillary tuberosity and second from posterior wall of the maxillary sinus. Care was taken while dissecting in posterior, lateral and medial margin to prevent any

Figure 5 Intraoperative photograph showing the lesion after incision and flap reflection.

unnecessary damage to the surround vital structures. Nasal antrostomy and antral packing were carried out. Closure was performed and specimen was sent for histopathological examination. Antral pack was removed after 36 h.

OUTCOME AND FOLLOW-UP

The patient was on monthly follow-up. The patient was last seen after period of 2 years, when she was in good health, with no recurrence and subsequently going to maintain follow-up (figure 6).

DISCUSSION

Benign FOLs are a group of pathological entities where the normal bone is replaced by fibroblast and collagen fibres along with varying amount of mineralised material. This group includes fibrous dysplasia, benign fibro-osseous neoplasms (central OF) and a heterogeneous group of reactive lesions (osseous dysplasias).^{1–3} These are poorly defined and are among the controversial group of lesions affecting the jaws and craniofacial bones. They have similarity in its histological picture, thus definitive diagnosis requires close clinical, histopathological and radiological examination.

The WHO currently defines OF as a benign neoplasm, often presents well-demarcated borders and is composed histologically

of fibrocellular stroma and variable amounts of mineralised material showing different morphological appearance.¹

The occurrence of multiple or recurrent OF in the jaws is considered to be rare and is associated with hormonal abnormalities, such as hypercalcaemia associated with hyperparathyroidism. OF affect younger generation, involves nose and paranasal sinuses and display as craniofacial dysplasia and juvenile dysplasia. Clinicians should be aware of the possibility of HPT-JT in young patients because this disease is usually associated with recurrent or multiple OFs of the jaws. HPT-JT syndrome is an autosomal dominant disorder characterised by parathyroid tumours, FOLs of the mandible and maxilla, renal cysts or Wilms' tumours.^{4 5}

One of the classic papers in 1968, by Hammer *et al*⁶ reviewed 249 cases which they considered to be benign FOLs of the jaws of periodontal membrane origin. They stated that the fibrous connective tissue of the periodontium contains mesenchymal blastic cells with the potential to form cementum, alveolar bone and fibrous tissue. They categorised as cementoid, osteoid, cemento-osteoid and fibroid lesions. They included fibrous dysplasia separately under medullary bone origin and claiming furthermore that it had distinctive histological features.

The difficulty in establishing a definitive diagnosis through any single diagnostic modality was confusing due to

Figure 6 Postoperative photograph of the disease-free patient (intraoral and extraoral views).

surrounding classification of the tumour. Adjunctive radiographic and nuclear medicine diagnostic aids were utilised, as were clinical, laboratory and histopathological studies, in resolving the diagnostic questions posed by this large and aggressive tumour. Surgical intervention through conservative enucleation produced few operative or postoperative difficulties and minimal residual deformity. Differentiation of OF from other benign and malignant neoplasms as well as fibrous dysplasia is important in the correct management of this lesion.⁷

Menzel in 1842 first described the term known as OF. Montgomery the one who in 1927 coined the term 'ossifying fibroma'.^{7, 8} The introduction of the term 'fibrous dysplasia' by Lichtenstein in 1938⁹ suggested that lesions of the jaws previously designated as fibrous osteoma or OF should be called fibrous dysplasias.

For the management of these lesions, there are varied schools of thought with several options (1) observation, (2) curettage and (3) resection. If treatment is with conservative approach, careful long-term follow-up is needed. Lesion treated with conservative surgery, but exhibits rapid multiple recurrences should be considered for resection.¹⁰ A careful excision with cauterisation of the surrounding region, during a period of quiescence. Some said that these lesions are of benign nature, massive resection, with its accompanying disfigurement and high mortality was unjustified. They suggested that the tumours can be operatively removed as thoroughly as possible without too great destruction of the jawbone. The other authors also believe that roentgen therapy serves to inhibit further growth of portions of the lesion not removed at operation.

Lastly, the treatment of benign OF is a conservative excision of the well-circumscribed lesion without extensive resection of adjacent structures unless involved by the lesion. In case of a sudden growth spurt with aggressive behaviour be witnessed, an en bloc resection should be considered for definitive therapy.

Learning points

- ▶ Clinicians should be aware of the possibility of hyperparathyroidism-jaw tumours syndrome in young patients, because this disease is usually associated with recurrent or multiples ossifying fibroma (OFs) of the jaws.
- ▶ Most OF can be treated by conservative surgical curettage or excision. Although the recurrence rate of this tumour seems to be extremely low, the patients should be on regular follow-up.
- ▶ Careful clinical examination and correlating it with histopathological findings and additional radiographic and laboratory investigation for planning the surgical excision should be carried out, since this terminology has been correlated with other osseous dysplasia with similar radiographic and clinical findings, to avoid recurrence.

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