Craniofacial fibrous dysplasia

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ABSTRACT
Fibrous dysplasia can present clinically in varied forms which may appear as collision of different pathologic processes. We report a rare case of craniofacial fibrous dysplasia with coexisting epithelial lined cyst and superimposed osteomyelitis with sequestrum formation. Its clinical features and management with possible hypotheses are described along with the post operative course. Pertinent literature has been reviewed with emphasis on pathogenesis of this unique occurrence.

Key words: Craniofacial fibrous dysplasia, epithelial lined cyst, osteomyelitis
Operative procedure

Under right maxillary nerve block supplemented with local infiltration for haemostasis, a mucoperiosteal flap was raised and the lesion was exposed till the infraorbital rim superiorly and body of zygoma posterosuperiorly. In the alveolar region the buccal cortex was destroyed and replaced by thick cystic lining. Teeth #14-17 were extracted and cystic lining was curetted out. The cystic lining contained little bit of pus and three small pieces of necrotic bone. Then the residual bone was curetted particularly over the anterior surface of maxilla, body of zygoma and infraorbital rim [Figure 4]. Since there was no paresthesia or compression symptom in infraorbital nerve, the rim of bone surrounding it was left untouched.

On histopathology, the bony pieces retrieved after recontouring revealed uniformly distributed curvilinear trabeculae in a fibrocellular stroma with numerous fibroblasts and a few blood vessels, overall picture appeared to be consistent with mature stage of fibrous dysplasia [Figure 5].

The lining exhibited a nonkeratinised stratified squamous epithelium with proliferating and arcading rete ridges and an underlying connective tissue capsule showing intense infiltration by chronic inflammatory cells chiefly lymphocytes and plasma cells [Figure 6]. The three pieces of bone contained within the lining were necrotic i.e. sequestrum [Figure 7]. Thus a diagnosis of osteomyelitis with coexisting epithelialised cyst in fibrous dysplasia was confirmed.

One year postoperatively good symmetry was maintained in the frontal view but slight expansion was still visible on the worm’s eye view. No recurrence of cystic cavity was observed in one year post operative CT scan and the bony defect healed completely. Three years postoperatively the patient came back seeking correction of facial asymmetry caused by bony growth in the same region [Figure 8]. Facial recontouring was performed again and the bone pieces removed showed microscopic picture consistent with fibrous dysplasia. No cystic lining or avascular bone was observed.
DISCUSSION

Facial region may be affected by a form of fibrous dysplasia that is not strictly monostotic, but may be confined to a single anatomical region. These lesions affect primarily the maxilla and simultaneously cross sutures and enter into the adjacent facial bones. This type of fibrous dysplasia does not meet the precise criteria for the monostotic or polyostotic forms and has been termed craniofacial fibrous dysplasia. In the present case, fibrous dysplasia affected the maxilla and adjacent zygomatic bone and thus may be termed as craniofacial type.

As per the current belief fibrous dysplasia results from a defect in bone maturation that begins in the embryo. The classical division of fibrous dysplasia into monostotic, polyostotic and McCune Albright forms may reflect the timing of the mutation and thereby, the initial size of the mass of fibrous dysplasia precursor cells. The polyostotic form may arise in foetal life whereas the monostotic form may arise postnataally. This correlates with the evidence that the monostotic form is not a precursor of the polyostotic form. Thus fibrous dysplasia may reflect a programmed field effect of abnormal osseous development in congenitally predisposed bone matrix. This may account for the fusiform expansion of affected bone. Although the shape of affected maxilla appears to be more complex, reflecting its structure, the overall effect is similar to that seen in the mandible. The lesion, if large completely obliterates the maxillary sinus. The above pattern is altered if the fibrous dysplasia undergoes cystic degeneration, then the affected part may lose its anatomical shape and becomes spherical. Aneurysmal bone cyst and simple bone cyst are well defined entities that sometimes occur as secondary phenomenon in many benign and malignant bone tumours. Secondary cystic lesions consisting of blood filled cavities in bone that are lined by a thick layer of fibrous tissue termed as nonspecific cystic degeneration have also been reported. The development of cystic degeneration in fibrous dysplasia can pose a diagnostic and therapeutic dilemma as it may present clinically as a rapidly enlarging mass that can be alarming to both patient and physician. Sarcomatous transformation
should always be ruled out in such clinical presentations.\[16,19,20\]
Another clinical implication of cystic degeneration, and
consequently ensuing rapid growth in a relatively quiescent
fibrous dysplasia, is alteration in the management protocol
necessitating early surgical intervention.\[16\] The affected part
may lose its anatomical shape and become spherical, thus may
appear more cosmetically deformed. However, this is a first
case of its kind reporting an epithelial lined cyst in fibrous
dysplasia occurring with chronic suppurative osteomyelitis.

Maxillary osteomyelitis occurs rarely in a healthy host and
fibro osseous lesions\[21\] particularly fibrous dysplasia,\[22\] which
is considered as one of the local factors that predispose to
this type of infection. Chang et al.\[23\] have described a case
of fibrous dysplasia with chronic osteomyelitis of mandible.
Osteomyelitis complicating the fibrous dysplasia affected frontal
bone subsequent to trauma has been reported in the literature.\[24\]
Sequestrum formation has been reported in fibrous dysplasia
affecting tibial bone without any clinical or pathological
evidence of osteomyelitis,\[25\] but never in maxillofacial region.
Increased predisposition may be due to vascular compromise
subsequent to replacement of bony medullary cavity with
immature fibrocellular tissue having relatively poor vascularity.

Presence of a carious exposed tooth in such setting may initiate
chronic inflammatory response which may cause:
1. Stimulation of epithelial rests of malassez resulting into
initiation of a radicular cyst.
2. Propagation of an inflammatory response which may
cause the usual sequelae of abscess formation and
discontinuity of cyst lining.
3. Presence of chronic foci of infection leading to
osteomyelitis and sequestrum formation.
4. Persistence of chronic infection resulting into
enlargement of cyst lining, granulation tissue and
eventually engulfing the sequestrum.

Simultaneous occurrence of three different pathologies
viz. fibrous dysplasia, epithelialised inflammatory cyst and
osteomyelitis with sequestrum formation at one site is an
exceptional and unique pathologic event. It is vital to sort
the time frame and sequence in which these three must have
occurred to understand the pathogenesis. Any one of these
could have been the primary event leading to rest which means
that there are three possible series of events.

1. Carious exposed tooth as the first event leading to
radicular cyst → formation abscess formation → initiation
of Garre’s type of response with fibrous dysplastic bone.
2. Existing craniofacial fibrous dysplasia with carious exposed
tooth leads to formation of radicular cyst → abscess
formation and rupture of cyst lining → osteomyelitis with
sequestration and engulfing by the cyst
3. Existing fibrous dysplasia → carious exposed tooth with
periapical infection → osteomyelitis with sequestration and
sinus tract → epithelial ingrowth encircling the sequestrum

The probability of a radicular cyst occurring as the primary
event in a pre-existing fibrous dysplasia is much higher
(hypothesis 2) because the cyst lining was attached to the
tooth roots as noted intraoperatively and in preoperative CT
scans. The reverse sequence of osteomyelitis and sequestrum
formation as the primary event with its surrounding
granulation getting epithelialised would lead to a cyst which
will be entirely disassociated from the carious teeth. Both these
hypotheses are based on the presumption that maxilla was
affected by fibrous dysplasia before the tooth became carious.
This is supported by the patient’s history and the extent of
fibrous dysplasia involving maxilla and zygoma and not just
confined to alveolar region (Garre’s type response). Thus, it
appears to be a unique scenario in which there is occurrence
of radicular cyst in the bone affected by fibrous dysplasia,
and osteomyelitis develops subsequently as a consequence
of altered vascularity and chronic infection. The sequestrated
bone eventually gets engulfed by the disintegrated cystic
lining.

Recurrence has been reported following surgical removal
of non epithelial lined bone cyst occurring in fibroosseous
lesions and prudence dictates similar possibility with its
epithelialised counterpart. Thus follow up is advised to
detect any recurrences and regrowth of fibrous dysplasia.\[3,10\]
Rapid growth in such a dysplastic bone should raise suspicion
about sarcomatous or cystic transformation thus necessitating
prompt intervention.

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