Otolaryngology–Head and Neck Surgery (2007) 136, S59-S61

CASE REPORT

Endoscopic treatment of fibrous dysplasia confined to the frontal sinus

Simon D. Charlett, BSc, MBChB, MRCS, Stuart G. Mackay, BSc, MBBS, and **Ray Sacks, MBBCh, MMED (ORL), FRACS,** Sydney, NSW, Australia

Fibrous dysplasia is a benign idiopathic skeletal disorder, characterized by replacement and expansion of medullary bone by disorganized fibro-osseous tissue.[1] The disease primarily affects long bones but may also develop in the craniofacial region. We report a rare case of a symptomatic patient with isolated frontal sinus involvement who was successfully treated using a modified endoscopic Lothrop procedure.

A 39-year-old sheep-shearer was referred by his local ENT surgeon with a six-month history of a constant and progressively worsening frontal headache. The pain was localized to the mid-forehead on the left side and had become so severe he was unable to work. Other symptoms were left-sided nasal obstruction and snoring, neither of which had improved with topical nasal steroids. His past medical history included a nasal injury from playing sports, and Crohn's disease controlled with Sulphasalazine 1 g twice daily.

Examination revealed a mild nasal septal deformity to the left side and polypoidal mucosa within the middle meatus. Computed tomography (CT) of the paranasal sinuses demonstrated a 1.5-cm mid-line lesion in the frontal sinus of mixed sclerotic and soft tissue density, with evidence of cortical thinning of the anterior table (Fig 1). These radiographic features were suggestive of a fibro-osseus lesion such as fibrous dysplasia or an ossifying fibroma; however, alternative diagnoses considered included chronic osteitis and eosinophilic granuloma.

The patient underwent an image-guided modified endoscopic Lothrop procedure. Intraoperative findings were a bony mass of low vascularity in the left frontal sinus consisting of a thick hard outer rim and a soft central fibrotic core. The specimen was sent for histopathological analysis. The patient was given two doses of intravenous ceftriaxone 1g twice a day postoperatively. The following day he was discharged home with cephalexin 500 mg four times a day for one week, and advised to use regular saline nasal douching for two weeks.

Histopathological assessment confirmed a fibro-osseous lesion, showing foci of irregular and rounded metaplastic woven bone embedded in predominantly spindle-cell fibrous tissue (Fig 2). A diagnosis of fibrous dysplasia was made following radiological correlation at a multidisciplinary bone tumor meeting. On review one week later, his frontal headache had improved significantly. There were no postoperative complications, and he returned to work shortly thereafter.

DISCUSSION

Fibrous dysplasia (FD) is classified into three types. Monostotic is the most common form, accounting for about 70% of cases.[2] It involves one or two contiguous bones and is typically found in the ribs, proximal femur, tibia, and calvarium.[3] Polyostotic fibrous dysplasia (20%-30%) occurs in multiple sites and has a predominantly unilateral distribution, although bilateral disease has been described.[4] McCune-Albright syndrome is a rare congenital disorder caused by sporadic gene mutations, and is a variant of polyostotic FD. It usually affects females and is associated with cutaneous hyperpigmentation, precocious puberty, and endocrine abnormalities.[3] Fibrous dysplasia is generally chronic and slowly progressive; how-

From Concord Repatriation General Hospital.

Reprint requests: Simon D. Charlett, BSc, MBChB, MRCS, Leeds General Infirmary, Great George Street, Leeds, West Yorkshire, LS1 3EX, United Kingdom.

E-mail address: sdcharlett@doctors.org.uk.

doi:10.1016/j.otohns.2006.10.025

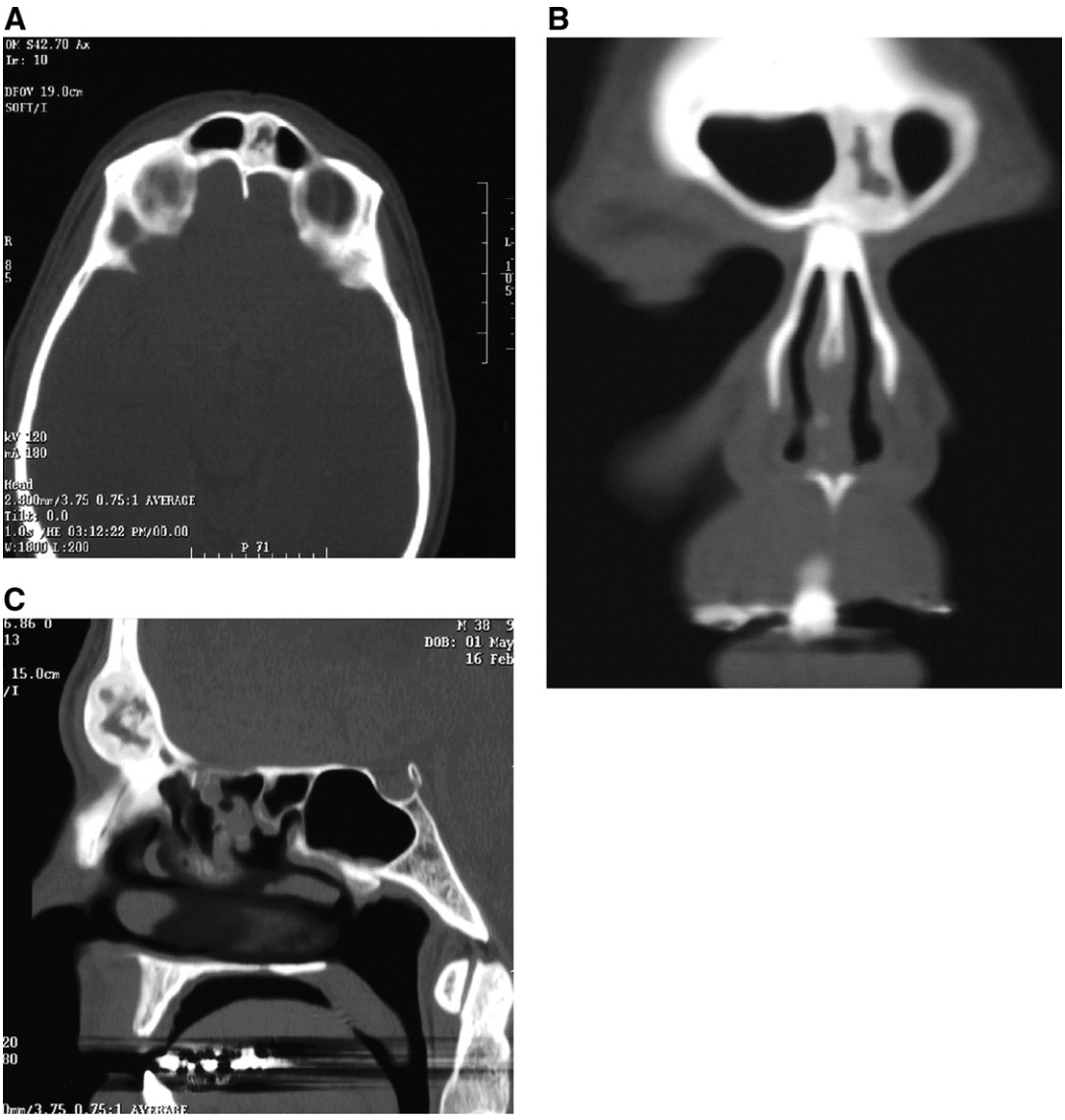

Figure 1 **(A)** Axial; **(B)** coronal and **(C)** parasaggital CT views of the paranasal sinuses demonstrating a fibro-osseous lesion in the left frontal sinus.

ever, malignant transformation has been observed in polyostotic disease, usually to osteosarcoma (0.4%).[5]

Craniofacial FD occurs in 10% to 25% of monostotic and over 50% of polyostotic cases.[3,6] Paranasal sinus involvement is rare, and there have been only two cases of FD confined to the frontal sinus reported in the past 30 years.[7,8] The monostotic form is often asymptomatic and discovered as an incidental finding on radiological imaging for other reasons.[2] Clinical manifestations are caused by the effects of an expanding mass, which may produce bony deformity, and morbidity resulting from nasal or sinus obstruction. Chronic sinusitis and mucocele formation have been described as sequelae of FD occluding the normal drainage pathway of the frontal and sphenoid sinuses.[4,7,9] When these disease processes are complicated by orbital and intracranial extension, patients may present with visual disturbance and proptosis, and neurological symptoms, including headaches, facial pain, cranial nerve palsies, convulsions, seizures, and meningitis.[3,6-8,10]

High-resolution CT is the investigation of choice for radiological examination of the paranasal sinuses.[1] Features of FD include expansion of the diploic space by fibrocellular connective tissue of mixed tissue density and a thickened sclerotic cortical margin.[4] Pathologically, the lesions are often yellow or greyish-white and have a gritty consistency.[11] Microscopic analysis characteristically demonstrates the replacement of normal cancellous bone with fibrous tissue. Haphazardly arranged spicules of woven bone laid down within the stroma produce the classical ground-glass appearance on CT imaging.[3] In most instances, a diagnosis of fibrous dysplasia cannot be made from clinical, radiological, or histological features alone, but requires a combination of all three.[2] Correlation of these modalities is best performed in a multidisciplinary team setting.

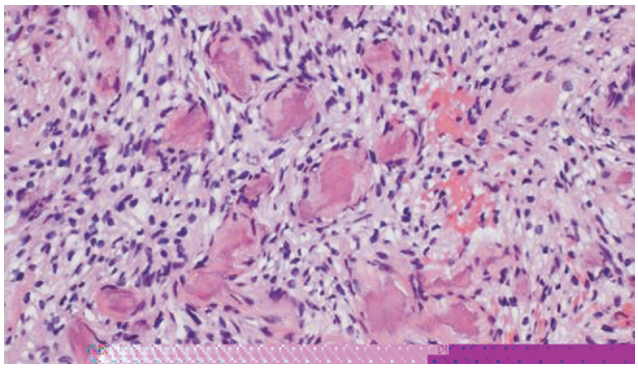

Figure 2 Histopathological specimen from the left frontal sinus at high power illustrating typical features of fibrous dysplasia. Islands of metaplastic woven bone embedded in spindle-cell fibrous tissue.

Patients with asymptomatic FD do not require surgical intervention. In this case the patient's symptoms were considered to be directly attributable to the frontal sinus pathology. Diseases of the frontal sinus have traditionally been treated using a bicoronal osteoplastic approach with fat obliteration. A subperiosteal scalp flap is created and the anterior table of the frontal sinus is raised, offering excellent access to the frontal sinus at the cost of significant postoperative morbidity. The modified endoscopic Lothrop procedure (MELP) is an intranasal technique that has become a recognized treatment for complicated chronic frontal sinusitis and tumors of the frontal sinus.[2,12,13] It is based on a combined intranasal and external approach originally described by Lothrop in 1916, and involves creating a common communication between the frontal sinuses and nasal cavity to improve ventilation and drainage.[14] This endoscopic technique has several advantages over external approach surgery, including a shorter operative procedure and inpatient stay, and lower complication rate.[15]

The MELP is a technically demanding procedure due to the narrow surgical field and difficult operating angle. Image-guidance systems allow operative progress to be visualized on CT images in three planes and help in the identification of important anatomical landmarks. They have therefore become a valuable tool for navigating through the complex frontal sinus outflow tract, particularly in cases of advanced disease and revision surgery.

CONCLUSION

Fibrous dysplasia of the frontal sinus is a rare cause of frontal headaches, but the diagnosis should be considered in patients who have characteristic radiographic features on CT imaging. Symptomatic relief can be successfully achieved using an image-guided modified endoscopic Lothrop procedure.

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