Craniofacial fibrous dysplasia
Report of a case with diverse radiological spectrum

Silky Rajesh Punyani1
Saurabh Srivastava2
Vishal Ramesh Jasuja3

1 Department of Oral Medicine & Radiology, People’s Dental Academy, Bhopal, India
2 Department of Oral Medicine & Radiology, Babu Banarsidas College of Dental Sciences, Lucknow, India
3 Department of Anatomy, People’s College of Medical Sciences & Research Centre, Bhopal, India

Address for correspondence:
Silky Rajesh Punyani, MDS
C-14, Senior MIG, PCMS Campus, People’s University
Bharapur bypass road, Bhopal-462037, India
E-mail: dr.srpunyani@gmail.com

Summary

A young male of Asian-Indian ethnicity reported with a complaint of a painless, slow growing swelling over the left side of lower jaw. A thorough clinical history was taken and detailed radiological exam performed. The conventional radiographic examination revealed a mixed radiolucent-radiopaque lesion with unique appearances on different radiographs. Additional computed tomographic examination discovered the involvement of several bones in the skull base. Subsequent to histopathological confirmation a final diagnosis of craniofacial fibrous dysplasia was made. This case is particularly unique and of didactic importance as well because the various textbook descriptions for radiological appearances of fibrous dysplasia were found in the same case.

KEY WORDS: craniofacial fibrous dysplasia; ground glass; cotton wool.

Introduction

Fibrous dysplasia (FD) is a slowly progressive, benign fibro-osseous condition characterized by replacement of normal bone by cellular fibrous tissue and irregular bony trabeculae. The involvement of cranial bones is seen in 10 to 27% in the monostotic form (single bone) and 50% in polyostotic (multiple bone) cases (1). The craniofacial variety of fibrous dysplasia is a localized form of this pathology characterized by confluent involvement of adjacent bones of the cranium and skull base. Though a benign process, involvement of the craniofacial skeleton can pose the problems of significant esthetic and functional disturbances. Depending on its location, the signs and symptoms can vary from an asymptomatic deformity to more grave consequence like vision changes, hearing impairment and nasal obstruction (2). This paper reports an interesting case of craniofacial fibrous dysplasia diagnosed incidentally after radiological investigations were performed for an asymptomatic mandibular swelling. Another noteworthy observation was the different radiological appearances seen on different radiographs.

Case report

A 19-year-old Indian male patient reported with a complaint of a slowly progressive swelling in the front region of the lower jaw on the left side since 8 years. The swelling was insidious in its onset and gradually increased over a period of eight years to the presenting size. The patient declined to history of any previous toothache and trauma to the affected site. The review of systems was non-contributory. The past medical and dental histories were unremarkable. Upon examination the patient was moderately built and had a normal intellect. The extra-oral examination revealed an appreciable facial asymmetry due to the presence of a diffuse swelling over the left parasymphysis region (Figure 1). Antero-posteriorly, the swelling extended from the left parasymphyseal region to 2 cm beyond the angle of the mandible on the left side. It’s supero-inferior dimensions extended from the mid-ramus region to the inferior border of the mandible on the left side. The swelling was firm and nontender. The submandibular lymph nodes were not enlarged. The patient was completely asymptomatic and there was no associated pain. The mandible was not deviated on protrusion. The tongue was not deviated. The occlusion was normal with no overjet or overbite. The patient’s general health was good and there was no history of any systemic disease.

Figure 1 - Extra-oral photograph showing diffuse swelling over the left side of mandible.
Ling was hard to firm in consistency and non tender on palpation. Further, intra-oral examination revealed a discrete, bony hard swelling extending from 41 to 35 region causing obliteration of the left buccal vestibule (Figure 2). The swelling caused drifting of the involved 33 mesially and 34 distally. Based on the history and clinical characteristics a working diagnosis of a benign fibro-osseous lesion was made.

Investigations that were advised for the patients were serum alkaline phosphatase level and radiographs including intra-oral periapical radiograph (IOPA), mandibular cross-sectional occlusal radiograph, panoramic radiograph and serum alkaline phosphatase levels. The serum alkaline phosphatase level was within the normal range (95U/L; normal range-25-100U/L). The IOPA of the involved region revealed altered trabecular pattern of the alveolar and basal bone and localized granular appearance of the bone including a single ill-defined lytic area, giving the characteristic “ground glass” appearance (Figure 3). The panoramic (Figure 4) and lateral oblique (Figure 5) radiograph of the body of the mandible showed an ill-defined mixed radiolucent radiopaque lesion merging gradually into the adjacent normal bone. The trabeculae appeared hazy and the lesion lacked a distinct defining margin giving the “cotton wool” appearance. The panoramic radiograph also revealed an increased height of the mandibular basal bone on the left side and increased width of the ramus, along with superior displacement of the inferior alveolar canal as compared to the other side. Concentrally arranged thin bony trabeculae, the “thumb-print” appearance was evident on the cross-sectional mandibular occlusal radiograph (Figure 6). To ascertain the precise extent of the pathology a plain computed tomographic scan was advised. The CT scans were performed on a multislice spiral CT unit (Somatom Definition Edge; Siemens, Erlangen, Germany). The exposure parameters were tube voltage-120 kV, tube current-270 mA, and slice thickness-1 mm. The axial section CT image of the mandible showed expansion of the body of the mandible with few lytic areas bilaterally but greater in degree on the left side and expansion of the left ramus. Ground glass appearance of the bone was clearly appreciable (Figure 7). Another noteworthy finding evident on another axial section was a similar involvement of multiple bones of the craniofacial skeleton including the body of sphenoid, greater and lesser wing of sphenoid, squamous part of the temporal bone on the left side, left pterygoid laminum, pterygoid plates, posterior part of the hard palate, ethmoid bone on the left side, nasal septum, left zygomatic arch and posterior wall of the left maxillary sinus (Figure 8). This prompted us to perform radiographic skeletal survey to identify possible involvement of other bones. No contributory findings were found in this regard. An incisional biopsy was performed from the left body of mandible. The H & E stained decalcified section exhibited lesional tissue composed of irregularly shaped immature lamellated bony trabeculae in a loosely arranged cellular fibrous tissue stroma. Some of the scattered bony trabeculae were curvilinear in shape with the presence of osteocytes within lacunae and osteoblastic rimming at places. A final diagnosis of craniofacial fibrous dysplasia was thus arrived at. Surgical recontouring of the bone was done for cosmetic correction. The patient was informed about the involvement of other bones and possible complications that could arise in future. He was advised a neurophysician and neurosurgeon consult and regular follow up.

Discussion

Fibrous dysplasia, first described by Lichtenstein, is a benign, developmental dysplastic disorder of the bone characterized by formation of immature woven bone in lieu of normal osseous tissue (3, 4). Affliction of the craniofacial skeleton by this disease process can have a myriad of clinical manifestations and resultant complications. The common age of occurrence for CFD is childhood and adolescence, average age of diagnosis being 10 years (5). The clinical features CFD encompass a broad clinical spectrum depending upon the location and extent of the lesions. These can vary from mild facial asymmetry or a painless “lump” to serious functional deficits owing to progressive expansion of
the craniofacial lesions (6). The ocular effects of FD are particularly worrisome. The involvement of the sphenoid and/or ethmoid bones can cause displacement of the eyeball. Epiphora and loss of vision may occur secondary to compression of the optic nerve by expansion of the sphenoid bone. Thereby in cases of optic canal involvement, a thorough ophthalmologic examination is warranted, including testing for visual acuity, visual field, color perception and visual evoked potential (7). Atypical facial pain, headache and symptoms similar to sinusitis can occur in the involvement of temporal bone, parietal bone and paranasal sinuses. Nasal obstruction and epistaxis can also occur in extreme cases (8). Cystic lesions may develop in areas affected by fibrous dysplasia, these include cystic degeneration and lesions such as aneurysmal bone cysts (9). Though this phenomenon is somewhat rare, an area of bone affected by FD undergoes rapid enlargement by cystic degeneration which can lead to grievous complications such as optic nerve compression (10). Due to its fast progressing nature, cystic degeneration may be confused with malignant transformation.

The conventional radiological features of fibrous dysplasia vary widely. Depending upon the stage of the disease the lesion may appear, lytic, mixed or sclerotic (11). As described in the case report section, “ground-glass”, “cotton-wool” or “thumb-print” appearance may be seen, all of which were appreciable in the present case. CT is the modality of choice for diagnosis and follow up as it provides superior bony detail and assesses the exact extent of involvement. It is also efficient for assessing cranial nerve entrapment and optic nerve compression (12). On CT CFD may exhibit a ground-glass pattern, a homogeneously
dense pattern or cystic variety (13). On MRI, the lesions characteristically show low signal intensity and well-demarcated borders on both T1- and T2-weighted images (14). Surgical treatment of CFD consists of conservative shaving/contouring or radical excision with immediate reconstruction depending on site and extent of involvement. Decompression of the optic nerve is done as a therapeutic procedure or prophylactic measure. Medical treatment includes the use of steroids, mainly in the treatment of visual symptoms and bisphosphonates (15).

CFD can be an imposter being diagnosed easily as an incidental finding or posing a diagnostic enigma owing to multiple complications caused by expansion of skull base lesions. The present case is unique as it revealed different radiographic appearances on the radiographs and further CT examination disclosed involvement of the skull-base. Due to the indolent nature of the disease the management should be done as conservatively as possible, with radical surgery reserved for cases exhibiting gross functional deficits.

Conflict of interest

None.

References