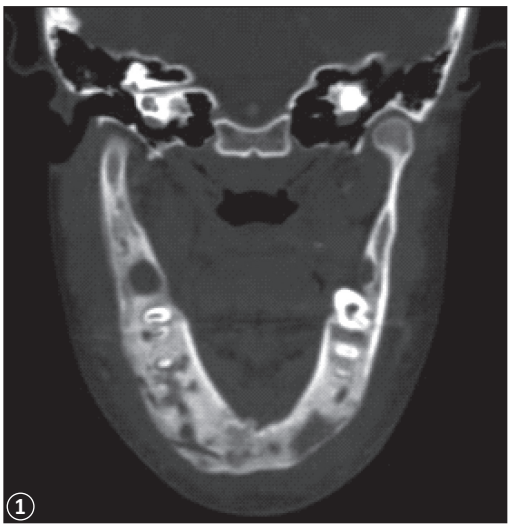
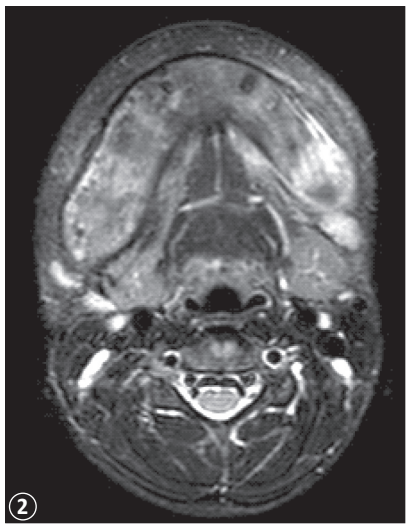
Fibrous Dysplasia of the Mandible in a Pediatric Patient





Our patient was an 11-year-old male with submandibular cellulitis and a history of fibrous dysplasia (FD) in the mandible. A maxillofacial CT scan was performed (Image 1) and showed osseous expansion and a ground glass appearance within the right mandibular body, parasymphysis, and symphyseal regions. These findings are in keeping with FD of the right mandible. There was also overlying soft-tissue swelling present.

A follow-up MRI exam (Image 2) showed bone expansion within the mandible, as well as low T1 and intermediate T2 signal intensity without associated enhancement. These findings were concordant with those of prior study. In addition, there were signs of soft tissue inflammation. Because of the abnormal bone characteristics, a diagnosis of superimposed osteomyelitis could not be entirely excluded in this study.

FD is a disorder that was first described in 1938 by Lichtenstein (1). While it was previously considered to be a benign chronic neoplasia, it is now considered to be a hamartomatous bone disorder associated with mutations in gene GNAS I, and which affects the proliferation and differentiation of preosteoblasts (2). These factors affect the bone metabolism and result in the replacement of components within the normal cancellous bone with abnormal fibrous tissue. This abnormal bone commonly has the appearance of ground glass on radiographs.

There are 3 major types of fibrous dysplasia: monostotic FD (involving a single bone), polyostotic FD (multiple lesions in multiple bones), and McCune–Albright syndrome (a polyostotic form of FD with associated endocrine abnormalities) (3). The monostotic form is the most common, accounting for about 70% of all cases. The average age of diagnosis in patients with FD is 10 years (1), and the disease usually becomes dormant by the third decade (4).

Cranial or facial bones are affected in about 30% of all cases (3), and the disorder usually affects the mandible (1). FD is usually asymptomatic, but, associated with osseous encroachment of canals and foramina, it may be a cause of pain and discomfort as well as of restricted facial movement (4). In infancy, FD must be differentiated from cherubism, which presents as painless bilateral swelling around the mandible (1). On rare occasions (0.4 – 3.1%), FD may be associated with malignant changes; when present it is usually in patients older than 30 with craniofacial FD accompanied by the rapid onset of pain and swelling (5). Conservative management involving shaving diseased bone via an intraoral approach management has been the standard of care in cases of facial FD (2).

References

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