SINONASAL FIBROUS DYSPLASIA: OUR 10-YEARS EXPERIENCE

Dmitry Kapitanov¹, Anastasia Kostousova¹, Marina Nersesyan¹, Dilyana Vicheva²

1) Department of Otorhinolaryngology, Burdenko Neurosurgical Institute, Moscow, Russian Federation,

2) Department of Otorhinolaryngology, Medical Faculty, Medical University, Plovdiv, Bulgaria..

ABSTRACTS:

Fibrous dysplasia (FD) is a rare benign bone tumor characterized by replacement of normal bone by fibro-osseous connective tissue. FD accounted for 7 % of all the benign bone tumors. The incidence of FD involving the sinonasal tract is unknown.

The purpose of this study was to assess the incidence of sinonasal involvement by FD, to analyze the clinical characteristics, radiological features, and management of patients with fibrous dysplasia in order to understand the indications for surgical treatment. We also present some CT images of our patients to demonstrate typical radiographic signs of FD.

Materials and methods: a retrospective review of 19 patients. Distribution of the cases according to the clinical and radiological features was described, management and results were presented.

Results: among 78 patients with craniofacial FD, 19 (24, 36%) patients presented sinonasal involvement. The average age was 47 years. 1 patient was with monostotic form, the rest 18 – with polyostotic. The most common site of the lesion was sphenoid and ethmoid sinuses, the most common complaint – headache, less often – nasal obstruction with or without nasal discharge. 16 patients underwent purely endoscopic removal, the rest 3 – combined neurosurgery with endoscopic assistance. Median follow-up period was 21 months, and there were no continued growth in our observed group.

Conclusions: FD is a very rare pathology. In this particular group of patients, endoscopic treatment showed a good result, but this pathology still needs to be further studied, and patients require a long-term follow-up. Endoscopic surgery is challenging as FD bleeds freely, and should be only contemplated by an experienced rhinologist.

Keywords: Fibrous dysplasia, sinonasal fibrous dysplasia, skull base, functional endoscopic sinus surgery,

INTRODUCTION:

Fibrous dysplasia (FD) is a benign tumor characterized by replacement of normal bone by fibro-osseous connective tissue. It is a congenital disorder related with a set of mutations in the GNAS1 gene, located on chromosome

20q2. FD comprises 2.5% of all bone tumors and 7.5% of all benign bone neoplasms [1-3]. The incidence of FD involving the sinonasal system is unknown [1]. According to the literature, maxilla, orbit and mandible are the most commonly affected areas in head and neck [1, 2, 4], however involvement of paranasal sinuses is also described [5]. In this regard, the most common complaints are facial asymmetry and painless swelling of jaws. Less frequently, patients suffered from headache or bone pain, nasal discharge with or without nasal obstruction. In cases with affected skull base - symptoms of cranial nerves compression such as anosmia, hearing loss, facial paresis, trigeminal dysfunction and ophthalmic symptoms such as visual impairment, exophthalmos, ptosis, eyelid edema may take place [5-7].

The causes of the disease are associated with disorders in the chromosome structure - a GNAS1 gene mutation, which results in the inhibition of proliferation and differentiation of stromal cells of normal bone tissue. Fibrous dysplasia can occur at any age, but the debut of the disease most often occurs in children and young age, due to the genetic nature of the disease. Men and women are usually equally affected [1-3].

FD can involve different bones, including the skull bones. Three forms of FD are usually distinguished: it can be monostotic (70-85%), involving only one bone, or polyostotic (15-25%), involving more than one bone and third rare form (<10%) is a McCune-Albright syndrome characterized by polyostotic form combined with precocious puberty and “café au lait spots”, preferentially involves children. Craniofacial bones are typically involved in about 10-20% of patients with monostotic form and in up to 80-100% with polyostotic form [1, 2, 4, 5].

Computed tomography is considered the best technique for diagnosis FD. Radiological characteristics of the lesions differ regarding to the bone and fibrous matrix ratio. There are three typical radiographic patterns: pagetoid (56%), sclerotic (23%), and cystic (21%) (Figure 1). Sclerotic pattern mostly involves facial and skull base bones and characterized by a high-density signal on CT. Pagetoid pattern characterized by equal rate of the bone-fibrous matrix, described as a classical «ground glass» sign, with typical «swelling» of dense bone structures, without clear boundaries, which looks differently from osteoma or ossifying fibroma [5, 8].

Fig. 1. Computed tomography, bone window mode. Radiographic patterns: pagetoid, sclerotic, and cystic. Asterisks indicates the FD lesion.

- a) The maxillary bone and maxillary sinus lesion (pagetoid pattern) in axial projection.
- b) Ethmoid and sphenoid bones and sinuses lesion (sclerotic pattern) in coronal projection.
- c) Ethmoid bone and sinus lesion (cystic pattern) in axial projection

Magnetic resonance imaging (MRI) is an additional method for the diagnosis of FD, which helps to distinguish the soft tissue character of the lesion and helps to visualize the intracranial growth of the tumor. MRI shows a hypointense signal on T1-weighted sequence in ossified or fibrous parts of FD. On T2-weighted images, MR-signal is also hypointense with some heterogeneity [5].

Microscopically FD characterized by replacement of normal bone tissue by fibrous connective tissue, and the new formed abnormal fibro-osseous tissue consists of numerous small, irregular-shaped trabeculas. Trabecular structures show an abnormal arrangement, and it looks like Chinese letters or alphabet soup [2].

Biopsy is not always required. Exacerbation of symptoms is an indication for biopsy. Non-symptomatic cases are usually require a long-term follow-up.

According to the literature, there are some opinions that FD can be treated with bisphosphonates conservatively, it may decrease the incidence of fractures and bony pain [1]. Wang Y. et al. consider that bisphosphonates may suppress high bone turnover to partially control the activity of the disease, and are well tolerated in most patients. Zoledronic acid has similar effects as pamidronate in controlling disease activity [9, 10]. There are also some publications about osteonecrosis of the jaw caused by bisphosphonates treatment in patients with FD [11]. This potential complication stopped us from using bisphosphonates, thus we do not have such experience.

Differential diagnosis is firstly conducted with osteoma and ossifying fibroma. However, there are a number of diseases that can also be included in this list such as, aneurysmal bone cyst, osteosarcoma, osteochondroma, cemento-osseous dysplasia, eosinophilic granuloma, deforming osteitis (Paget's disease) and others [1, 2, 12-14] (Table 1, Figure 2) .

Table 1. General characteristics of patients.

	Patients n, %
Sex (Female/male)	10 (%) / 9 (%)
Age	13 – 55 (mean 47) years
Sinonasal FD (from patient with craniofacial FD)	19 (24%)
Monostotic	18 (95%)
Polyostotic	1 (5%)
Previous surgery	2 (10,5%)

Fig. 2. CT scans: axial projection (a, c), sagittal projection (b, d), bone window mode. Differential diagnosis of FD. Asterisks indicates different lesions. a) Ethmoid bone and sinus fibrous dysplasia. b) Frontal sinus osteoma. c) Ethmoid bone and sinus cemento-osseous dysplasia. d) Ethmoid bone, sphenoid bone and nasal cavity aneurysmal bone cyst.

Many authors in their publications describe the massive craniofacial lesions, located in maxilla, orbit and mandible. In this regard, the most common complaint is the facial asymmetry [1-3]. With such manifestation of the disease, patients are likely to seek the neurosurgeon or maxillofacial surgeon examination and often need surgical treatment using external approaches.

In our paper, we would like to share our experience of management patients with FD, which extended into the nasal cavity and paranasal sinuses, and also skull base can be involved. Hope our experience will be helpful for otolaryngologists.

MATERIALS AND METHODS:

78 patients with craniofacial fibrous dysplasia had been treated from 2008 to 2018 in Burdenko Neurosurgical Institute. We included 19 patients (24,36%) with sinonasal fibrous dysplasia from 13 to 55 years old (mean - 47) in our study.

Male-to-female ratio was approximately equal (10 women and 9 men). The rest 59 (75, 64%) patients were excluded from our study because they had extended craniofacial FD and needed neurosurgical treatment.

The diagnosis was based on clinical symptoms, CT

findings and confirmed histologically in each case. Some of the patients additionally underwent MRI.

At our department, 19 patients with sinonasal FD had been treated surgically in 10 years. 16 (84, 21%) of them – underwent purely endoscopic removal, the rest 3 (15, 79%) – combined neurosurgery with endoscopic assistance.

Median follow-up period was 21 months, and there were no recurrences in our observed group. However, 11 patients were lost from follow-up group.

RESULTS:

Among 78 patients with craniofacial FD 19 (24, 36%) patients presented the following involvement: facial bones and skull base lesion spreading into the paranasal sinuses and/or to the nasal cavity. All of them had been treated in our ENT department.

There was one patient (5, 26%) with polyostotic form and the rest 18 (95, 74%) – with monostotic form. There was none with McCune-Albright's syndrome in our series. 17 (89, 47%) patients were with primary diagnosed FD, 2 (10, 53%) patients had previous surgical treatment at another hospital, so they had continued growth of the tumor (Table 2).

Table 2. Differential diagnosis

	Fibrous dysplasia	Ossifying fibroma	Osteoma	Aneurismal bone cyst
Incidence	Not known (2.5% of all bone tumors)	Not known	0.43% to 3%	Not known (1%–2% of all primary bone lesions)
Age of presentation	1 - 2 decade	2 – 4 decade	3 - 4 decade	Under 20 years
Male to female ratio	1 : 1	1 : 5	1,5 – 3, 1 : 1	1 : 1
Site	Maxilla, mandible, orbital bones	Mandible	Frontal sinus	Skull base, ethmoid sinus
Symptoms	Facial asymmetry	Painless swelling, headache, nasal obstruction	Frontal headache	Neurological symptoms when skull base is involved
Radiographic signs	«Ground glass» appearance on CT	Expansile mass with sharp demarcation	Homogenous, dense, well-circumscribed	CT reveals bony expansion, a widened diploic space, and fluid-fluid interfaces
Treatment	Observation, surgery in symptomatic cases	Observation; if possible complete resection in extended cases	Observation, surgery in symptomatic cases	Surgical curettage and grafting. Radiotherapy may trigger malignant changes

FD of facial and skull base bones in our series hadspreaded mostly into the sphenoid sinus – 13 (68, 42%) patients, also into the ethmoid sinus – 9 (47, 37%), nasal cavity – 4 (21, 05%), frontal sinus – 3 (15, 79%), orbit - 3 (15, 79%) and maxillary sinus – 1 (5, 26%). 12 (63, 16%) patients had isolated FD and in 7 (36, 84%) patients lesions involved multiple sites.

The most common symptoms were headache – 16 (84, 21%), nasal obstruction – 7 (36, 84%), rhinorrhea – 7 (36,

84%), less often were cranial nerves compression symptoms – 3 (15, 79%) and ophthalmic symptoms – 3 (15, 79%). Among three patients with ophthalmic symptoms: one (5, 26%) patient was with visual impairment, one with ptosis, and another one with exophthalmos. Among three patients with neurological symptoms, there were 2 (10, 53%) patients with hyposmia and 1 (5, 26%) patient with trigeminal dysfunction (Table 3).

Table 3. Site of the lesion and main symptoms

Site	Patients n, %
Sphenoid bone and sinus	13 (68, 42%)
Ethmoid bone and sinus	9 (47, 37%)
Nasal cavity	4 (21, 05%)
Frontal bone and sinus	3 (15, 79%)
Orbital bones	3 (15, 79%)
Symptoms	Patients n, %
Headache	16 (84, 21%)
Nasal obstruction	7 (36, 84%)
Rhinorrhea	7 (36, 84%)
Anosmia/hyposmia	2 (10, 52%)
Ptosis	1 (5, 26%)
Trigeminal dysfunction	1 (5, 26%)
Visual impairment	1 (5, 26%)
Exophthalmos	1 (5, 26%)

The CT findings in our series of FD were typical: there were increased bony thickness with widening of the diploic spaces with marrow involvement, which was usually homogeneous radiodensity with the loss of the trabecular pattern and varying degree of obliteration of the sinuses (Figure 3, 4). Although the CT is the best technique for visualization of FD lesions, MRI can be helpful to evaluate the cranial nerves involvement, including the optic nerve. But in spite of the contrast administration, the cellularity and activity of the lesions do not correlate well on MRI with signal intensity and enhancement degree or pattern.

Fig. 3 a, b. CT scans, bone window mode. Example of indications for endoscopic removal. Asterisks indicates the FD lesion. **a)** Sphenoid bone and sinus FD lesion in coronal projection. **b)** Sphenoid and ethmoid bones and sinuses FD lesion in axial projection.

Fig. 4. CT scans: coronal projection (**a**), sagittal projection (**b**); bone window mode. Example of indications for combined removal. Asterisks indicates the FD lesion.

On these CT scans FD of the skull base spreading into the ethmoid sinus, nasal cavity, sphenoid bone, orbit and anterior cranial fossa.

In our series, most patients were with sclerotic and pagetoid radiographic patterns. Sclerotic pattern characterized by dense, homogeneous lesion on CT, and pagetoid pattern presented with “ground-glass” opacities and bone expansion. A few patients were with cystic pattern, which was seen in younger patients and characterized by round or oval “cystic” lesions and sclerotic borders (Figure 1).

All the patients with sinonasal FD underwent endoscopic procedures. 3 (15, 79%) patients with exacerbation of the symptoms and not clear radiographic picture underwent endoscopic biopsy to confirm the diagnosis of FD (Figure 5).

Fig. 5. Endoscopic view of the sphenoid sinus, 0° endoscope. **a)** Using high-speed bur for removing the lesion. **b)** At the location of the suction, we can see dense fibro-osseous masses of the tumor

Most of the patients with headache underwent endoscopic surgery with the purpose of surgery to create the cavity of the sphenoid or ethmoid sinuses, to drain the sinuses and to drain the mucocoele. All the patients relieved or symptoms decreased after the surgery. The similar situation with the cases with bony nasal obstruction to improve nasal breathing.

A few patients with headache and cranial nerves compression with sclerotic form of FD underwent endoscopic surgery under intraoperative navigation in order to decompress the cranial nerves and to create the cavity. During the drilling of the unstructured bony architecture in these cases, we constantly have to cool the drill by overfilling water

irrigation in order not to bur the nerve.

As the result, we had one patient with improved vision, and one patient with exophthalmos decreased. However, another patients with cranial nerves compression symptoms were lost from follow-up group.

We used intraoperative navigation in 4 (21, 05%) cases (when it was available) because in patients with FD

the bony expansion makes difficult to find out the landmarks and therefore easy to mix them during the surgery and to get the complication.

Nowadays, we use navigation in each case with changed nose or sinus anatomy and because of the technical difficulties during the surgery such bony uncontrolled bleeding (Table 4).

Table 4. Management of patients with FD.

Endoscopic biopsy	3 (15, 79%)
Endoscopic resection	13 (68, 42%)
Combined approach (Endoscopic + bifrontal craniotomy)	3 (15, 79%)
Using navigation system	4 (21, 05%)
Mean follow-up period, months	21 [4;37]

DISCUSSION:

According to the literature, the monostotic form occurred in 75-80% of patients with FD, and polyostotic – in 15-20%. The most frequent localization of the FD lesion is the maxilla, orbital bones and mandible. Thus, the most common complaint is the painless swelling of jaws leading to facial asymmetry [1, 2, 4, 5, 8].

In spite of variety of publications, there is insufficient information about incidence of sinonasal FD and about follow-up of patients with FD in general.

C. Duan et al. wrote that the ratio of monostotic and polyostotic forms was equal [4, 6]. They reviewed 28 patients with sinonasal FD, the most common sites of the lesion were sphenoid sinus (50%), nasal cavity (39%) and ethmoid sinus (36%). The median follow-up period was 29 months, three patients were lost for follow-up and 2 patients from 25 had continued growth [6].

In our series incidence of sinonasal FD was 24, 36% (from patients with craniofacial FD), almost all patients were with polyostotic form (94, 74%), the most common complaints were headache, nasal obstruction/nasal discharging, rare – neurologic and ophthalmic symptoms and the most common sites of the lesion - sphenoid, ethmoid sinuses and nasal cavity, which complies with the world literature [1-5].

Since the patients with FD need a long-term follow-up, the important point is to determine the indications for surgical treatment.

According to the literature, it is considered that patients with FD should be operated endoscopically in case of neurological and ophthalmologic symptoms. In our opinion, each such case should be considered individually and discussed with a neurosurgeon. In case of technical feasibility and in case of extensive experience of the rhino surgeon, such patients can be treated endoscopically.

In our hands, a biopsy is indicated in cases of exacerbation of the symptoms, not clear radiographic signs and rapid growth of the lesion.

The indications for endoscopic surgery are:

1) Ophthalmic and neurological symptoms above mentioned.

2) Blocked sinus ostium, which leads to persistent, intractable headache – to relieve the pain.

3) Blocked sinonasal tract leading to nasal obstruction with or without regular nasal mucous - purulent discharge from the nose – to improve nasal breathing.

The indications for combined surgical treatment are:

1) FD spreading intracranially, rough facial deformity.

2) Ophthalmic symptoms such as visual impairment, oculomotor nerve dysfunction, proptosis, eyelid edema – spreading into the orbit, optic nerve canal, cavernous sinus.

In our study median follow-up period were 21 months (range from 4 to 37) and there were no continued growth in our observation group. However 11 patients were lost from follow-up group, but they didn't return back neither for additional treatment nor for examination during this period of time, in spite of Burdenko Neurosurgical Institute is the main institute for skull base surgery in Russia. Thus, we indirectly assume that these patients may do not have continued growth.

CONCLUSIONS:

FD is a very rare pathology. This study found that the incidence of sinonasal FD was 24, 36%, that monostotic form prevails over polyostotic form (95% / 5%). The most commonly involved sites were sphenoid, ethmoid sinuses and nasal cavity, the most common complaints – headache, nasal obstruction/nasal discharging, rare – neurologic and ophthalmic symptoms. In this particular group of patients, endoscopic endonasal surgery was the method of choice, but this pathology still needs to be further studied, and patients require a long-term follow-up. Endoscopic surgery is challenging as FD bleeds freely, and should be only contemplated by an experienced rhinologist.

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Address for correspondence:

Kostousova Anastasia, E-mail: kostousova@yahoo.com
Professor Kapitanov D.N., E-mail: Dkapitanov@nsi.ru