CASE REPORT

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Prosthetic treatment of adolescents with craniomaxillofacial fibrous dysplasia

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Keywords

craniomaxillofacial fibrous dysplasia, prosthetic rehabilitation, adolescent patient

Summary

Craniomaxillofacial fibrous dysplasia may cause structural anomalies in the facial part of the skull, leading to anatomical and functional abnormalities in the stomatognathic system, requiring multi-disciplinary treatment. Depending on the location of dysplastic lesions, stage and course of the disease, each patient presents with a specific picture of the disorder and requires an individual therapeutic approach. Prosthetic treatment of patients at developmental age requires regular check-ups and frequent corrections or replacement of prosthetic restorations. This is particularly important in patients whose prosthetic field is deformed in such disorders as fibrous dysplasia or following numerous surgical procedures. The effectiveness of therapeutic procedures, which in such cases must be multidisciplinary, depends mainly on the commitment and cooperation of the patient and their carers. We present difficulties in the prosthetic rehabilitation of maxillofacial dysplasia based on the example of a patient diagnosed with fibrous dysplasia, severe oligodontia multiorgan congenital malformations, and a history of numerous surgical procedures within the stomatognathic system.

Introduction

Fibrous dysplasia is a genetic disorder involving local replacement of bone with fibrous tissue. The craniomaxillofacial form usually manifests in small children and is usually polyostotic – multifocal, i.e. many bones are affected.

Craniomaxillofacial fibrous dysplasia is a chronic disease. Initially, local soft tissue oedema and localised pain may occur (1). Later stages involve bone deformities, which may lead to stomatognathic anatomical disorders and, consequently, functional impairment, such as difficulty opening the mouth, chewing and articulating sounds (2). Progressive dysplastic lesions may cause premature loss of primary teeth and dental-occlusal abnormalities affecting permanent dentition as a result of impaired tooth eruption and development. Severe, shooting pain is typical for exacerbations involving bone and soft tissue inflammation (3). Nerve compression caused by tissue hyperplasia may also cause facial paresthesia. Depending on the location and size of lesions, other disorders may also occur: change of visual acuity, exophthalmos, tear duct obstruction, hearing impairment, nasal obstruction. Pathological fracture of the affected bone may occur in extreme cases (4-6).

Many patients are asymptomatic in the initial stages of the disease, and the diagnosis is based on radiography performed for a different reason, and then confirmed histologically. The radiological picture undergoes a specific evolution in the course of the disease. Cyst-like osteolytic formations without a distinct border between compact and trabecular bone are observed at early stages of the disease.

These are followed by blurred bone structures producing a characteristic appearance compared to "ground glass", "cotton wool", "snow storm" or "orange peel". Advanced stages produce mixed appearance due to the presence of both sclerotic lesions and areas of bone thinning. Differential diagnosis should include ossifying fibroma, Paget's disease, oral giant cell lesion, a unicameral bone cyst and cherubism (7).

Disturbances in bone structures of the facial part of the skull, which occur in the course of disease, may cause anatomical and functional abnormalities within the stomatognathic system, requiring multi-disciplinary treatment. Surgical correction of the affected tissues is often needed. Oligodontia and tooth dislocations often require orthodontic treatment and prosthetic rehabilitation. Prosthetic treatment of adolescents with craniomaxillofacial fibrous dysplasia poses difficulty due to progressive changes in the prosthetic field, especially in patients with incomplete bone growth who are in the active phase of the disease.

Case report

A 13-year-old boy was reported to the Department of Prosthetic Dentistry of the Medical University of Warsaw for prosthetic consultation. Medical history of diagnosed craniomaxillofacial fibrous dysplasia and multiorgan congenital malformations: coarctation, thoracic and abdominal aortic hypoplasia, congenital oesophageal and anal stricture, bile duct anomalies, bilateral cryptorchidism; severe growth deficiency (ongoing growth hormone therapy), hypertension, hypocalcaemia (calcium supplementation).

The patient was treated in the Institute of Mother and Child in Warsaw, where he underwent multiple surgeries of the stomatognathic system. Craniomaxillofacial fibrous dysplasia was diagnosed based on histopathology of tissue specimens (2010). Dysplastic lesions of the maxillary alveolar process and the alveolar portion of the mandible never produced pain. First manifestations of the disease occurred already in the neonatal period in the form of epulis of the maxillary alveolar ridge, which was removed along with the primary tooth bud. The aim of subsequent surgical procedures in later years was to remove mandibular epulis and dysplastic lesions of the maxillary alveolar process and the alveolar portion of the right mandible. Surgical procedures involved successive removal of tooth buds from the affected areas.

Only tooth 21 and tooth 12 with grade II mobility were present in the oral cavity before prosthetic treatment (fig. 1, 2). Severe deformation of the tissues in the prosthetic field involved the anterior and right-sided alveolar portion of the edentulous mandible and was a consequence of previous surgical removal of dysplastic lesions and impacted teeth (fig. 3). Reduced height of occlusion and changes in facial features were noticeable (fig. 4). Panoramic radiography showed bone thinning in the region of conglomerate mass of odontogenic tissue in the central part of the mandible (fig. 5). CBCT revealed multiple maxillary and mandibular anomalies (fig. 6). Due to the state of prosthetic field, mandibular class V and maxillary class IV defects according to Galasińska-Landsbergerowa, treatment using removable dentures, i.e. partial removable lower denture and a complete lower denture, was scheduled. Prosthetic restorations were performed in accordance with the general guidelines, using the functional selective pressure impression technique and determining correct spatial mandibular relationships (fig. 7). Additionally, a soft silicone-based denture liner (Mollosil, Detax) was used. Excellent retention and stabilisation of the upper denture were achieved. However, the retention of lower denture was unsatisfactory due to the very unfavourable conditions in the prosthetic field. During follow-up visits, the patient reported difficulties adapting to

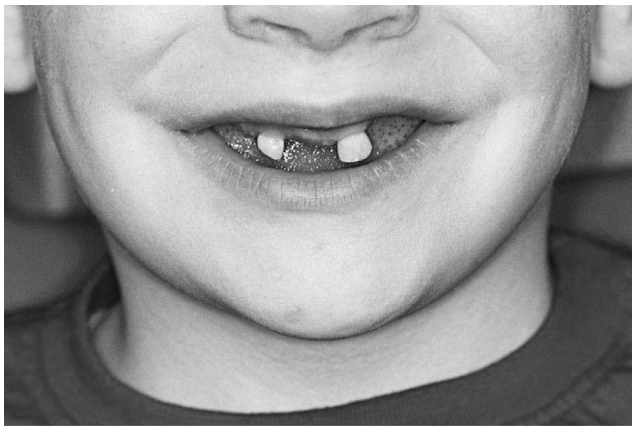

Fig. 1. Extraoral photograph – before prosthetic treatment

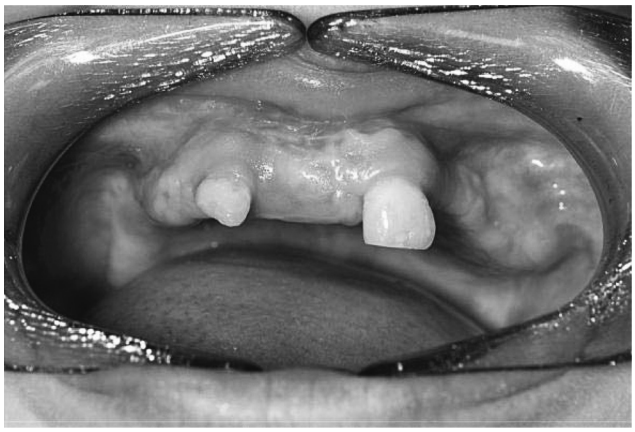

Fig. 2. Intraoral photograph – maxillary prosthetic field before prosthetic treatment

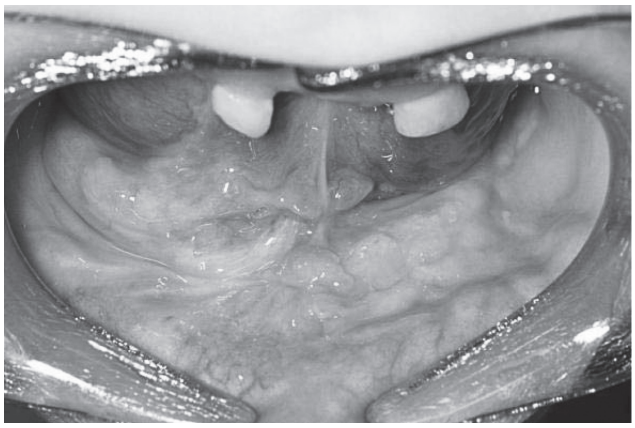

Fig. 3. Extraoral photograph – mandibular prosthetic field before prosthetic treatment

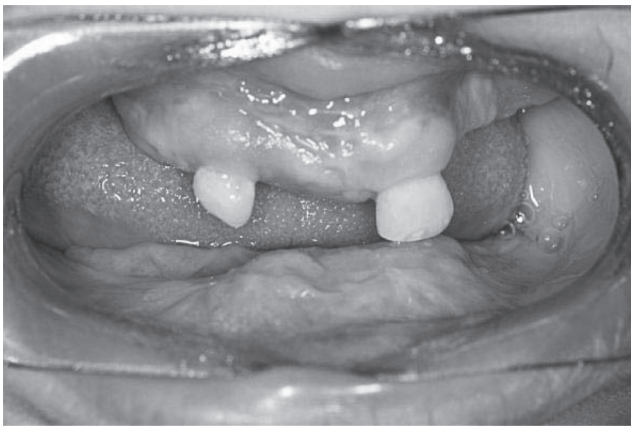

Fig. 4. Intraoral photograph – intermaxillary conditions

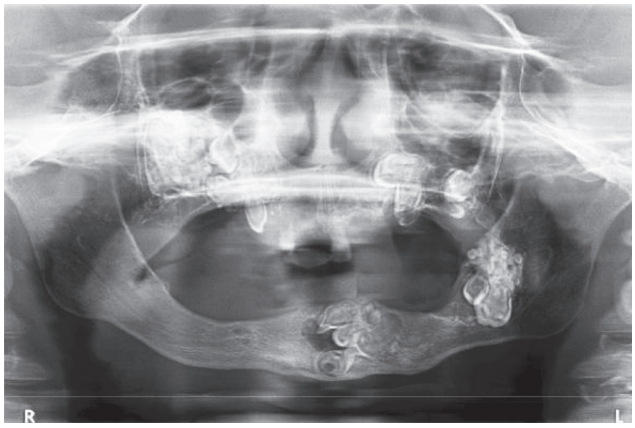

Fig. 5. Panoramic radiograph before prosthetic treatment

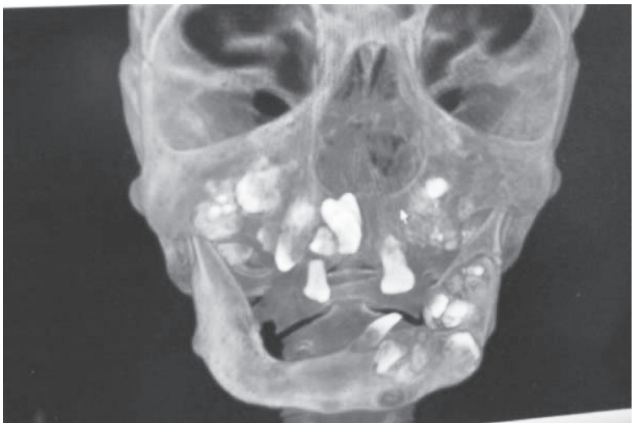

Fig. 6. Three-dimensional CBCT image – before prosthetic treatment

dentures due to increased salivary secretion and difficulty maintaining the lower denture in the oral cavity while chewing, speaking and swallowing. Further diagnosis was planned to assess the progression of lesions and the possibility of interdisciplinary treatment, including implantoprosthetic treatment.

Prosthetic restorations were not used systematically by the patient. Tooth 12 was lost in a short time due to facial injury during a football match. The denture was repaired, and the tooth 12 was placed in the upper denture (fig. 8). The patient failed to regularly report for follow-up visits due to general medical complications associated with multiorgan congenital defects and frequent hospital stays. The boy discontinued the use of dentures due to a decubitus ulcer under the upper denture plate and an erupting tooth 13 (fig. 9). Following a telephone conversation with the mother, a check-up visit was scheduled, during which prosthetic restorations were corrected and again lined with smooth material. In the next stage of treatment, further surgical procedures were planned to correct midline mandibular lesions and to monitor lesions in the prosthetic field in the region of conglomerate odontogenic tissue and impacted teeth. Correction of prosthetic restorations or placing new restorations, depending on the extent of postoperative changes in the prosthetic field and growth-related facial changes, were planned after surgical intervention.

Further prosthetic rehabilitation and planned interdisciplinary treatment were postponed due to frequent hospital stays coinciding with our visits and, most of all, the lack of willingness to cooperate on the part of the mother and the boy.

Discussion

Fibrous dysplasia is a congenital metabolic disorder, in which normal trabecular bone is converted into compact fibrous tissue containing cartilaginous and osseous components, as well as calcifications (7). Fibrous dysplasia is a genetic disorder. As a result of mutation of GNAS1 (chromosome 20q13.2-13.3) coding for the α subunit of Gsα protein and an increased synthesis of IL-6, which is

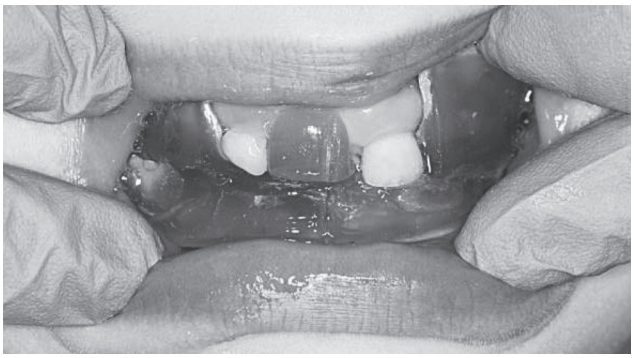

Fig. 7. Extraoral photograph – determining central occlusion

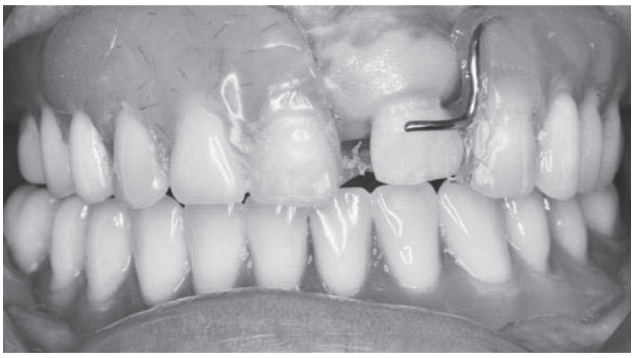

Fig. 8. Intraoral photograph – after prosthetic treatment

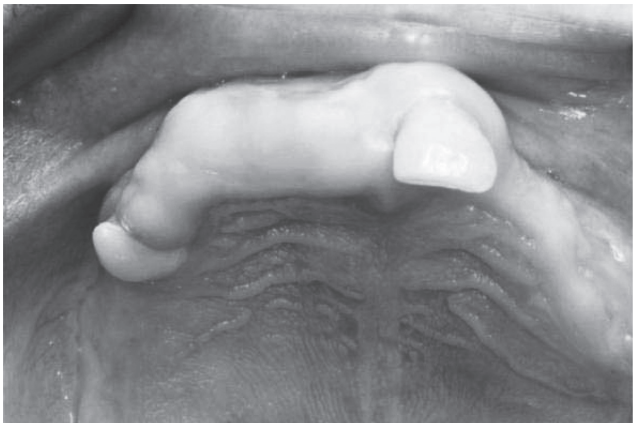

Fig. 9. Extraoral photograph – erupting tooth 13

responsible for osteoclast differentiation, proliferation of morphologically and functionally abnormal osteogenic cells occurs (3, 8, 9).

Monostotic fibrous dysplasia, which usually involves the femur or the ribs, is the most common form (74% of cases) (1). It usually manifests at the age of 20-30 years. Polyostotic fibrous dysplasia, which accounts for about 13% of cases, is usually more severe and has an onset in early childhood. It often affects, among other things, the facial part of the skull. Craniomaxillofacial fibrous dysplasia is classified by the WHO as a separate disease, accounting for about 13% of cases (10). Symptoms develop before the age of 5 years in 90% of patients.

Treatment needs of patients with fibrous dysplasia depend on the stage and severity of disease, as well as functional and aesthetic disorders associated with the disease. In the absence of indications resulting from the above mentioned disorders, potential surgical treatment should be delayed until the patient reaches maturity, when progression of lesions usually stops (6). Pharmacological treatment may involve bisphosphonates, calcitonin, calcium and vitamin D. Antibiotics (lincomycin, clindamycin) are used during exacerbations (2, 11-13). Previous use of radiation therapy is debatable as there is a 400 times increased risk of malignant transformation of dysplastic lesions (14).

At the same time, patients need comprehensive, multidisciplinary dental treatment ranging from the management of dental caries and periodontal diseases, through orthodontic treatment of accompanying malocclusions, to maxillofacial surgery and prosthetic rehabilitation. Dysplastic lesions may involve the maxillary alveolar process and the alveolar part of the mandible, affecting dental growth and arrangement, as well as causing malocclusions. Malocclusions, dental crowding or interdental gaps are the most common dental disorders in such cases (15). Indications for certain stomatognathic rehabilitation approaches should be tailored individually for each clinical case as the presentation varies among patients, depending on location, stage and course of the disease. Prosthetic treatment is often difficult due to recurrent exacerbations.

In the presented clinical case, not only the bone tissue, but also the number, quality, formation and location of tooth buds were affected, leading to severe morphological and functional alterations. The development of the stomatognathic system was also certainly affected by surgical procedures and the lack of stimulation of the growth of the maxillary alveolar process and the alveolar portion of the mandible, which occurs during tooth eruption. These factors were the reason for severe oligodontia, underdeveloped alveolar processes, and deformed tissues in the prosthetic field. Growth hormone therapy could also account for the irregular growth of the facial skull; however, as pointed out by van Erum et al., this depends on patient's age, doses used and treatment duration (16). These changes also resulted in reduced height of occlusion, changes in facial features and chewing dysfunction (fig. 8, 9). The influence of these factors on the child's appearance, self-esteem, interpersonal contacts or position in the peer group was not without significance.

There are only few reports describing prosthetic treatment in patients with craniomaxillofacial fibrous dysplasia. The authors focus mainly on surgical or orthodontic

aspects of stomatognathic rehabilitation. There are also scarce reports and missing long-term data on implant treatment involving placement of intraosseous implants in a dysplastic bone. However, such attempts have been made, and successful osseointegration of implants was achieved (17).

Prosthetic treatment in patients at developmental age requires regular check-ups and frequent corrections or replacement of restorations, depending on the clinical situation. In the case of patients with dysplastic lesions, especially during active phase of the disease when the progression of lesions is difficult to predict, prosthetic rehabilitation requires individual, multidisciplinary approach and close collaboration with the patient and their parents/legal guardians. There were several reasons for treatment failure in the described case: non-compliance with follow-up due to frequent hospital stays, difficult adaptation to dentures, unsatisfactory retention of restorations due to difficult conditions in the prosthetic field, and poor cooperation with the parent. In the case of treating adolescent patients with severe mandibular oligodontia or anodontia and objective difficulties using removable dentures, optional implant treatment involving the introduction of intraosseous implants between mental foramina is considered. However, the therapy was postponed until completing the diagnosis of dysplastic lesions, potential surgical treatment and osseous remodelling in the anterior portion of the mandible in the described case due to the presence of dysplastic lesions and factors contributing to the difficulty in adapting to restorations. It should be also emphasised that the therapeutic success, especially in the case of multistage interdisciplinary treatment of adolescent patients largely depends on the cooperation with the patient and their parents/legal guardians and understanding the limitations in the therapeutic management in such difficult clinical cases.

Conflict of interest

None

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