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CLINICAL RECORD

Fibrous dysplasia of the nasal bone: case reports and literature review

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Abstract

Introduction: Craniofacial fibrous dysplasia commonly affects the larger mandible and the maxillary bones. Although involvement of the frontal, temporal and sphenoid bones has been described, fibrous dysplasia of the nasal bone has not been previously described; the two cases reported here have been made rarer by their isolated involvement. Clinical management is dependent on disease activity and patient symptoms.

Objective: To present two cases of isolated fibrous dysplasia of the nasal bone: a 46-year-old woman with gradual widening of the nasal bridge and a 47-year-old man with an incidental finding of a nasal bone mass.

Method: Two case reports.

Results: The 46-year-old woman underwent excision of the lesion while the 47-year-old man opted for watchful waiting.

Conclusion: We have presented the first case reports of fibrous dysplasia of the nasal bone. The care of these patients should be customised to their needs and wishes.

Key words: Nasal Bone; Fibrous Dysplasia of Bone; Diagnosis; Pathology

Introduction

Fibrous dysplasia is a benign skeletal disease resulting in bony expansion due to the abnormal development of fibrous tissue. It can affect single (monostotic) or multiple (polyostotic) bones. Craniofacial bones are affected in approximately 10 per cent of monostotic fibrous dysplasia cases. This rises to 50–100 per cent in polyostotic fibrous dysplasia.[1] Fibrous dysplasia is an indolent bone disorder. Common sites of involvement in the head and neck are the maxillary bone and the mandible, resulting in craniofacial deformity.

Other symptoms can range from headache, facial pain and auditory impairment to visual loss due to optic nerve compression. Rarely, fibrous dysplasia may be associated with endocrinological abnormalities, particularly if the pituitary gland is involved. McCune–Albright syndrome, a variant of fibrous dysplasia, is suspected when two of the following three features are present: endocrine dysfunction, polyostotic fibrous dysplasia, unilateral café-au-lait spots.[2] Most commonly, the diagnosis of fibrous dysplasia is made radiologically, with the classical ground-glass appearance seen on computed tomography (CT) imaging. Fine needle aspiration cytology has been shown to be a useful diagnostic adjunct in lesions around the sinonasal region, mainly to exclude a more sinister pathology.[3,4]

While fibrous dysplasia has been shown to affect a number of bones in the craniofacial region, involvement of the nasal bones in isolation has never been described. We describe two such cases and highlight the different management options.

Case report 1

A 46-year-old woman was referred by her general practitioner after they noticed a gradual widening of her nasal bridge over the last few years. There was no history of trauma and the patient did not describe any rhinological or neurological symptoms.

Examination confirmed asymmetrical nasal bones, with a solid, firm mass on the right side. Computed tomography of her facial bones demonstrated fibro-osseous change in the right nasal bone (Figure 1). She was initially managed expectantly with serial imaging; however, the lesion began to grow after several months and she opted for surgical intervention. An external incision was made intra-operatively on the edge of the nasal bone, revealing spongy bone. A microdebrider was then used to smooth out the nasal bones. A cartilage construct, harvested from the septum, was then used to fill the defect and this was fixed with a vicryl suture threaded through the skin. Histology confirmed a benign fibro-osseous lesion, consistent with the radiological diagnosis of fibrous dysplasia. The patient has remained well post-operatively, with no evidence of recurrence at the one-year follow up.

Case report 2

A 47-year-old man presented to the ENT clinic following an incidental CT finding, which was requested as part of the diagnostic work-up for dental implantation. This showed a small, bony and radiopaque mass attached to the lateral aspect of the left nasal bone. The mass measured approximately 11 × 8 × 6 mm, blended with the nasal bone surface and showed a faintly trabeculated pattern (Figure 2). Radiological findings were consistent with fibrous dysplasia. As there were no rhinological symptoms, a policy of watchful waiting was adopted. After four years, there remains no evidence of growth of the lesion.

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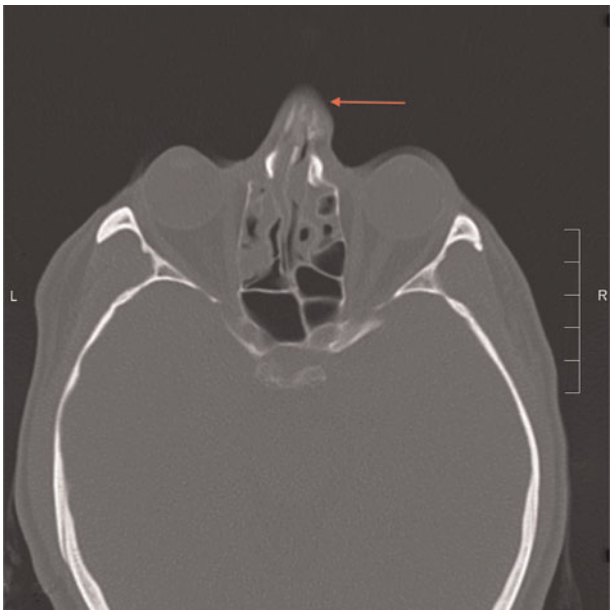

FIG. 1
Axial computed tomography scan demonstrating fibro-osseous change in the right nasal bone (L = left side; R = right side).

Discussion

Fibrous dysplasia is an uncommon, benign disorder characterised by a tumour-like proliferation of fibro-osseous tissue. It is a genetic disease, caused by activating mutations of the *GNAS complex locus* gene on chromosome 20, resulting in the somatic activating mutation of the $G_s\alpha$ subunit of a G protein-coupled receptor, with upregulation of cyclic adenosine monophosphate.[5]

Commonly, the diagnosis of fibrous dysplasia is made radiologically and it is then correlated with clinical and pathological findings. The radiological features of fibrous dysplasia are diverse and are dependent on the proportion of mineralised bone to fibrous tissue in the lesion. Maturing fibrous dysplasia lesions acquire a mixed radiolucent/radiopaque appearance; established fibrous dysplasia exhibits mottled radiopaque patterns resembling ground glass – small, diffusely distributed opacities.[1]

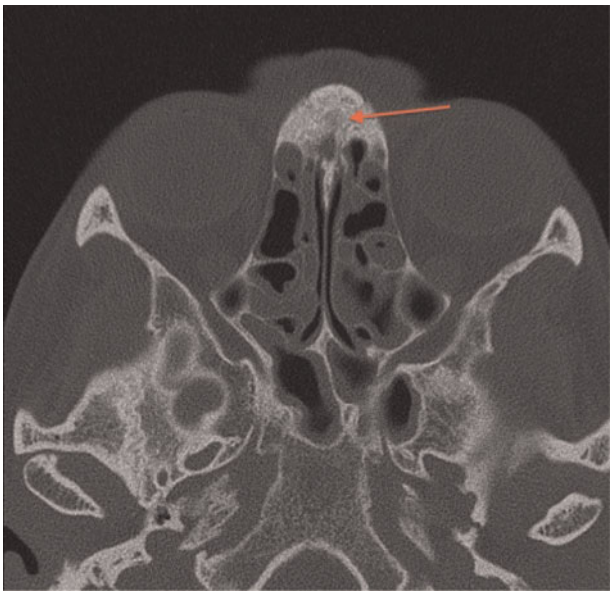

FIG. 2
Axial computed tomography scan demonstrating a small, bony, radiopaque mass attached to the lateral aspect of the left nasal bone, blending with the nasal bone surface and showing a faintly trabeculated pattern.

Unlike fibrous dysplasia elsewhere in the axial skeleton, the margins of craniofacial fibrous dysplasia are ill-defined, often blending with normal adjacent bone. Histologically, fibrous dysplasia is composed of interconnected trabeculae of mixed woven and lamellar bone in fibrous stroma, and the lesional tissue is usually in continuity with the surrounding, normal cortical bone.[6,7]

In monostotic fibrous dysplasia, 75 per cent of patients present with lesions in the ribs, femur, tibia or skull. The polyostotic form affects the proximal femur, craniofacial bones, tibia, humerus and ribs in decreasing order of incidence.[8] The mandible and the maxillary bone are the most commonly affected craniofacial bones. Involvement of other craniofacial bones, such as the frontal, temporal, sphenoid, parietal and orbital bones have been described.[9]

Fibrous dysplasia is often managed conservatively, but surgical intervention is advocated when complications occur, e.g. visual impairment, cosmetic deformity, mucocele, haemorrhage and aneurysmal bone cysts.[10,11] A recent meta-analysis comparing watchful waiting versus surgery found that optic nerve decompression is the preferred treatment in patients with symptomatic ophthalmic disease.[10] Surgery, however, is challenging since these lesions are highly vascular and excision should not be undertaken lightly and without adequate expertise. Some authors have advocated lifelong follow up in view of the continuous evolving nature of certain lesions, although the natural history of fibrous dysplasia is that it tends to 'burn out' in the third and fourth decade.[12]

In the case reports described here, both patients had isolated fibrous dysplasia of the nasal bone that has never been described in the literature. These two small, oblong bones, which form the bridge of the nose, articulate with four other bones: two of the cranium, the frontal and ethmoid; and two of the face, the opposite nasal bone and the maxilla. Fibrous dysplasia of the larger frontal and maxillary bones has been frequently described and there have been a small number of cases described involving the ethmoid bones. The general consensus on management is to offer regular follow ups to ensure that the lesion does not increase in size, recurs or involves other bones as it can cross suture lines.

- **These are the first described case reports of fibrous dysplasia of the nasal bone**
- **Diagnosis was based on clinical, radiological and pathological correlation**
- **Treatment was based on patient symptom(s) and disease progression**
- **Regular, long-term follow up is advocated due to the risk of growth or recurrence**

Other indicators of surgery may be persistent sinonasal symptoms, such as headache, nasal obstruction and pain, secondary to involvement of the paranasal sinuses.[13] When the temporal bone is involved, a patient may present

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with 80 per cent hearing loss because the inner-ear canal narrows. Further, it may cause facial nerve paralysis or vertigo.

There is continual research into the role of medical therapy. Treatment with bisphosphonates is predicated on the rationale that in fibrous dysplasia, there is an increase in interleukin-6-induced osteoclastic bone resorption, similar to another osteoclast-driven condition, osteitis deformans (Paget's disease of bone).[14] Various reports on the use of pamidronic and alendronic acid have shown significant reduction in bone pain and bone turnover.[15,16]

Conclusion

To the best of our knowledge, we have described the first isolated case series of fibrous dysplasia of the nasal bones. Diagnosis relies on a combination of clinical, radiological and pathological correlation. Similar to fibrous dysplasia of other craniofacial bones, treatment options depend on clinical presentation and patient choice. Regular, long-term follow up of these patients is advised as these lesions evolve and have the potential for recurrence.

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Mr G Wong takes responsibility for the integrity of the content of the paper
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