Case Report
Fibrous dysplasia of the inferior turbinate

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Abstract: Fibrous dysplasia (FD) is a benign skeletal disorder in which abnormally overgrowing bony lesion replaces normal bone. FD can affect one bone (monostotic form) or multiple bones (polyostotic form). The craniofacial bones are involved in about 10% of subjects with monostotic FD. However, its occurrence in the sinonasal tract is very rare. We report a case of monostotic FD developed only in the inferior turbinate in a 29-year-old woman. To the best of our knowledge, it is the second report of monostotic FD involving the inferior turbinate in the medical literature. We, therefore, report this rare case with a review of literature.

Keywords: Fibrous dysplasia, monostotic, inferior turbinate

Introduction

Fibrous dysplasia (FD) is a rare fibro-osseous lesion characterized by progressive replacement of normal bone elements with benign cellular fibrous connective tissue. FD occurs in monostotic or polyostotic forms, and the latter may be associated with McCune-Albright syndrome, which is the most severe form of polyostotic FD. Because FD has predilection for membranous bone such as femur or tibia, it infrequently occurs in the sinonasal tract, where the inferior turbinate is extremely rare bone to occur. Recently we experienced a case of monostotic FD involving the inferior turbinate. To the best of our knowledge, only 3 cases of FD involving the inferior turbinate have been previously published, two of which are polyostotic forms [1-3]. We report the case with a review of literature.

Case report

A 29-year-old woman was referred for chief complaint of chronic bilateral nasal obstruction, which started more than 5 years ago. She didn’t complain any other nasal symptoms, such as rhinorrhea, sneezing, itching, postnasal drip and hyposmia. She denied other past or family medical history. On the nasal endoscopic examination, the right inferior turbinate was enlarged and bony hard on palpation. The nasal septum was deviated to the right side. There was no polyp or discharge in the nasal cavity. No abnormal skin pigmentation could be found. Laboratory blood test including serum alkaline phosphatase and urine test were normal. A computed tomography (CT) scan showed ground-glass appearance of the right inferior turbinate with well defined borders and normal mucosal covering, which was compatible with FD (Figure 1). Bone scan revealed increased uptake only in the right inferior turbinate (Figure 2). Under the general anesthesia, the mass was partially removed via endoscopic endonasal approach leaving approximately proximal one tenth of the lesion along with its covering nasal mucosa in situ. Septoplasty was performed thereafter. The microscopic findings were typical of FD (Figure 3). There was no evidence of enlargement of the remaining inferior turbinate at 2-year follow-up.

Discussion

FD is a sporadic benign skeletal disorder and accounts for approximately 7.5% of the benign bone neoplas [4, 5]. It presents 2 basic clinical forms: monostotic (75-80%) and polyostotic (20-25%) [5, 6]. Monostotic form with the involvement of the ribs, femur and tibia is most common. Polyostotic form may be rarely compli-
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cated by café-au-lait spots and hyperfunctional endocrinopathies (McCune-Albright syndrome) or multiple soft-tissue myxomas (Mazabraud’s syndrome) [5-7]. In monostotic form, males and females are thought to be affected with equal frequency, however polyostotic form is more common in females. Approximately 50% to 100% of patients with polyostotic disease and 10% to 30% with monostotic disease have craniofacial involvement. Craniofacial FD typically presents at around 10 years of age and then progresses throughout adolescence [5]. Craniofacial FD frequently affects the maxilla and mandible, followed by frontal, parietal, and occipital bones.

Most patients are asymptomatic, and the lesions typically are found incidentally. The most common clinical sign of craniofacial FD is bone pain or bone deformity. When the anatomic spaces and foramina are constricted because of encroachment of the lesions, the patient may experience a variety of symptoms, including headache, visual loss, proptosis, diplopia, hearing loss, anosmia, nasal obstruction, epistaxis, epiphora, and recurrent rhinosinusitis [2, 8].

The feature of FD is a focal or generalized inability of bone-forming tissue to produce mature lamellar bone. Tissue is arrested at the stage of immature woven bone. Recent molecular biology studies suggest that FD is a genetic non-inherited condition caused by the missense sporadic mutation of the gene GNAS1 on chromosome 20, which encodes the alpha subunit of the stimulatory G protein-coupled receptor (Gsα) [6, 9, 10]. This mutation results in an arginine-to-histidine or an arginine-to-cysteine substitution in the Gsα subunit leading to inhibition of GTAase activity and constitutive stimulation of AMP-protein kinase A. Many organs such as bone, skin, ovaries, thyroid and pituitary glands, which have Gsα protein-coupled receptor, can be affected as a consequence of this mutation. Therefore, clinical spectrum of FD can be varied according to the stage of embryogenesis when this mutation occurred. This mutation was first identified in patients with McCune-Albright syndrome but was later demonstrated in the lesions of patients suffering from either monostotic or polyostotic FD [4, 6].

The radiological appearances are variable and depend upon the proportion of mineralized bone to fibrous tissue in the lesion [4-6]. Radiologically, characteristic appearances of FD consist of three varieties: ground-glass pattern (56%), homogeneously dense pattern (23%) and cystic variety (21%) [11]. CT scan and MRI are useful in detecting and defining neuro-
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vascular and ocular involvement. Findings on CT scan can be from radiolucent to sclerotic according to the degree of mineralization of the tissue [4, 8]. On MRI scan, the bone lesions have variable signal intensity on T2-weighted images, whereas the T1 signal is more commonly hypointense [6, 8]. Bone scan is a useful technique for identifying the distribution of lesions in patients with polyostotic disease and typically reveals increased uptake of radioisotope in the lesions, but a cold scan does not exclude the diagnosis [6]. On gross appearance, the lesions are centrally located and consist of fibrous, tan to gray grainy tissue and vary in consistency and vascularity [6, 12]. Microscopically, FD comprises irregular trabeculae of woven bone resembling Chinese characters, blending into the surrounding normal bone and lying within a cellular fibrous stroma [4, 6]. Three stages of FD can be distinguished. Acute stage has rich cellular connective tissue with mitotic figures and woven immature bone. The connective tissue in the subacute stage becomes less cellular, and fibers tend to be arranged in whorls. The bony trabeculae become thicker and lamination occurs. In the chronic stage, bony trabeculae occur in abundance. The osseous trabeculae show lamination, with a rim of osteoblasts [13, 14]. The present case is in accordance with the subacute stage of FD.

Bone markers have been used to assess the activity of the disease and follow response to treatment. Total serum bone alkaline phosphatase and urine hydroxyproline are examples of useful markers. During the active phase of FD, levels of both of these markers are elevated in approximately 75% of patients [6].

FD should be differentiated from osteoclastoma, non-Hodgkin’s lymphoma, dentigerous cyst, dental cyst, and ossifying fibroma through a combination of clinical, radiographic and histological information.

Spontaneous resolution of FD does not occur. Asymptomatic lesions that do not progress and do not cause deformities or functional impairment can be observed [4, 6, 8]. The treatment is required in symptomatic cases and depends upon the extent of the lesion, rate of growth, aesthetic disturbance, and functional disruption. Surgery is recommended when the lesion becomes marked with pain, progressive deformity or interference with functions, to relieve symptoms or to correct aesthetic deformities but usually without curative intent, varying from simple shaving of the bone to more extensive surgery. In the present case, nasal obstruction was the only symptom related to the tumor and the lesion was confined to the inferior turbi-
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Considering the patient's age and the side effects such as atrophic rhinitis following complete resection, we partially removed the tumor leaving approximately proximal one tenth and surrounding mucosa via endoscopic endonasal approach. Although spontaneous sarcomatous transformation of FD is reported to occur, it is in less than 1% of the cases and most reported cases have occurred after radiation therapy [5, 12, 14]. In addition, because the inferior turbinate is easy to check for any change and excise endoscopically, we decided partial resection and regular follow up. The prognosis for FD is generally good, although outcomes are poorer in young patients and those with the polyostotic forms [6]. Recurrence is rare in adults, but the lesions can show surprising growth potential if they are surgically altered during their active growth phase.

Conflict of interest statement

All authors have no conflict of interest.

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