Fibrous dysplasia of middle turbinate associated with Widal syndrome: endoscopic treatment of a rare case

Trattamento endoscopico di un raro caso di displasia fibrosa del turbinato medio associata a triade di Widal

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Key words

Nasal disorders • Fibrous dysplasia • Middle turbinate • Widal syndrome • Endoscopic treatment

Parole chiave

Malattie del naso • Displasia fibrosa • Turbinato medio • Sindrome di Widal • Trattamento endoscopico

Summary

Fibrous dysplasia, a rare bony disease, is characterised by substitution of normal bone with immature tissue embedded in a fibrous stroma. It can be either monostotic or involve several bones. Fibrous dysplasia is usually asymptomatic but, in the advanced stage, pain due to neural compression or pathological fractures may occur. In the case of craniofacial involvement, ocular, masticatory, respiratory or auditory functional alterations are possible. A case of fibrous dysplasia, limited to the middle turbinate and associated with Widal triad (sinus-nasal polyposis, asthma, acetyl salicylic acid intolerance), is described. Craniofacial computed tomography revealed enlargement of left middle turbinate with characteristic "ground-glass" appearance. The patient underwent anterior bilateral functional endoscopic sinus surgery with near-total resection of left middle turbinate. Histopathological examination confirmed the diagnosis of fibrous dysplasia. After 2 years the patient is still asymptomatic. Videorhinoscopy shows good sinus-nasal patency without disease recurrence. Even though exceptional, localization of fibrous dysplasia at middle turbinate has been described, therefore, it must be considered in the differential diagnosis of the craniofacial ossifying disorder. For localized and symptomatic lesions, endoscopic surgery is an effective option. Scrupulous life-long follow-up is necessary due to the high percentage of recurrence and possible malignant degeneration.

Riassunto

La displasia fibrosa è una rara patologia ossea, benigna ed idiopatica, caratterizzata dalla sostituzione dell'osso sano con tessuto immaturo immerso in uno stroma fibroso. Può presentarsi in forma monostotica o interessare più distretti. Generalmente decorre asintomatica; tuttavia, negli stadi avanzati, possono comparire dolori da compressione nervosa o fratture patologiche; in caso di interessamento cranio-facciale possono aversi alterazioni funzionali masticatorie, oculari, respiratorie ed uditive in relazione alle ossa colpite. Riportiamo un raro caso di displasia fibrosa limitata al turbinato medio ed associata a triade di Widal (rinosinusite polipoide, asma bronchiale allergica, intolleranza all'acido acetil-salicilico). Il paziente è stato sottoposto a videorinoscopia ed a TC cranio-facciale che ha evidenziato un aumento di volume del turbinato medio di sinistra con degenerazione sclerotica caratterizzata da zone calcifiche alternate ad aree di rarefazione. Vista la localizzazione ben definita della lesione e la presenza di sintomatologia nasale, il paziente è stato sottoposto a chirurgia funzionale endoscopica rinosinusale (FESS) del comparto anteriore bilateralmente ed a resezione sub-totale del turbinato medio di sinistra. L'esame istologico del pezzo operatorio ha confermato la diagnosi di displasia fibrosa del turbinato medio. A 2 anni dall'intervento il paziente risulta asintomatico senza ripresa di malattia. Per quanto eccezionale la localizzazione della displasia fibrosa al turbinato medio deve essere considerata nella diagnosi differenziale di patologie ossificanti di questo distretto. In caso di lesioni localizzate e sintomatiche il trattamento chirurgico endoscopico costituisce una valida opzione terapeutica. L'elevata percentuale di recidiva e la possibile degenerazione maligna rendono necessario un attento follow-up.

Introduction

The term fibrous dysplasia (FD) refers to a rare bony disease, benign and idiopathic, characterised by fibroblastic proliferation and progressive substitution of the normal bone with fibrotic tissue and disorganised bony trabeculae. This disorder is usually unilateral with poorly defined edges between the normal bone and the pathologic tissue [1]. Although several aetiological theories have been advanced (trauma, neoplastic degeneration, genetic or embryologic modification …), the causes of FD are still unknown [2 3]. According to reliable interpretations, this disease could be caused by congenital alterations of the mesenchyme due to traumas or neuroendocrine dysfunctions [1]. Recently, attention has been focused on a defect in the adenylate cyclase signal transduction system found in the pathological tissues [4].

In 70% of cases, FD involves only one bone (monostotic form), especially the ribs or proximal portion of the femurs and tibias. In only 10% of cases a craniofacial bone is affected, in particular the maxilla or the mandible. The polyostotic form which accounts for approximately 30% of cases is usually limited to one side of the body and, in approximately half the patients, craniofacial lesions are present. Polyostotic fibrous dysplasia can be associated with cutaneous hyperpigmentation and endocrine disorders (hyperparathyroidism, precocious puberty …) in the McCune-Albright syndrome which is probably due to somatic alterations during the early phases of embryogenesis [5]. FD, in all cases, is characterized by a slow and asymptomatic increase in volume of the affected bones and is usually an incidental radiological finding. In the advanced stages, cosmetic changes and pain due to neurological compression or pathological fractures may appear; if craniofacial bones are affected, ocular, uditive, respiratory and masticatory dysfunctions may occur in relation to the bones involved.

FD is more frequent in females, in particular between the ages of 10 and 30 years. Progression of the disease slows down after puberty reaching a complete halt after the third decade; however, cases presenting evolution of dysplasia, independently of age, as well as the possibility of malignant degeneration, have been described [1,2].

The present report focuses on a rare case of FD limited to the middle turbinate and associated with Widal triad (sinus-nasal polyposis, bronchial asthma, acetyl salicylic acid (ASA) intolerance), successfully treated with the endoscopic approach. Clinical features, diagnostic problems and therapeutic possibilities are also discussed.

Case report

A 28-year-old male was referred to us complaining of progressive hyposmia and respiratory nasal obstruction, prevalently on the left side. He reported a history of bronchial asthma, assessed with the metacholine test, ASA intolerance and allergy to grass pollen and dermatophagoides.

Videorhinoscopy revealed right ethmoid polyposis and left middle turbinate hypertrophy. Axial-coronal craniofacial computed tomography (CT), used to complete the clinical assessment, revealed ethmoidal inflammatory tissue with bilateral obstruction of the ostium-meatal complex and inspissation of the maxillary sinus mucosa. The left middle turbinate appeared enlarged with the characteristic "ground-glass" appearance (Fig. 1).

The patient underwent anterior bilateral functional edoscopic sinus surgery (FESS) with near-total resection of the left middle turbinate. Histological examination of the surgical specimen revealed the presence of disorganized trabeculae of immature bony tissue embedded in a connective fibrous stroma without cellular atypia: features consistent with fibrous dysplasia (Fig. 2). The diagnostic assessment was, therefore, completed with dermatological evalua-

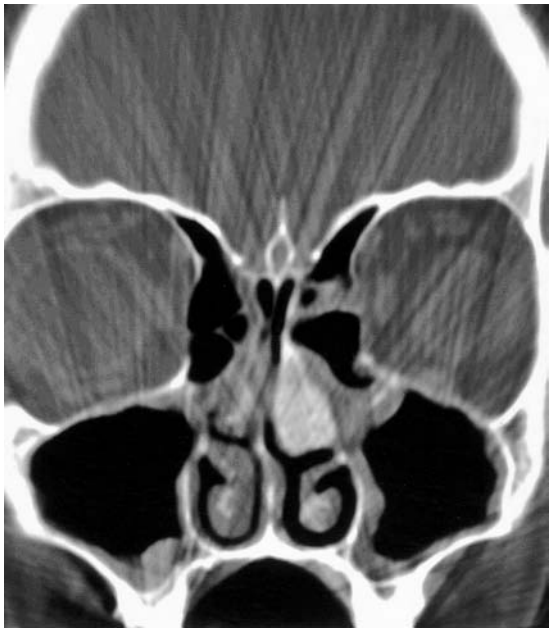

Fig. 1. Coronal preoperative CT: enlargement of left middle turbinate with characteristic "ground-glass" appearance.

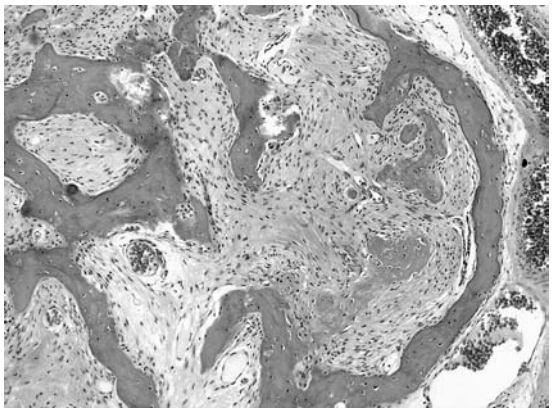

Fig. 2. Histological examination: disorganized trabeculae of immature bony tissue embedded in connective fibrous stroma without cellular atypia.

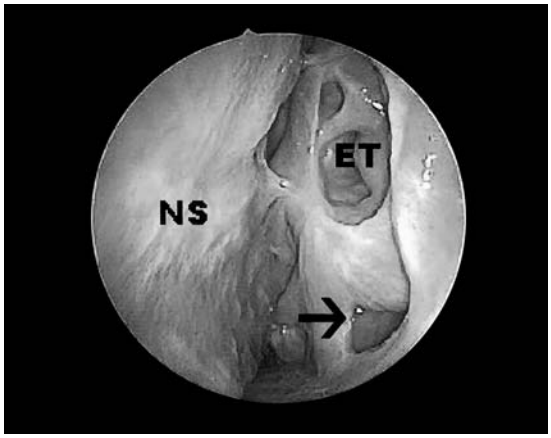

Fig. 3. Postoperative videorhinoscopy: good sinus-nasal patency without disease recurrence.
NS = nasal septum; ET = ethmoid; arrow = maxillary ostium.

tions and haematological tests that excluded the McCune-Albright syndrome. No other craniofacial dysplastic lesions were found at CT. Since the patient refused to undergo other radiological examinations, a polyostotic form could not be excluded.
Complete olfactory recovery and resolution of the respiratory nasal obstruction were reported, by the patient, 15 days post-operatively. After 2 years, the patient is still asymptomatic, no further episodes of asthma have occurred. Videorhinoscopy has confirmed good sinus-nasal patency without disease recurrence (Fig. 3).

Discussion

Localization of FD only at the middle turbinate is an extremely rare event; only two other cases having been reported in the English literature [2 6] neither of which underwent surgical treatment. Nevertheless, the incidence of this disease is probably underestimated due to the asymptomatic bony proliferation that remains undetected until it gives rise to pathological fractures, pain due to neurological compression or functional disorders in the affected organ. Therefore, without related symptoms, diagnosis is usually incidental during radiological examinations performed for other causes.
On the middle turbinate, the bony degeneration gives rise to an increase in volume that can cause nasal obstruction or alterations in sinus drainage. Videorhinoscopy shows turbinate enlargement that can be mistaken for a concha bullosa; this examination also allows evaluation of other, associated or correlated, sinus-nasal diseases. Axial-coronal CT, in thin sections, is the radiological exam of choice; it highlights the bony nature of the lesion with the charactestic "ground-glass" appearance. The radiographic findings may change in relation to the evolutive stage of the disease, therefore, three classic radiological patterns have been described:
– Lytic or cysti-like form characterized by roundish rarefactive areas with a sclerotic border;
– Sclerotic form with homogeneous bone expansion;
– Pagetoid form with dishomogeneous areas of radiodensity and radiolucenty [1 4].

With CT, it is also possible:
– to define the boundaries and connections of the lesion;
– to exclude a polyostotic craniofacial form;
– to identify other possible associated disorders;
– to monitor evolution of the disease.

FD has to be differentiated from other fibrodysplastic disorders (Paget disease, hyperparathyroidism, osteogenesis imperfecta …) and from ossifying lesions (osteoma, osteosarcoma, chondroma, ossifying fibroma …). 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 [7]. However, to distinguish diseases with such similar clinical and histological features, close cooperation between the clinician, the pathologist and the radiologist is essential [1], 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.
FD may be associated with hormonal changes as well as defects in the calcium-phosphorus metabolism; therefore, blood tests such as calcaemia, alkaline phosphatase and parathormone assessment [6 8] are mandatory to exclude a syndromic form, in particular those of McCune-Albright. On the contrary, the association of FD with polypoid sinusitis, allergic bronchial asthma and ASA intolerance (Widal triad), that we consider incidental, has not been described, so far, in the literature.
No medical treatment has been found to be effective in the management of FD; therefore, in those cases presenting localized lesions with functional disorders or cosmetic alterations, surgical treatment is indicated. On account of the poorly defined limits between normal and dysplastic bone, "en-bloc" excision is often very difficult and associated with recurrence in approximately 20-25% of these patients. Partial resections or bony reshapes are more frequently performed also in consideration of the fact that the disease tends to exhaust itself after puberty.
In the case described here, the well-defined localization of the lesion at middle turbinate level and the

presence of nasal symptoms, also due to associated polyposis, led us to proceed with endoscopic resection of the lesion associated with functional endoscopic sinus surgery.
Surgical treatment, particularly if not radical, requires long-term follow-up in order to monitor the risk of recurrence and to evaluate possible malignant degeneration. Indeed, FD is associated with sarcomatous evolution in 0.5-1% of cases, in particular in cases of craniofacial lesions [9 10]. The risk of malignant degeneration increases considerably after radiation-therapy that must be avoided in all cases.

Conclusions

Even though exceptional, localization of FD at the middle turbinate has been described, and, therefore, must be borne in mind in the differential diagnosis of the craniofacial ossifying disorder [2 4]. Likewise, possible syndromic associations should be sought.
Videorhinoscopy and craniofacial CT are fundamental examinations in the correct assessment of this disease, diagnosis of which, however, depends on a scrupulous study of the histopathological features.
In the case of localized and symptomatic craniofacial lesions, endoscopic surgery is an effective option [7] that must be considered, particularly if sinus-nasal disorders are associated.
Scrupulous life-long follow-up is mandatory on account of the high percentage of recurrence and possible malignant degeneration.

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