Case 14456

Eurorad

Maxillary fibrous dysplasia

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Section: Head & neck imaging
Area of Interest: Head and neck Ear / Nose / Throat
Procedure: Diagnostic procedure
Imaging Technique: Digital radiography
Imaging Technique: CT
Imaging Technique: MR
Special Focus: Connective tissue disorders Case Type:
Clinical Cases
Authors: La Pietra P, Molinari M, Sommario M, Cavicchi M, Scarcella E
Patient: 35 years, male

Clinical History:

A 35-year-old male patient complained for several days of gradually increasing right cheek pain.
His dentist ruled out dental problems and performed a dental panoramic radiograph that showed a radiopacity in the right maxillary sinus.

Imaging Findings:

The dental panoramic radiograph demonstrated a faintly radiopaque lesion in the right maxillary sinus. In order to obtain a better definition of the lesion, an unenhanced CT scan and MRI were performed.
The unenhanced CT scan showed mild hypoplasia of the right maxillary sinus, which is almost entirely filled by a hyperdense lesion with thickening of the medial and superior walls of the sinus.
MRI shows low signal intensity of the lesion on all sequences.
Based on these results a diagnosis of maxillary fibrous dysplasia was made and confirmed on histology.

Discussion:

Fibrous dysplasia (FD), also known as Lichtenstein-Jaffe disease, is a bone developmental anomaly due to a genetic defect in the osteoblastic differentiation caused by mutation in the GNAS1 gene and characterized by a focal replacement of normal bone with fibrous stroma and islands of immature woven bone [1, 2].
According to the involvement of one or multiple bones, it is classified as monostotic or polyostotic form.
It can affect any bone, but most often the ribs, femur, tibia, fibula and facial bones. Involvement of facial bones is most common in polyostotic forms and accounts for 30% of monostotic cases [3].
The polyostotic form can be associated with endocrinopathies, skin pigmentation and precocious puberty (McCune-Albright syndrome).
FD typically occurs in teenagers or young adults but is not exclusive to this age range, moreover both sexes are equally affected [4, 5]. Symptoms depend on the site and size of lesions: small monostotic lesions may be asymptomatic and identified incidentally.
Common signs in symptomatic cases are bone pain, swelling, deformity and pathological fractures.
In craniofacial locations, slow growth can lead to facial asymmetry, deformity and compression of adjacent structures leading to visual impairment, exophthalmia, strabismus, hearing loss, anosmia, headache, repeated sinus infections and cranial nerve palsies.
Imaging is fundamental in the assessment of extension and localization of the disease. The radiological appearance

depends on the ratio between fibrous and bony tissue within it; predominantly fibrous tissue produces a radiolucent lesion whilst a higher quantity of bony tissue produces a denser lesion. Their characteristic ground-glass appearance is due to the mixture of tissue constituents [5].
CT has a key role in pre-operative planning and follow-up.
MRI typically shows intermediate or low signal intensity on T1-weighted images and intermediate or high signal on T2-weighted images with heterogeneous contrast-enhancement after gadolinium administration [6, 7]. These nonspecific findings are not particularly useful in differentiating the lesion from other tumours; a potential pitfall. Bone scan shows tracer uptake and may be useful to differentiate between monostotic or polyostotic forms. Medical therapy has a limited role in the management of FD.
Small asymptomatic lesions do not require surgical resection but the patient may require follow-up with CT to monitor disease progression.
Surgery remains the mainstay of therapy for symptomatic forms to prevent pathological fractures, control pain and reduce bone deformities.
Sometimes the presence of important adjacent structures can hamper radical excision; more conservative surgery risks recurrences, especially during growth periods [8, 9]. Malignant transformation is very rare.
Differential Diagnosis List: Monostotic maxillary fibrous dysplasia, Cemento-ossifying fibroma, Paget disease, Sclerotic metastases, Ameloblastoma, Silent sinus syndrome

Final Diagnosis: Monostotic maxillary fibrous dysplasia

References:

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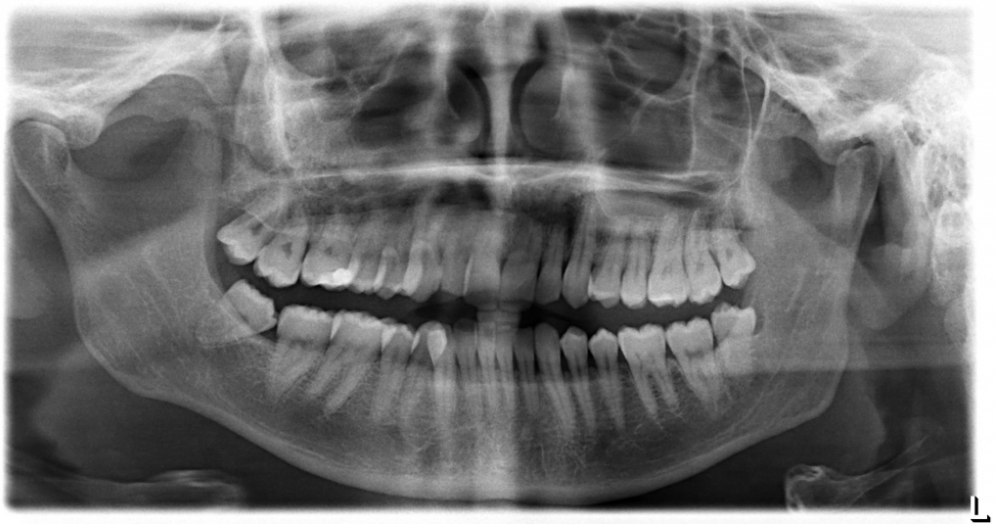
Figure 1

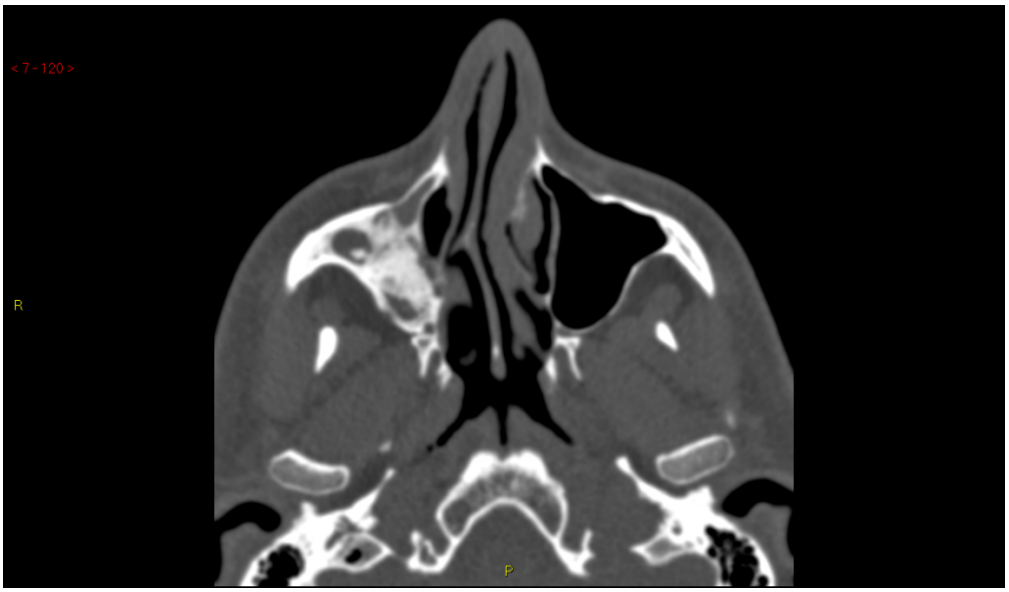
a



Description: Panoramic radiography shows normal appearance of teeth and a large radiopaque structure in the right maxillary sinus. **Origin:** Image owned of the patient

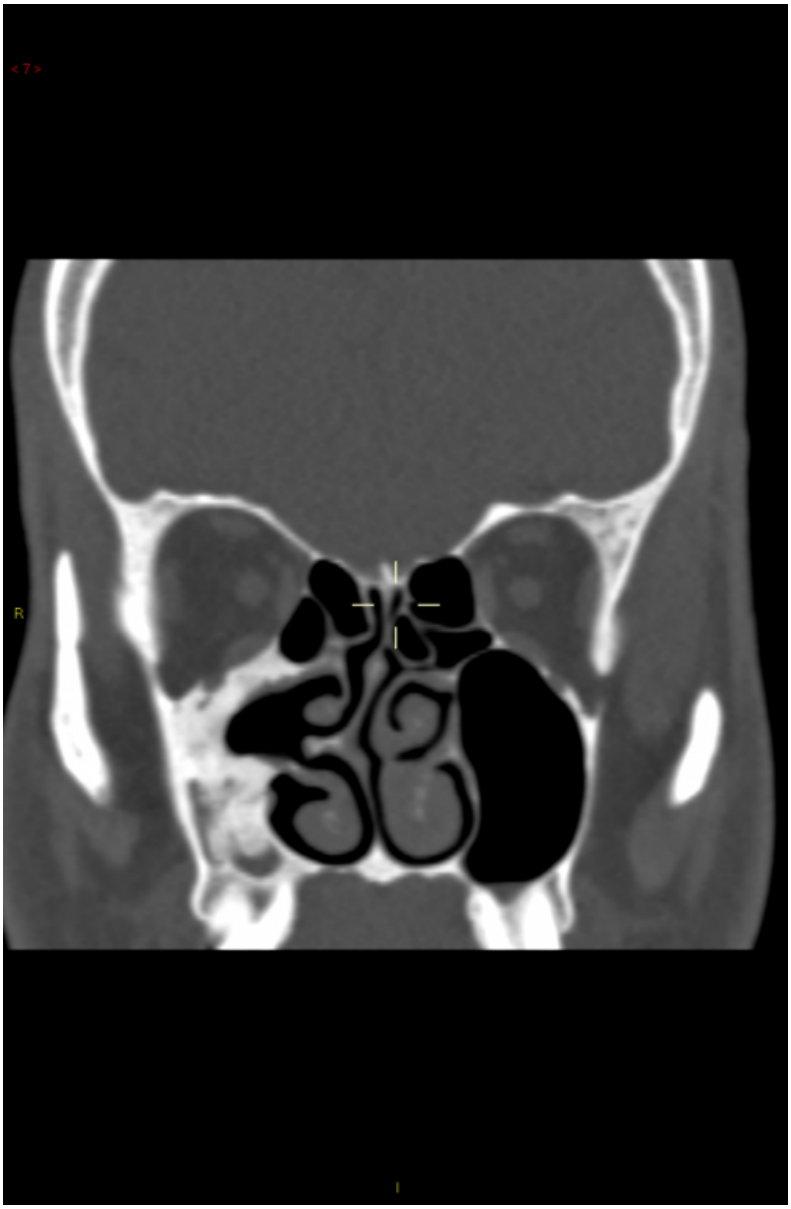
Figure 2

a



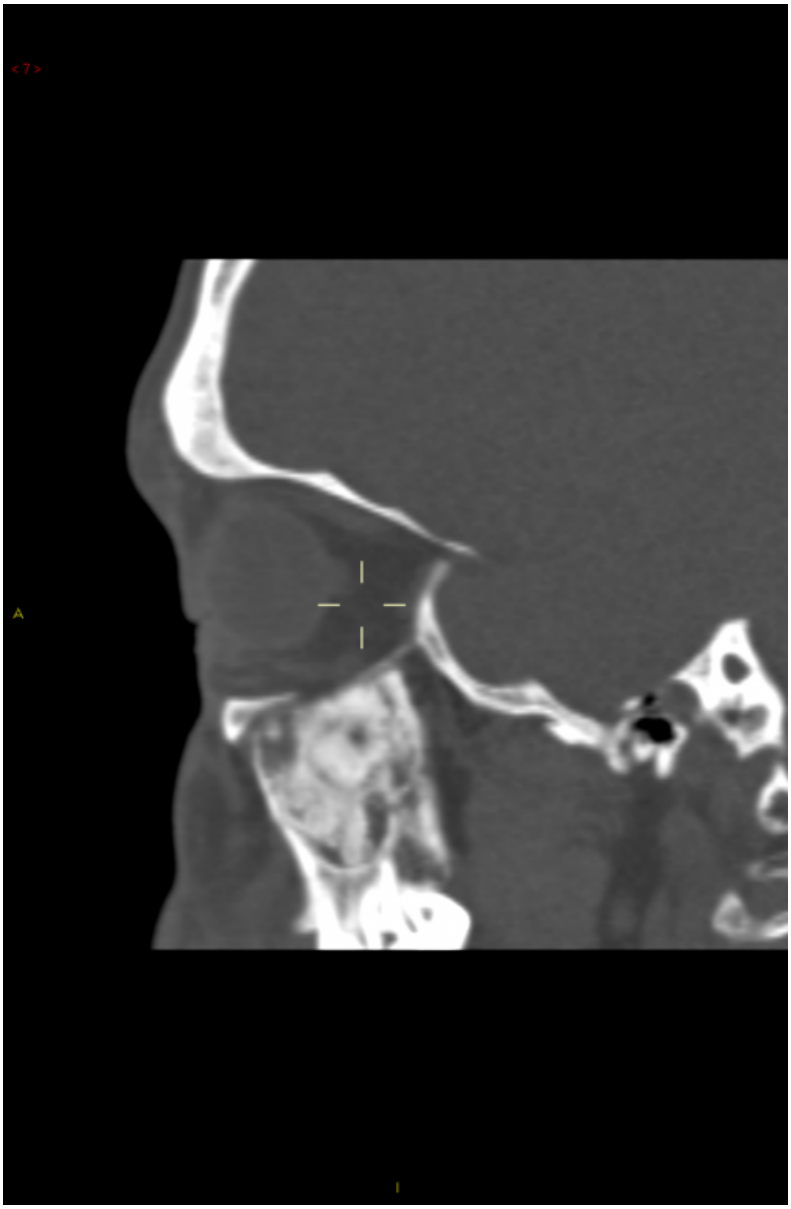
Description: Unenhanced CT scan (axial view) shows: hypoplasia of the right maxillary sinus, almost entirely filled by a hyperdense lesion. **Origin:** Image origin: U.O.C radiologia area nord AUSL BOLOGNA

b



Description: Unenhanced CT scan (coronal view) shows: hyperdense thickening of the walls of right maxillary sinus and consequent hypoplasia. **Origin:** Image origin: U.O.C radiologia area nord AUSL BOLOGNA

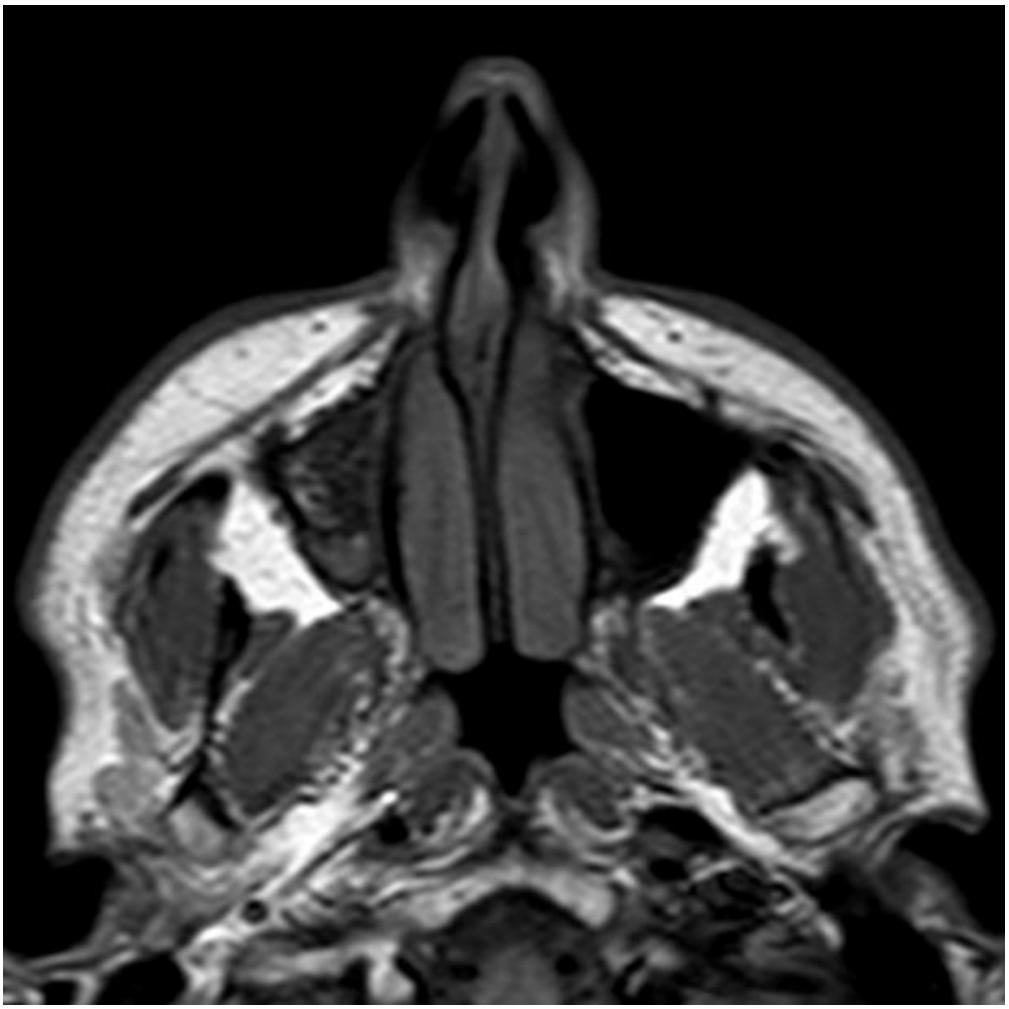
c



Description: Unenhanced CT scan (sagittal view) shows: the right maxillary sinus is almost entirely filled by the lesion. **Origin:** Image origin: U.O.C radiologia area nord AUSL BOLOGNA

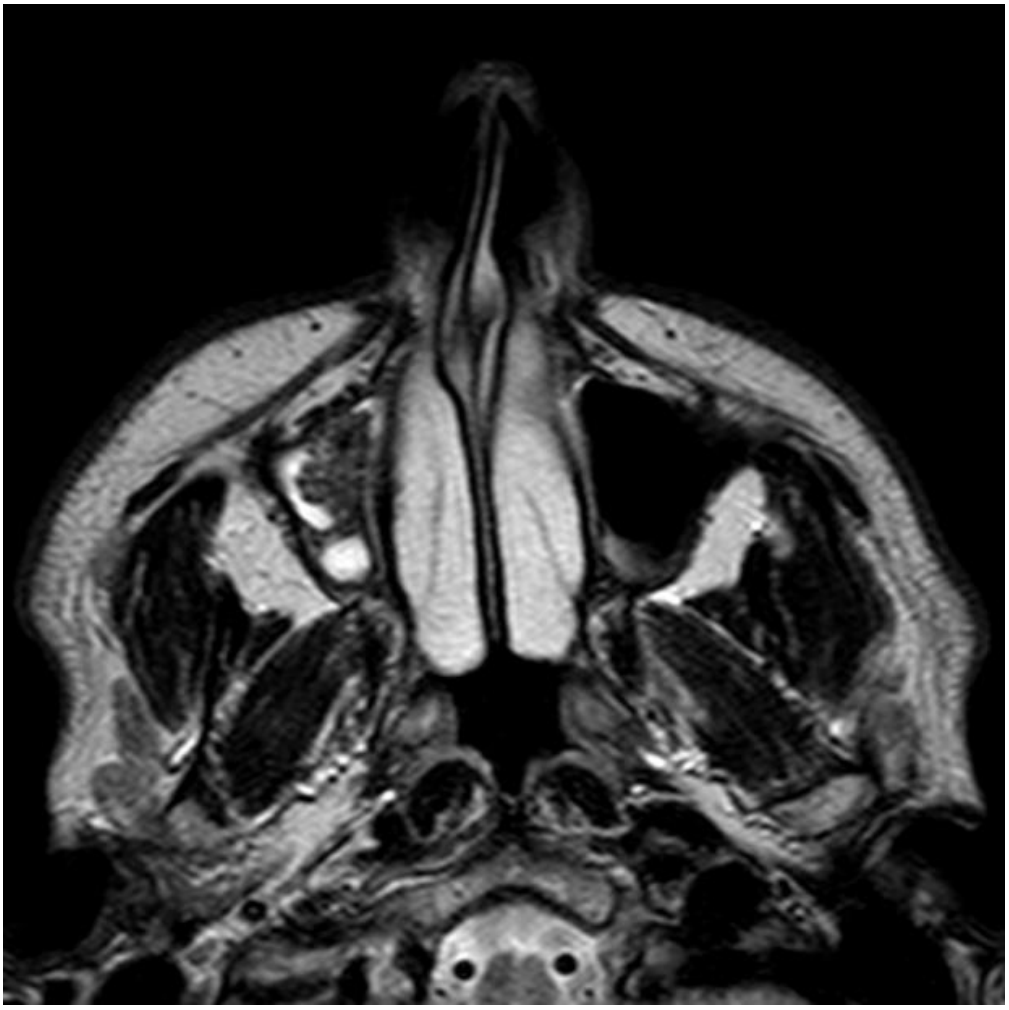
Figure 3

a



Description: Axial unenhanced T1-weighted MR image shows hypoplasia of the right maxillary sinus filled by a heterogeneous low-signal-intensity lesion. **Origin:** Image owned of the patient

b



Description: Axial unenhanced T2-weighted MR image shows a low signal intensity of the lesion and a small peripheral amount of fluid. **Origin:** Image owned of the patient