CLINICAL CASE OF MONOSTOTIC FIBROUS DYSPLASIA IN A CHILD

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Annotation

Fibrous dysplasia is a rare tumor-like condition characterized by the replacement of normal bone with fibrous tissue, with an etiology of uncertain origin. Diagnosis relies on clinical and radiological data, with biopsies used in doubtful cases. The aim of our study is to demonstrate our experience in working with a rare pathology such as fibrous dysplasia of the pterygoid process. We report a clinical case of isolated fibrous dysplasia affecting the pterygoid process of the left sphenoid bone, treated in the Pediatric Head and Neck Surgery Department of the University Medical Center, Astana, Kazakhstan. The patient, a 15-year-old girl, presented with a diagnosis of fibrous osteodysplasia of the left pterygoid process of the sphenoid bone. As a result of surgical treatment, we did not observe an increase in the tumor during a 3-year follow-up. Diagnosis of some tumors, such as fibrous dysplasia, is difficult and requires modern diagnostic methods. The presence of fibrous dysplasia in the sphenoid bone is a caustic pathology, which makes this clinical case unique. In this case, we used transnasal endoscopic approaches, which demonstrated the effectiveness of the treatment.

Introduction

The onset of fibrous dysplasia (FD) is usually subtle, with initial symptoms such as headache,[1,2] or ocular disturbances. Early diagnostic efforts typically involve neurologists, ophthalmologists, and pediatricians.[3] Diagnosing craniofacial bone disorders in children is challenging due to the variability of clinical presentations, the subtlety of symptoms, and the extended latent periods.[4] This complexity often leads to delayed diagnosis, which can negatively affect the child's development or lead to disability.[5]

According to international data, fibrous dysplasia affects the craniofacial region in 10-29% of children.[1,2] However, there are no specific epidemiologic data for Kazakhstan or other CIS countries. Among all bone pathologies, fibrous dysplasia accounts for approximately 2.5% [6] and up to 7.5% in cases of bone-related cancers.[7] The disease affects boys and girls equally, with the age of diagnosis typically ranging from 5 to 30 years.[8] Most cases manifest in childhood, with disease progression often coinciding with skeletal growth. Bone lesions usually develop soon after puberty,[9] with a sarcomatous transformation risk ranging from 0.5% to 4%.[10]

Monostotic forms are most commonly found in the ribs, craniofacial bones (mandible and maxilla), and femur.[11] Polyostotic forms often involve the lower limbs, pelvis, and may include skin and endocrine pathologies as part of McCune-Albright syndrome.[12]

Cranial fibrous dysplasia may be asymptomatic or present with symptoms such as exophthalmos, facial pain, dizziness, facial asymmetry, cranial nerve impairment, sinusitis, or headaches, depending on the lesion's location.[13,14,15] Radiologically, affected areas can appear dense or lucent compared to surrounding bone, sometimes resembling "ground glass." Lesions may have sclerotic borders or diffuse margins, particularly in cranial bones. Fractures may present with pronounced periosteal reactions.[16,17,18]

Clinical presentation

A 15-year-old girl presented with complaints of persistent headaches. Her

symptoms reportedly began in 2019 with episodes of dizziness, primarily at night. Initial physical examination revealed no significant abnormalities except for signs of medicamentous rhinitis. An otorhinolaryngologist identified nasal congestion linked to frequent use of decongestant nasal sprays and referred her for radiographic imaging of the paranasal sinuses, which revealed an abnormality. Subsequent computed tomography (CT) identified a lesion in the sphenoid bone.

Diagnostic Imaging

CT of the paranasal sinuses (November 9, 2021) (Figure 1):

✓ Axial and 3D reconstructed scans revealed abnormal "ground glass"-like changes in the left sphenoid sinus with intact cortical layers, measuring approximately 3.7 × 1.8 × 2.0 cm.

✓ A polypoid formation in the left maxillary sinus was noted, measuring 1.2 × 1.0 cm with clear borders and a density of 51 HU.

✓ Additional MRI (July 25, 2019) confirmed the lesion in the left sphenoid bone.

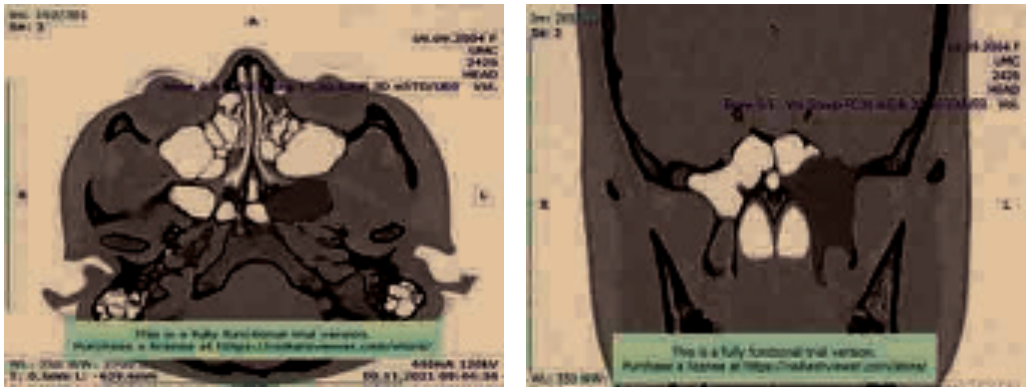

Figure 1.
Patient's CT scans before surgery (a-axial, b-coronary) The CT scans show a tumor of the pterygoid bone (left process) of uniform consistency (80 HU) with clear edges and no signs of growth into the surrounding tissue or into the nasal cavity or nasopharynx.

Treatment

The patient underwent endoscopic surgical removal of the lesion on December 14, 2021, performed under general anesthesia during a master class in the Pediatric Head and Neck Surgery Department of "UMC." Specialists from Dmitry Rogachev National Medical Research Center, Moscow, participated in the procedure. The lesion was removed using a surgical burr without complications. Histopathological analysis confirmed the diagnosis of fibrous dysplasia. Postoperative recovery was uneventful, and the patient was discharged on the 10th day in satisfactory condition. Subsequent sinus tomography 10 months after surgery revealed partial fibrous dysplasia of the pterygoid process (Figure 2).

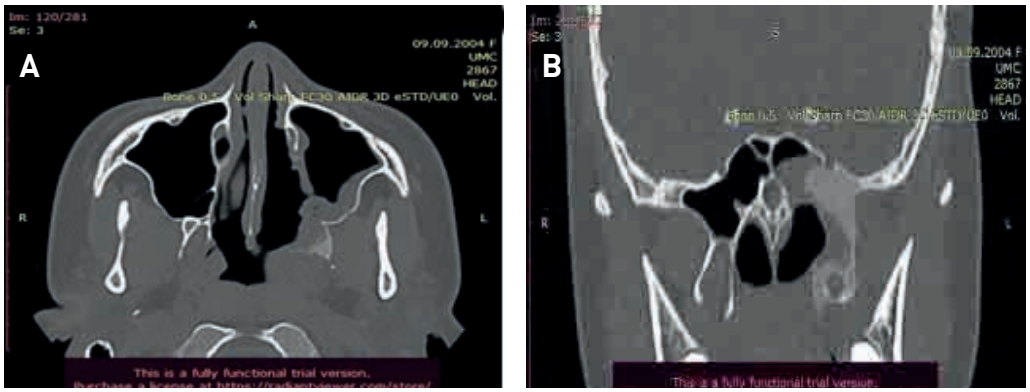

Figure 2.
After surgery (a-axial, b-coronary).
Post-surgical defect of the posterior nasal wall (access site) and partial tissue with fibrous dysplasia as a ground glass opacity

Follow-Up

Three years postoperatively, CT imaging (2024) showed residual fibrous dysplasia in the left sphenoid bone without progression (Figure 3). A postoperative defect in the medial wall of the left maxillary sinus was observed.

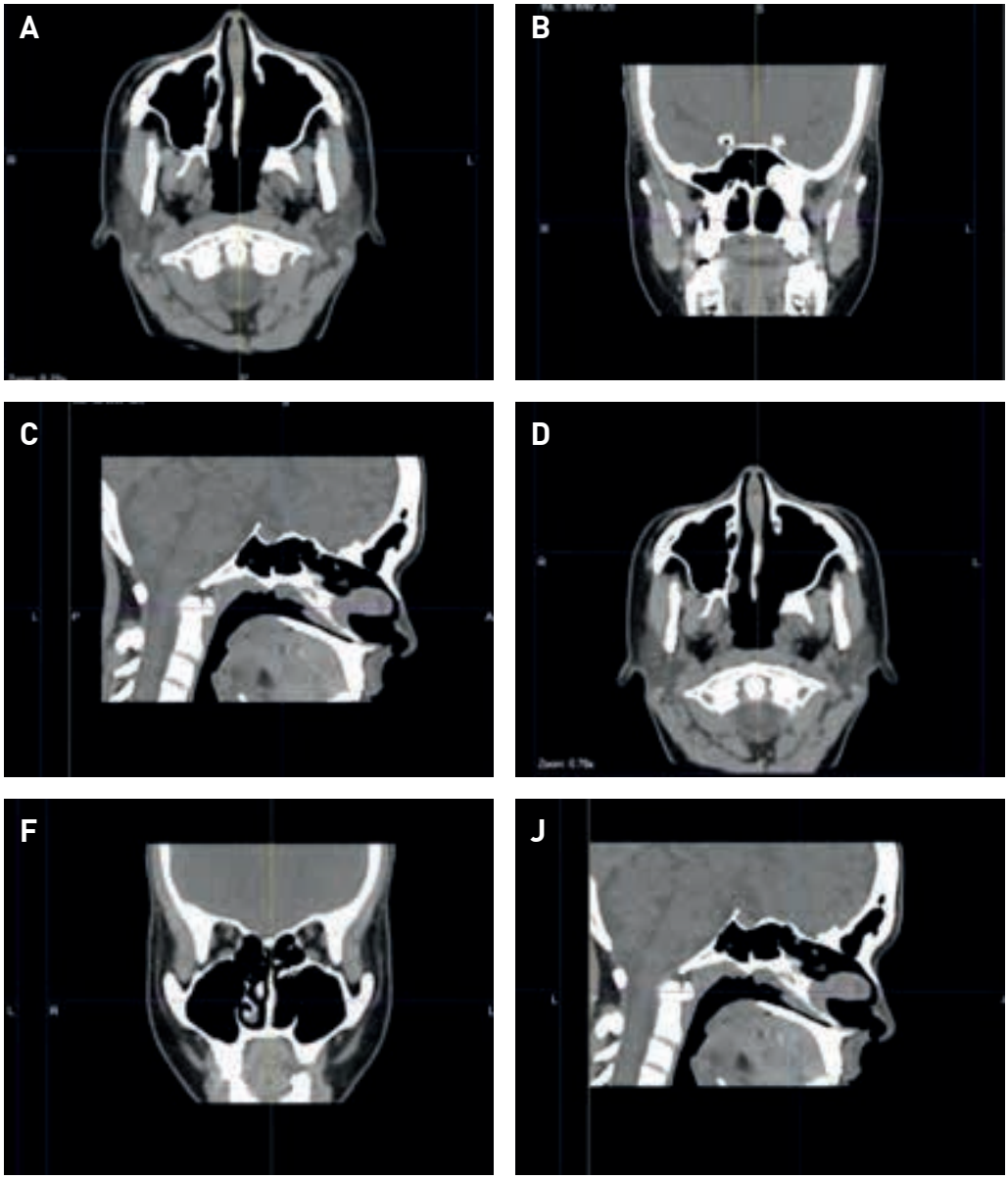

Figure 3.
Follow up after surgery (a/d-axial, b/f-coronary, c/j - sagittal).
A picture taken three years after surgery shows no recurrence of FD.
The series of images shows an overview of the skull bones in three positions in the early (a, b, c) and late after 3 years (d, f, j) postoperative periods.
Axial images show a large defect in the posterior nasal wall, the upper coronal image shows plus tissue with clear borders measuring 1.5 * 1.0 cm.
Both sagittal images do not show any destructive changes in the bone.

Discussion

This work is devoted to such a rare pathology as fibrous dysplasia, which is classified as a benign formation.[19] This formation can affect several bones at the same time, which can subsequently lead to its change, progress in the form of bone atrophy and, accordingly, to deterioration.[20] It is noted that fibrous bone lesions in childhood occur more in the craniofacial bones, especially in the upper jaw, lower jaw and sphenoid bone.[21]

Our study is devoted to a rarer form of FD of the cranial bones, which, according to the literature, occurs in less than 3%,[22] especially the monostotic form of sphenoid bone lesion. A clinical case of a 15-year-old female patient with isolated fibrous dysplasia of the left pterygoid process of the sphenoid bone is presented. With scanty symptoms (headaches), the tumor was detected by routine examination by neurologists using magnetic resonance imaging, followed by contrast computed tomography of the brain.

The pathognomonic sign of FD of bone tissue was "ground-glass" type changes on the radiograph, which corresponds to the literature data.[23,24] This image was crucial for the correct diagnosis of FD in a child, as it is necessary to differentiate from similar conditions such as ossifying fibromas or other benign bone tumors. Treatment of the patient is based on transnasal endoscopic partial resection of the affected tissue. This therapeutic tactic allows us to re-

duce the risks of invasive interventions in the head in the postoperative period, which has been confirmed by other authors.[3]

The patient's recovery was successful, and no evidence of progression was found during subsequent observations. Nevertheless, it is important to note that bone tissue FD, especially is monoaxial, are subject to gradual growth and malignancy.[1]

It is important to follow such patients for a long time, with repeated CT scans of the brain every year. We periodically examined her during this period and did not notice any signs of progression.

This clinical case is unique in that the lesion of the sphenoid bone is casuistic and we did not find a similar description of the case in the literature. FD is most commonly observed in the long bones (forearm, femur), sometimes in the pelvic bones.14 There are cases in the literature of craniofacial lesions of fibrous dysplasia in children.[14] There are cases of craniofacial lesions of fibrous dysplasia in the literature among children,[16] where endoscopic approaches have also been used in treatment. The peculiarity of our case is that a gentle minimally invasive approach was used in the treatment, which reduced the cosmetic and psychological damage to the child, but was no less effective in therapy.

Limitations. Despite the favorable outcome, the postoperative period was only 3 years. This category of patients requires long-term follow-up (10-20 years), and sometimes lifelong follow-up. In addition, it is necessary to perform a genetic study of patients admitted to FD to exclude mutation of GNAS genes, which are often associated with FD, which was not available in our case.

What's known? The diagnosis of FD of the sphenoid bone is usually incidental during a routine brain examination by MRI or CT, since the clinical symptoms of this pathology can rarely manifest themselves as headache or visual disturbances. The characteristic radiographic feature of FD in children is the presence of bone sparseness in the form of "ground glass", which helps to distinguish it from other pathologies.

What's New? This case report presents a casuistic case of isolated monostotic fibrous dysplasia of the sphenoid bone in a child, which is documented for the first time. We successfully applied minimally invasive treatment methods, which allowed us to minimize cosmetic and psychological consequences. Subsequent annual imaging showed no progression of the disease, underscoring the importance of long-term follow-up.

Conclusion

Isolated lesion of the sphenoid bone with fibrous dysplasia is a rare pathology. Diagnosis requires the use of computed tomography with contrast, which reveals the complex structure of the disease. Endoscopic transnasal resection of the tumor is currently a gentle and effective tactic in the treatment of these patients. However, longer follow-up and annual imaging (CT of the brain) are required to monitor disease progression.

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Authors' Contribution. A.D.: conceptualization, methodology, and writing of the manuscript, Surgical intervention and postoperative care. B.A.: clinical management of the patient and data collection. S.N.: Radiological analysis and imaging data interpretation, conceptualization, methodology. B.M.: conceptualization, methodology, clinical management of the patient and data collection.

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