Fibrous dysplasia of sphenoid bone – diagnostic difficulties

Dysplazja włóknista kości klinowej – trudności diagnostyczne

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SUMMARY:
Fibrous dysplasia (FD) of the bones is a benign disorder of unknown etiology in which normal medullary bone is replaced by fibrotic tissue. Lesion is rarely located in the sphenoid bone. Radiological features are characteristic but nonpathognomonic, so definitive diagnosis requires histopathological confirmation by biopsy. The authors present a case of a 58-year-old woman suffering from FD of sphenoid bone. The patient complained of a headache for several years, with no other associated symptoms. Magnetic resonance imaging (MRI) and computed tomography (CT) have not confirmed benign nature of the disease. However, biopsy specimen obtained by endoscopic surgery with neuronavigation showed the presence of fibrous dysplasia of the sphenoid bone.

KEY WORDS: fibrous dysplasia, neuronavigation, functional endoscopic nasal surgery, sphenoid sinus

INTRODUCTION
Fibrous dysplasia (FD) of bones is a benign disorder of yet unknown etiology and pathological mechanisms [1,2]. It is believed to be caused by an activating mutation within GNAS gene in chromosome 20 [3]. The natural history of the disorder consists in slow, progressive replacement of normal bone tissue by irregular, proliferative, isomorphic fibrous tissue separated by poorly developed irregular trabeculae [4]. Three subtypes of the disorder have been identified: monoosteotic (70% of cases), polyosteotic (30% of cases) and McCune-Albright syndrome [5]. Monoosteotic disease affects a single bone and is...
usually located within the costa and facial cranium bones. It is a benign disease occurring within the second or third decade of life. The polyostotic subtype affects multiple bones and is observed mainly in children. In 3-5% of cases, polyostotic FD is accompanied by endocrine disorders. The most severe form of the disease is McCune-Albright syndrome in which fibrous dysplasia of bones is accompanied by café-au-lait skin pigmentation and precocious puberty [4,6].

Fibrous dysplasia was first described as a separate nosocomial entity by Lichenstein in 1938 [2,7]. As much as 80% of cases is observed in Caucasian individuals [4]. Most commonly, lesions stabilize during the puberty and are observed mainly in female subjects. Lesions within the facial cranium bones include mainly the mandible and maxilla as well as paranasal sinuses, particularly the maxillary and, less commonly, ethmoid sinuses [8]. Sphenoid location of fibrous dysplasia is extremely rare [4]. Facial cranial locations are estimated to comprise about 25% of cases of the monoosteotic disorders and ca. 40-60% of cases of polyosteotic disorder [4]. The involvement of the sphenoid sinus causes headaches, disturbed vision, double vision, nasal congestion and ptosis [5,9]. The key role in the diagnostic process is played by magnetic resonance and computed tomography imaging. A characteristic feature observed in CT scans is the ground-glass matrix image [10]. Differential diagnosis should include various nosocomial entities [Table 1].

In addition, FD is often accompanied by other lesions such as myxomas [11]. Due to the benign nature of the lesion, symptomatic treatment is the method of choice in the management of fibrous dysplasia of the sphenoid bone. Surgical procedure is recommended only in patients with severe functional disorders, progressive deformations or malignant transformation. Radiation therapy is contraindicated due to the risk of neoplastic transformation [2,12]. Literature includes few reports regarding FD being transformed into a malignancy, most commonly osteosarcoma (~ 70% of transformation cases) [4]. The risk of lesion becoming malignant is assessed to be less than 1% for monoosteotic disease and about 4% for polyosteotic disease [4, 13]. Malignancy develops most frequently within the facial cranium bones (46% of cases) [14]. To date, no cases have been described regarding fibrous dysplasia located within the sphenoid sinus turning into a malignant lesion [7].

**CASE REPORT**

A 58-year-old female patient presented at the neurologist’s in the autumn of 2013 due to persistent headaches she had been experiencing for many ears. The patient did not complain of any other symptoms of sinusitis such as the flow of secretion along the posterior wall of the throat, nasal congestion or running nose. Neurological examination revealed no deviations from normal. However, contrast-enhanced CT scan of the head was performed in October 2013 to reveal thickening of sphenoid sinus walls [and an] abnormal skeletal structure with inner area filled with a soft tissue of the density of ca. 95 HU with uneven outline and approximate size of 15x10 mm. Further diagnostic examinations were suggested. A magnetic resonance scan performed in March 2014 suggested the presence of tumor or atypical mucocele within the sphenoid sinus, without penetration into the neighboring cerebral tissues. Due to the ambiguity of radiological diagnosis, a decision was made to perform a tissue biopsy. The patient was hospitalized at the Department of Otolaryngology of the Municipal Hospital in Sosnowiec where CT scan was repeated to reveal the typical ground-glass matrix image within the sphenoid sinus [Fig.1]. Subsequently, the patient was subjected to endoscopic collection of a tissue section under neuronavigation facilitating targeted collection of material from the visible foci of reduced bone density. The result of pathomorphological examination unambiguously pointed to fibrous dysplasia. Due to the extent of the lesion and the absence of infiltration onto the adjacent structures, either pericerebral fluid spaces or the brain, and in line with the management described in the literature, surgical treatment was abandoned. Follow-up MRI scan taken in July 2014 (Fig. 2, 3) and sinus CT scan taken in October 2014 confirmed the lack of lesion progression.

In addition, the patient was hospitalized at the Department of Internal Medicine and Metabolic Diseases of the Independent Public Clinical Hospital No. 7 of the Medical University
of Silesia in Katowice, where parosteal osteoma of the lateral malleolus of the left ankle was detected, but any disturbances of calcium and phosphorus metabolism were excluded.

To date, the patient has remained under continuous supervision by the Laryngological Outpatient Clinic.

DISCUSSION

Diagnostic difficulties in the diagnosis of sphenoid bone dysplasia are due not only to the rare location of lesions within this anatomical region, but also to the high diversity of CT and MRI scans due to varied proportions between fibrous and bone tissue in the involved bones. In MRI scans, fibrous dysplasia presents with low-intensity signals in T1-weighted sequences while generating signals of highly variable intensity, from high and medium to low intensity, in T2-weighted images [4,8,15]. These differences are due to the variable content of collagen within the cells and the trabecular tissue. In computed tomography, the disorder generates the “ground-glass” image and contract enhancement [1,16]. Also characteristic is high saturation in the range of 70-130 HU as compared to other benign bone lesions and e.g. osteomyelitis (20-40 HU) [4,17]. This feature was also confirmative of diagnosis in the reported case (ca. 95 HU).

It is worth mentioning that first radiological description and classification of fibrous dysplasia was performed by Fries in 1957 [18]. He identified 3 radiological types of the disease: the most characteristic form of a “ground-glass” cyst (currently observed in computed tomography scans), a sclerotic form and a pagetoid form [4,7,16]. In the era of the common use of computed tomography, the technique has also become the main technique used for diagnosing bone diseases.

Similarity of radiological images to those characteristic for a number of other bone diseases is a factor that makes the diagnosis even more difficult [Table 1]. Most common diagnostic errors are encountered in imaging studies (CT, MRI) in which fibrous dysplasia is confused with chondroma, chondrosarcoma, meningioma, sinusitis, mucocele, inflammatory polyp, inverted polyp, invasive pituitary adenocarcinoma, aspergillosa, or old bone fractures [1,19,20]. For example, mucocele would be characterized by a higher intensity of signals in T2-weighted images [7]. In case of computed tomography, similarities to FD image may be encountered in cases of aspergillosa due to central “turbidity” of bone tissue ima-
...and osteosarcoma, although it is characterized by different location of turbidities [8].

In case of the patient of interest, diagnoses suggested from imaging studies included atypical mucocele and proliferative disease. Similar diagnostic difficulties were encountered by other authors. Khalil et al. described magnetic resonance imaging similarities to nasopharyngeal carcinoma or chordoma [7].

Due to these diagnostic doubts, it was decided that the decisive role in the differential diagnosis of fibrous dysplasia would be played by the result of the histopathological examination used as the basis for final diagnosis [4]. Macroscopically, the lesion is of granular structure and grey or brown color. Small effusions or cysts may be visible. Usually, the lesion is surrounded by a hem of sclerotic bone [21, 22]. Microscopically, FD forms mainly a fibrous stroma, usually poorly vascularized and hypocellular. Fibroblasts characterized by improper maturation and differentiation are predominant among the reported cells. Of note is the absence of osteoblasts and osteoclasts. No atypia is observed while mitotic division figures are rare. A small part of the tissue may consist of cartilaginous elements [13, 22]. The trabeculae within the lesion, built of immature bone tissue, are of irregular shape with numerous bends and bifurcations [21]. Numerous authors described this image as resembling “alphabet soup” or Chinese writing [4, 13, 16, 21], the transverse lines across the trabeculae may imitate clinical presentation of Paget’s disease. The absence of osteoblastic sheath around the trabeculae, which is typical for healthy bone tissue, is crucial for the diagnosis of FD [13].

Although the collection of examination material poses no problems in case of “standard” locations, more surgical experience is required in case of sphenoid bone lesions. According to the international literature, biopsy is collected from endoscopic access as it warrants a minimally invasive way for accessing locations requiring classic transcranial approach. In addition, endoscopic access ensures a more anatomical approach to these locations while eliminating the need for an extensive lateral to the skull base, ensuring panoramic view and facilitating better identification of lesion borders than that achieved in the case of microscopic access [1, 23, 24]. Neuronavigation facilitating precise sections targeted at the foci of reduced density or the dense bone so as to obtain representative study material is a very helpful tool. Intraoperatively, FD is macroscopically described as dysmorphic, dysplastic bone causing deformations of adjoining structures [1, 2].

Table I. Microscopic differential diagnosis of fibrous dysplasia [1, 2, 4, 11, 22, 27, 30, 31, 32, 33].

<table>
<thead>
<tr>
<th>Nosocomial entity</th>
<th>Feature similar to FD</th>
<th>Feature dissimilar to FD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Osteosarcoma</td>
<td>hypercellular connective tissue disordered young bone tissue islets possible cartilaginous tissue elements</td>
<td>significant atypia, numerous division figures cortical bone damage, possible bone marrow infiltration</td>
</tr>
<tr>
<td>Osteoblastoma</td>
<td>hypercellular connective tissue disordered young bone tissue islets possible cartilaginous tissue elements</td>
<td>osteoblastic sheaths around the trabeculae absence of bone marrow morphotic elements</td>
</tr>
<tr>
<td>Chondroma</td>
<td>mature hyaline cartilage possible features of atypia</td>
<td>predominance of cartilaginous tissue</td>
</tr>
<tr>
<td>Paget’s disease</td>
<td>hypercellular connective tissue disordered young bone tissue islets</td>
<td>activation of osteoclasts trabecular thickening mosaic-like arrangement of new bone tissue Cellular inclusions of measles virus and RSV</td>
</tr>
<tr>
<td>Recklinghausen’s disease</td>
<td>hypercellular connective tissue</td>
<td>proliferation of peripheral nervous tissue elements</td>
</tr>
<tr>
<td>Chronic osteitis</td>
<td>hypercellular connective tissue disordered young bone tissue islets</td>
<td>activation of osteoclasts granulation tissue</td>
</tr>
<tr>
<td>Central giant cell granuloma</td>
<td>hypercellular connective tissue disordered young bone tissue islets</td>
<td>mononucleate inflammatory cells giant multinucleate cells</td>
</tr>
<tr>
<td>Aneurysmal bone cyst</td>
<td>ditto</td>
<td>ditto</td>
</tr>
<tr>
<td>Ossifying fibroma</td>
<td>hypercellular connective tissue disordered young bone tissue islets</td>
<td>osteoblastic sheaths around the trabeculae numerous cement deposits and dysmorphic calcifications lower reactivity to keratin, neurofibromin, S-100 protein and Leu 7</td>
</tr>
<tr>
<td>Brown tumor of bone in the course of primary hyperparathyroidism</td>
<td>hypercellular connective tissue atrophy of normal trabecular bone structure</td>
<td>cystic lesions, focal deposits of hemosiderin hemosiderin-laden macrophages osteoclast-like multinucleate giant cells</td>
</tr>
</tbody>
</table>
Additional identification of optic nerve compression is an important aspect that should be taken into account when diagnosing fibrous dysplasia of sphenoid bone. Although the lesion did not involve the optic nerve in the reported case, it may be helpful to mention this problem. Numerous authors highlight that in the case of asymptomatic compression of the optic nerve, decompression is not absolutely necessary, or even recommended [25,26,27]. Decompression, most commonly achieved by endoscopic method involving neuronavigation, is necessary in case of symptoms of compression of the optic nerve or structures crossing the superior orbital fissure [7,28,29].

In addition, Mladina et al. point to the necessity of ruling out possible infiltration of adjacent vessels, i.e. internal and external carotid arteries, by means of angiographic examination [8].

When analyzing the possibilities for the treatment of fibrous dysplasia of cranial bones, most authors are inclined towards an anticipatory approach with regular assessment of the extent of infiltration [7]. Surgical procedure is recommended only in cases when infiltration involves structures of importance for patient’s life and functioning. Khalil et al. described a case of a 26-year-old male with fibrous dysplasia of sphenoid bone progressing to the adjacent structures and manifesting by impaired vision, who was subjected to frontotemporal craniotomy with optic nerve decompression and resulting improvement in the field of view [7].

The efficacy of bisphosphonates that inhibit osteoclastic bone resorption and are used in the treatment of osteodystrophies, is highly dubious and the treatment is not recommended [27].

In conclusion, fibrous dysplasia of sphenoid bone is a rare disorder. The diagnosis is based on clinical radiological and histomorphological examination. Early diagnosis is difficult due to non-specific symptoms. The choice of the treatment method depends on the clinical condition of the patient.

References

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