Abstract:
Craniofacial fibrous dysplasia especially the polyostotic form is an uncommon condition. Its management is further complicated by the rarity of comprehensive reviews though databases are replete with case reports. Here we report one such case and review the current relevant literature.

Keyword: Polyostotic Craniofacial Fibrous Dysplasia, Polyostotic dysplasia, Fibrous dysplasia

Polyostotic Craniofacial Fibrous Dysplasia- Rare Case Report and Review Of Literature

Introduction
Fibrous dysplasia (FD) is a benign, sporadic skeletal disorder that predominantly involves craniofacial bones. It accounts for up to 10% of all skeletal tumors. It can involve in a monostotic or polyostotic form. It can present in combination with precocious puberty and hyperthyroidism and is known as the McCune Albright syndrome. The monostotic form is more common in the 20 to 30 years age group while polyostotic form occurs in children younger than 10 years of age. The bony lesions tend to grow with the child and stabilize after puberty. Here we present a case of polyostotic craniofacial fibrous dysplasia.

Case report
A 22 year old gentleman came with complaints of swelling over his face and skull since childhood. It started as a small swelling over the right side of the face 12 years ago and has gradually progressed to the present size of about 8x5cms. He does not give any history of pain related to the swelling. He also complained of a similar swelling over the right side of the face 12 years ago and has gradually progressed to the present size of about 8x5cms. He does not give any history of pain related to the swelling. He also complained of a similar swelling over the right side of his scalp. Examination revealed a diffuse, non tender, bony swelling present over the right zygoma extending onto the maxilla and fronto-zygomatic buttress area. Another similar 6x7cms swelling was noted in the right temporal region. There was minimal hypertrophy of the right
side of mandible. Occlusal canting was present and facial nerve examination was normal. Vision and eyeball movements were normal. CT scan revealed bony lesions with expansion and ground glass appearance of the right hemimandible, the right maxilla, the right pterygoids, the right greater and lesser wings of sphenoid, the posterior portion of the right frontal bone, the squamous portion, the mastoid and a part of the petrous temporal bone, sparing the petrous apex. The right globe was mildly displaced anteriorly due to bony expansion of the orbital walls.

Figure 1: Pre-operative photograph.

Figure 2: Coronal section of CT scan showing the lesions over right temporal bone and maxilla. Figure 3: Axial section of CT scan showing the lesions involving right maxilla and zygoma.

Surgical facial contouring (Partial excision) was planned. The right zygoma, maxilla, frontal and temporal bones were exposed with right lower eyelid subciliary incision, bicoronal incision and right upper gingivobuccal sulcus incision. The bony swellings over right
temporal region and lesions over frontal bone, maxilla, zygoma including zygomatic arch and infra-orbital rim were contoured by using burrs and oscillating saw. Around 3x3x4cms of right temporal bone swelling, 2x3x3cms each of right zygoma and maxillary bones were excised. Postoperatively adequate symmetry was achieved.

Figure 5: Post-operative photograph

Discussion:

The etiology of fibrous dysplasia though not completely elucidated is found to follow a missense mutation in the gene GNAS1 on chromosome 20. It encodes the alpha subunit of the stimulatory G protein-coupled receptor, Gs. The systemic manifestations of the mutated Gs protein manifest in the bone through parathyroid hormone receptor and in skin through melanocyte-stimulating hormone receptor. Microscopically irregular trabeculae of woven bone are seen blending into the surrounding normal bone. They stay within a cellular fibrous stroma with osteoblast progenitor cells resembling fibroblasts. This picture is said to resemble Chinese script writing. Clinical it presents with bone pain, pathological fractures and bone deformities. The craniofacial bones are affected in about 10% of cases of monostotic FD and in 50% to 100% of cases of polyostotic FD. Any craniofacial bone can be affected by FD. The clinical features depend upon the bones affected. Signs and symptoms include facial pain, headache, cranial asymmetry, facial deformity, tooth dislodgement, and visual or auditory impairment.

Diagnosis of FD is based on clinical, radiographic or histological criteria. The radiological features of FD are non-specific and vary widely. The typical appearance is that of radiolucent lytic lesions with a homogenous ground-glass appearance and ill-defined borders. Its nonspecific radiological appearance makes it difficult to differentiate from other skeletal lesions like Paget’s disease or ossifying fibroma. CT scan is essential to define the extent of involvement and to rule out optic nerve involvement.

Treatment of FD consists of either conservative contouring or radical excision with reconstruction. The choice of surgery depends on the site of involvement, rate of growth, aesthetic implications, functional disruption, patient preference and surgeon’s experience. Optic nerve decompression has generally been advised; especially in those patients with decreasing visual acuity. The value of therapeutic decompression is as controversial as the need of prophylactic decompression in asymptomatic cases. Studies have shown that vision is less likely to return if the decompression is done more than one week after blindness. Visual loss has been proposed to be due to a mass lesion rather than optic canal stenosis. Surgical excision or contouring is aimed to manage the mass effects of expansion and the consequent facial deformity. In a majority of cases,
surgery of the craniofacial skeleton comprises contouring. Rarely resection and reconstruction may be needed. Contouring surgery though is a more limited operation; there is a higher likelihood of recurrence. The excision on contouring or complete resection is made based on the rate of growth, aggressiveness and site of the lesion. Resection is followed by reconstruction with either alloplastic materials or bone autograft. The use of autologous tissues like calvarial bone and rib is preferable. The inner cortex is used as the graft while the outer cortex is placed back to its original position. Microvascular free flap reconstruction has a role, especially for lesions involving the mandible where segmental excision is necessary. Medical management has a role in FD. Various drugs have been used with varying success, prominent among them being bisphosphonates, vitamin D and steroids. The lack of objective evidence of their success makes it harder to evaluate results.

References:


