Cukurova Medical Journal

Olgu Sunumu / Case Report

A Rare Cause of Unilateral Chronic Nasal Obstruction: Fibrous Dysplasia of the Middle Turbinate

Tek Taraflı Kronik Burun Tıkanıklığının Nadir Bir Sebebi: Orta Konka Kaynaklı Fibröz Displazi

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Cukurova Medical Journal 2015;40 (Ek Sayı 1):106-111.

ABSTRACT

Fibrous dysplasia is a nonneoplastic fibro-osseous lesion characterized by fibroblastic proliferation and progressive replacement of normal bone with fibrotic tissue and disorganised bony trabeculae.Fibrous dysplasia infrequently occurs in the sinonasal tract. Location in the middle turbinate is extremely rare. It is usually asymptomatic but, in the advanced stage, nasal obstruction due to extension of disease and pain due to neural compression, or pathological fractures may occur. It is usually secondary to extension of disease from adjacent bones. In this case report we presented a 54 year old woman with unilateral chronic nasal obstruction

Key words: Nasal obstruction, middle turbinate, fibrous dysplasia

ÖZET

Fibröz displazi fibroplastik proliferasyon ve normal kemiğin progresif fibröz doku ile yerdeğiştirmesi ile karakterize nonneoplastik fibroosseöz bir lezyondur. Fibröz displazi sıklıkla sinonazal bölgede olmak ile birlikte orta konka kemiğinde oluşumu oldukça nadirdir. Genellikle asemptomatiktir ancak ileri olgularda hastalığın yayılımı ile burun tıkanıklığı, sinir basısından ötürü ağrı veya patolojik kırıklar olabilir. Genellikle bağlı kemiklerden hastalığın uzanıma seconder olarak gelişir. Bu olgu sunumunda tek taraflı burun tıkanıklığı olan 54 yaşında bayan hasta literature bilgileri gözden geçirilerek sunuldu.

Anahtar kelimeler: Burun tıkanıklığı, orta konka, fibröz displazi.

INTRODUCTION

Fibrous dysplasia (FD) is a nonneoplastic fibro-osseous lesion, characterised by fibroblastic proliferation and progressive replacement of normal bone with fibrotic tissue and disorganised bony trabeculae. It can be either monostotic or involve several bones. This disorder is usually unilateral with poorly defined edges between the normal bone and the pathologic tissue[1,2]. Although a variety of causative factors are described (trauma, neoplastic degeneration, genetic or embriologic modification), the causes of FD is still unknown[2-4]. According to reliable interpretations, this disease could be caused by congenital alterations of the mesenchyme due to traumas or neuroendocrine dysfunctions[2].

FD infrequently occurs in the sinonasal tract, where the middle turbinate is extremely rare bone

to occur. It is usually asymptomatic but, in the advaced stage, nasal obstruction due to extension of disease, pain due to neural compression, or pathological fractures may ocur. It is usually secondary to extension of disease from adjacent bones[2,5]. The signs and symptoms of FD of head and neck invariably relate to the location and extent of bony abnormalities. Facial asymetry is the most common sign of FD, while pain and ocular proptosis are the next most frequent symptoms, followed by neurological changes[5]. The paranasal sinuses are often collapsed, when the bony changes of the disease process cause obstruction and dysfunction of the ostia. Recently we experienced a case of the base of skull FD, including the frontal sinüs, sphenoid sinüs, ethmoid complex, maxiller sinus and middle turbinate. Nasal obstruction secondary to this rare disorder presents a diagnostic and therapeutic challange. The nasal obstruction in this case is due to expansion of fibrous lesions in adjacent bones as well as direct involvement of the middle turbinate with fibrous dysplasia. We report the case with a review of literatüre.

CASE

A 54 year old woman was referred to our clinic with unilateral chronic nasal obstruction, which was started more than 5 years ago. She didn't complain any other nasal symptoms (rhinorrhea, sneezing, itching, postnasal drip and hyposmia) and pain. Facial asymetry and ocular proptosis were not found. She was treated previously with topical corticosteroids and decongestants, but failed to reduce her symptoms. She denied other past or family medical history. Only she was defined a head trauma history 6 years ago. On the nasal endoscopic examination, the left middle turbinate was enlarged and bony hard in palpation. The nasal septum was not deviated. There was no polyp or discharge in the nasal cavity. No abnormal skin pigmentation could be found. No endocrine disorders or cutaneous hyperpigmentation or other bone lesions were observed. Laboratory blood tests including serum alkaline phosphatase and urine test were normal. Haematological tests excluded the McCune-Albright syndrome. Axial-coronal craniofacial computed tomography (CT) and magnetic resonance imaging (MRI) were performed. The CT scans showed characteristic 'ground-glass' appearance of the base of skull, including the frontal sinus, sphenoid sinus, ethmoid complex, maxiller sinus and middle turbinate with poorly defined borders and normal mucosal covering, which was compatible with FD (figüre1,2). No other craniofacial dysplastic lesions were found at CT. MRI revealed a well defined mass, with an intermediate to slightly hypointense signal on unenhanced T1-weighted images and homogenous, high signal intensity on T2-weighted images(figüre 3). The radiological features were considered to be consistent with fibrous dysplasia. Under the general anesthesia, a 4.0 mm diameter nasal endoscope was used to visualize a large bony mass extending from the skull base and involving the left side osteomeatal complex and middle turbinate. The bone mass was partially removed via endoscopic endonasal approach. Because we believe that the conservative bone shaving or partial curettage of the lesion to relieve pressure and symptoms. All of the endoscopic resection was done utilizing 0 and 30 sinus endoscopes. The microscopic findings were typical of FD. The patient has been free of symptoms for 1 year post surgery. At 1 year of follow-up, there was no evidence of recurrence on the nasal endoscopic examination.

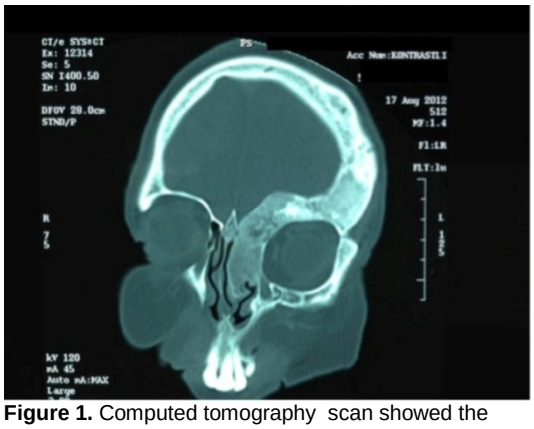

Figure 1. Computed tomography scan showed the affected bony structures and fibrous dysplasia of middle turbinate (Coronal slice view)

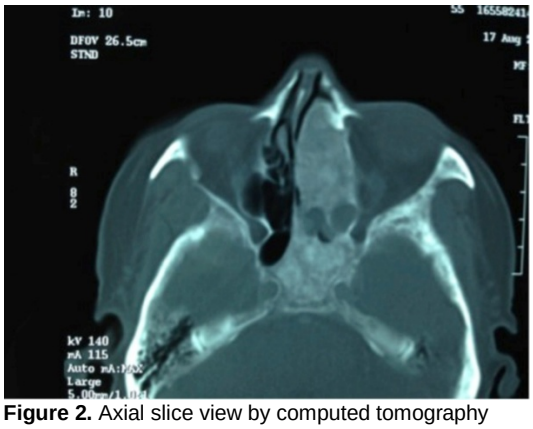

Figure 2. Axial slice view by computed tomography of the turbinate.

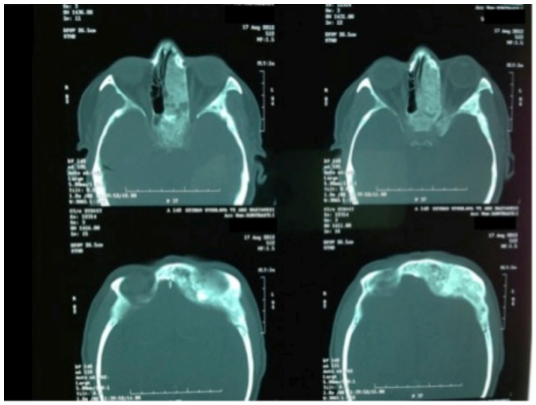

Figure 3. Computed tomography demonstrate the Obstruction of nasal airway by the enlarged middle turbinate

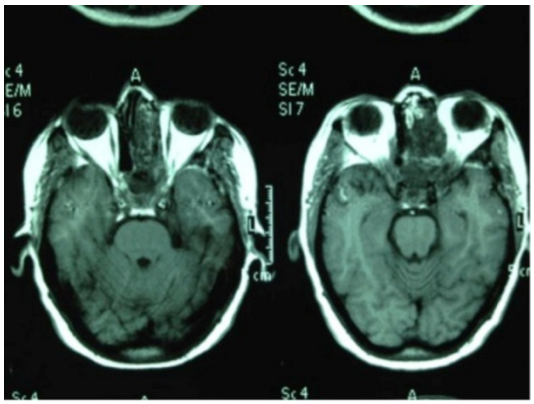

Figure 4. MRI scan showed the expansion of the middle turbinate.

DISCUSSION

FD is a sporadic benign skeletal disorder and accounts for approximately %7,5 of the benign bone neoplasm[1,6-8]. The fibro-osseous tumors can be further divided into monostotic FD in % 30 of cases and polystotic in %70[5,9]. Monostotic form with the involvement of the ribs, femur and tibia is most common. Polystotic form may be rarely complicated by cafe-au-lait spots and hyperfunctional endocrinopathies (McCune-Albright syndrome) or multiple soft-tissue myxomas (Mazabraud's syndrome) [1,8]. In monostotic form, males and females are thought to be affected with equal frequency, however polystotic form is more common in females[1,6,8,9]. Aproximately %50 to %100 of patients with polystotic disease and %10 to %30 with monostotic disease have craniofacial involvement. Craniofacial FD typically presents at around 10 years of age and then progress throughout adolescence[8]. Among FD of the head and neck, the maxilla and mandible are the most frequent sites to be affected. However, FD of the paranasal sinuses and middle turbinate is rare[5,10-12]. In the present case, the lesion was settled the base of skull including the frontal sinus, sphenoid sinus, ethmoid complex, maxiller sinus and middle turbinate with poorly defined borders and normal mucosal covering, which was compatible with FD. And there was no other craniofacial dysplastic lesions were found at CT.

The incidence of this disease is probably underestimated due to the asymptomathic bony proliferation that remains undetected until it gives rise to pain due to neurological compression or functional disorders in the affected organ[2]. Therefore, without related symptoms, lesions are typically found incidentally during radiological examinations performed for other causes. The most common clinical sign of craniofacial FD is bone pain or craniofacial deformity in the advanced stage. When the paranasal sinuses, orbits and foramina of the skull base are involved because of enlargement of the lesions, the patient may experience a variety symptoms, including headache, visual loss, proptosis, diplopia, hearing loss, anosmia, nasal obstruction, epistaxis, epiphora and recurrent sinusitis[1,9,10]. We know that, if the lesion involves the sinuses especially ostiomeatal complex, headache, nasal obstruction, or recurrent sinusitis symptoms are inevitable. In this case, the nasal obstruction is due to expansion of fibrous lesion in adjacent bones as well as direct involvement of the middle turbinate with fibrous dysplasia and blocking the ostiomeatal complex. On the middle turbinate, the bony degeneration gives rise to an increase in volume that can cause nasal obstruction or alterations in sinus drainage. After opening of the ostiomeatal complex by surgery, our patient's nasal obstruction symptom was relieved. The disease has a submucosal pattern of growth, and endoscopic examination shows turbinate enlargement that can be mistaken for a concha bullosa. In this step, if you do not performed radiological examinations, the diagnosis can be easily missed. On our patient's nasal endoscopic examination showed that, the left middle turbinate was enlarged and bony hard in palpation. We were able to diagnose with CT scans. CT is the radiological exam of choice; it highlights the bony nature of the lesion with the characteristic 'ground-glass' appearance. Due to the degree of mineralization of the tissue, findigs on CT scan can be from radiolucent to sclerotic[7]. The most common form is 'ground-glass' appearance with %56. Other forms are homogeneously dense pattern (%23) and cystic pattern (%21) [7,13]. MRI is useful especially in detecting and defining neuruvascular and ocular involvement[7,8].

FD may be associated with hormonal changes especially defects in the calcium-phosphorus metabolism. Blood tests such as calcaemia, alkaline phosphatase, urine hidroxyproline and parathormone assessment are mandatory to exclude from McCune Albright syndrome[2,14]. Bone markers have been used to assess the activity of the disease and follow response to treatment. During the active phase of FD, levels of both of these markers are elevated in aproximately %75 of patients[15]. In our case, laboratory blood tests including serum alkaline phosphatase, urine test and parathormone assessment were normal.

The differential diagnosis of FD includes the other fibrodysplastic disorders (Paget disease, hyperparatiroidism, osteogenesis imperfecta) and from ossifying fibroma[2]. To this end, histopathological studies are of fundamental importance since they reveal the presence of irregular bony trabeculae embedded in a cellular fibrous stroma without osteoblastic rimming[16]. To distinguish diseases with similar clinical and histological features, more especially for the ossifying fibroma that, unlike FD, usually occurs in adult age, is characterized by monostotic expansive lesions with a well-defined edge and, for this reason, shows a better response to surgical treatment. Complete surgical resection is recommended for the case of ossifying fibroma. This tumour tends to behave more aggressively than fibrous dysplasia and following incomplete resection recurrence rate is high[12]. Galvan et al, concluded that when the diagnosis is 'fibro-osseous lesion, not further specified', surgery is the treatment of choice whenever possible, when the diagnosis is FD, 'wait and see' is the treatment strategy choice[12].

Spontaneous resolution of FD does not occur. No medical treatment has been found to be effective in the management of FD. Solitary and small lesions may remain asymptomatic and requiring no treatment. Surgery is recommended to relieve symptoms including pain, nasal obstruction, progressive deformity or interference with functions[9]. There are also two surgical options: conservative bone shaving and radical excision[2, 17]. The extent of surgical resection of FD must be based on the location of the tumour, its proximity to important structures, and the severity of symptoms. Some authors advise partial cürettage of the lesion to relieve pressure and symptoms[2,17] .Also, on account of the poorly defined limits between normal and dysplastic bone, 'en-bloc' excision is often very difficult. In the present case, nasal obstruction was the only and important symptom related to the tumor and the lesion was confined to the ostiomeatal complex and skull base. To our knowledge, the first objective should be to reduce the patient's symptoms and because of the benign nature of the disease and the location of bony lesions including to the extend into the skull base, excision of the lesion in our patient was limited to the goal of opening the obstructed ostiomeatal complex. Considering the side effects such as CSF rhinorrhea or orbital complications following complete resection, and the priority to relieve the symptoms made us adopt the conservative surgery. So we have relieved the patient's chief complaint of nasal obstruction significantly by the partial surgery. Some authors have suggested that endoscopic approach used to operate on such a lesion[2,5,9,13]. We thought that the endoscopic approach is the preferable surgical method for our patient. Because endoscopic approach offers some significant advantages including no facial deformity and scars, excellent functional results and possible repetition in cases of recurrence. Radiotherapy should be avoided because of the possible malignancy transformation.

The prognosis of FD is generally good. Outcomes are poorer in young patients and with the polyostotic forms[15]. The recurrence rate is between %20-25 of these patients[5,15]. There is a % 1 risk of malignancy, especially in previously irradiated patients[1,8,18]. Because of the risk of recurrence and possible malignant degeneration, patients should be closely monitored long-term after surgery particularly if not radical..

In conclusion, fibrous dysplasia is a rare disease in sinonasal tract and middle turbinate. This case demonstrated that clinician must consider fibrous dysplasia, especially patient complaning of a unilateral chronic nasal obstruction with an enlarged middle turbinate for differantial diagnosis. An endoscopic transnasal approach is preferable surgical method for the opening of nasal airway.

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Geliş tarihi/Received on : 01.06.2015
Kabul tarihi/Accepted on: 08.07.2015