

CASE REPORT OLGU SUNUMU

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# Monostotic Fibrous Dysplasia of the Condyle and Ascending Ramus in a 7-Year-Old Child Treated with Conservative Surgery: 8-Year Follow-Up

## Konservatif Cerrahi ile Tedavi Edilen 7 Yaşındaki Bir Çocukta Kondil ve Yükselen Ramusun Monostotik Fibröz Displazisi: 8 Yıllık Takip

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**ABSTRACT** Fibrous dysplasia, a genetic, developmental bone disorder disrupting osteogenesis, results in excessive fibrous tissue replacing normal bone. It can manifest as monostotic or polyostotic forms, with about 30% of monostotic cases observed in the maxilla and mandible, predominantly unilateral in the posterior region. Common in adolescence, stability often occurs in adulthood. Clinical manifestations include swelling, facial asymmetry, pain, or numbness. Treatment options include conservative surgical approaches, radical interventions, and bisphosphonate-based medical therapies. This report presents a case of monostotic fibrous dysplasia in a 7-year-old female, demonstrating a one-year history of swelling in the posterior right region of the mandible. A conservative surgical approach was employed, resulting in no recurrence during an 8-year follow-up, with significant improvements in bone reconstruction. Early implementation of a conservative approach instead of radical and continuous follow-ups is vital for favorable outcomes and minimizing recurrence risk in young patients with monostotic fibrous dysplasia.

**Keywords:** Monostotic fibrous dysplasia; mandible; condyle; child

**ÖZET** Osteogenezi bozan genetik, gelişimsel bir kemik bozukluğu olan fibröz displazi, normal kemiğin yerini aşırı fibröz dokunun almasıyla sonuçlanır. Monostotik veya polioostotik formlarda ortaya çıkabilir; monostotik vakaların yaklaşık %30'u maksilla ve mandibulada, ağırlıklı olarak posterior bölgede tek taraflı olarak görülür. Ergenlik döneminde yaygındır, stabilite genellikle erişkinlikte ortaya çıkar. Klinik bulgular arasında şişlik, yüz asimetrisi, ağrı veya uyuşukluk yer alır. Tedavi seçenekleri arasında konservatif cerrahi yaklaşımlar, radikal girişimler ve bifosfonat bazlı medikal tedaviler yer almaktadır. Bu yazıda, mandibulanın sağ arka bölgesinde bir yıldır şişlik olan 7 yaşında bir kız hastada monostotik fibröz displazi olgusu sunulmuştur. Konservatif bir cerrahi yaklaşım uygulanmış, 8 yıllık takip süresince nüks görülmemiş ve kemik rekonstrüksiyonunda önemli gelişmeler kaydedilmiştir. Monostotik fibröz displazili genç hastalarda radikal yerine konservatif yaklaşımın erken uygulanması ve sürekli takipler, olumlu sonuçlar ve nüks riskinin en aza indirilmesi için hayati önem taşımaktadır.

**Anahtar Kelimeler:** Monostotik fibröz displazi; mandibula; kondil; çocuk

Fibrous dysplasia (FD) represents a noncancerous process within the bone, marked by the substitution of marrow bone with poorly mineralized fibrous connective tissue.<sup>1-3</sup> The origin of this condition stems from a genetic anomaly in the stimulating G protein (GSα), inducing an imbalance in the activities of both bone-forming and bone-resorbing cells.

Overactive GSα triggers enhanced activity in precursor cells of bone formation and cells that break down bone, prompted by heightened production of interleukin-6 from supportive cells.<sup>4-6</sup> As per the bone tumor classification by the World Health Organization, FD falls into the category of tumors without a defined neoplastic character. Hormonal irregularities,

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notably early puberty in McCune-Albright syndrome (MAS) or intramuscular myxomas in Mazabraud's syndrome (MS) are linked to this condition.<sup>1,3</sup>

Typically emerging during the initial or subsequent ten years of life, FD predominantly affects females, with a ratio of 2:1 compared to males, often exhibiting no symptoms and minimal advancement.<sup>1,3</sup> The disease can appear either as a solitary form, affecting a single bone [monostotic fibrous dysplasia or (MFD)], or as a multifocal form, impacting multiple bones [polyostotic fibrous dysplasia or (PFD)]. MFD, which is about ten times more prevalent than PFD, generally tends to occur on one side.<sup>2,3</sup> The absence of steady bone matrix formation can lead to diverse presentations, such as facial distortion and lack of symmetry.<sup>4-6</sup>

In this case report, we present a case of MFD in a 7-year-old female patient who complained of gradually increasing swelling on the posterior right region of the mandible persisting for one year. The diagnosis of MFD was confirmed following an extensive assessment that integrated clinical, radiographic, and histopathological characteristics. A conservative surgical approach addressed the patient's condition, involving surgical excision and recontouring of excessive bone to restore facial symmetry. After an 8-year follow-up, the case exhibited no recurrence, with significant improvements observed in the reconstruction of the condylar and coronoid processes and the incisura mandible.

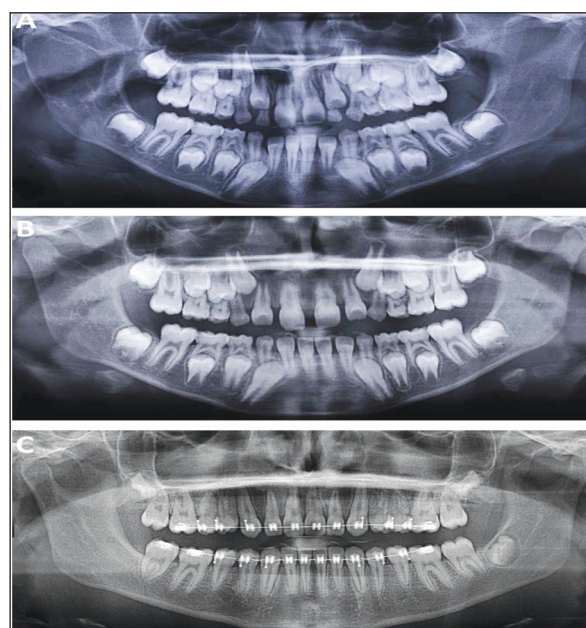
## CASE REPORT

A 7-year-old female was admitted to our department, accompanied by her parents, with a complaint of swelling and asymmetry on the right side of her mandible. Before presenting to our department, the patient underwent evaluations in several medical centers, where aggressive surgical interventions, including total ramus resection, were recommended based on suspicion of malignancy. However, due to the family's concern regarding the potential risks associated with such extensive surgeries, they pursued alternative opinions and ultimately sought consultation at our institution. The patient history indicated a gradual, unhurried enlargement of the swelling over the

course of the last year, with no accompanying pain.

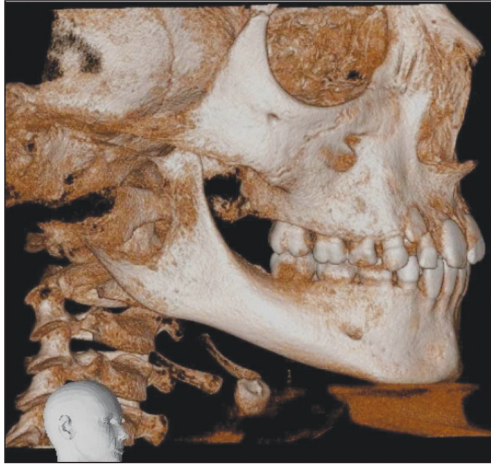
Upon extraoral examination, mild facial asymmetry was noted, concomitant with a noticeable swelling on the right side of the face. During mouth movement, the patient exhibited limited mouth opening and a deviation of the chin to the right. There were no pigmentation abnormalities on the facial skin, and no enlargement of submandibular lymph nodes was observed. Palpation indicated a firm and fixed swelling without mucosal inflammation or paresthesia. The intraoral examination did not reveal significant changes. A panoramic radiograph displayed a loss of trabecular structure with a radiopaque "ground-glass" appearance encompassing the right mandibular ramus and condyle (Figure 1). Cone-beam computed tomography (CBCT) images confirmed the lesion's extent, which affected the right mandibular ramus cortical bone and condyle, with only partially present the right ramus coronoid process (Figure 2).

Written informed consent for further investigation and permission to use their photographs and clinical data were taken. Based on clinical and radiological assessments, an initial diagnosis of MFD was made, with differential diagnoses including ossifying fibroma, MAS, and MS. The lesion was sur-



**FIGURE 1:** Orthopantomographic images of the patient; **A)** Preoperative status; **B)** First-year postoperative follow-up; **C)** Eighth-year follow-up.





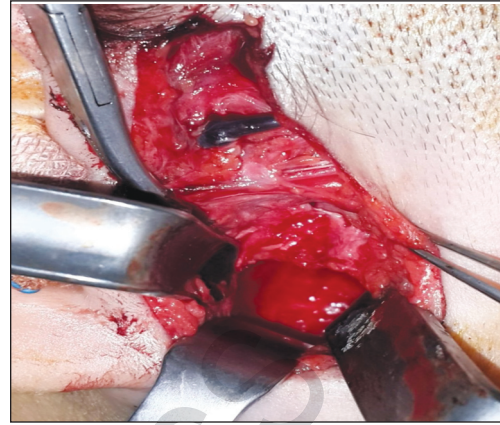
**FIGURE 2:** The three-dimensional model obtained from cone-beam computed tomography images showing the preoperative status of the patient.

gically excised using a conservative extraoral approach under general anesthesia, which impacted the region's remodeling (Figure 3).

Macroscopic evaluation of the excised bone revealed soft and spongy aspects (Figure 4). The biopsy specimens, consisting of pink-white elastic tissues ranging in size from 0.6x0.3x0.3 to 3x2.5x1.5 cm and an additional 1 cm of bone tissues, were subjected to routine histological processing. Hematoxylin and eosin staining demonstrated irregularly shaped immature bone trabeculae within a fibroblastic and vascularized stroma.

The microscopic examination further revealed a stroma composed of fibroblastic-fibrocytic spindle cells interspersed with irregular immature/felt bone trabeculae and delicate vascular structures. The stromal spindle cells exhibited no cytological abnormalities or mitotic activity, and the immature bone trabeculae lacked osteoblastic rimming. Notably, microscopic sections of the bone tissues exhibited clear continuity with the surrounding cortical bone tissue. These histological features were consistent with a diagnosis of FD (Figure 5).

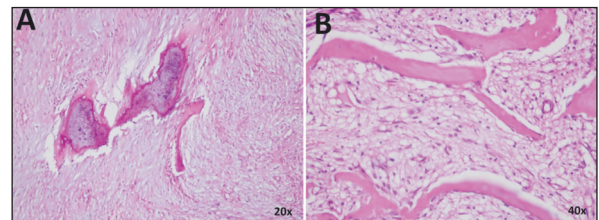
After the surgery, the patient underwent annual follow-ups involving clinical assessments and radiological examinations, including CBCT imaging performed six years post-operation. The CBCT images displayed significant improvements in the reconstruction of the condylar and coronoid processes and the mandibular incisura (Figure 6). No complications



**FIGURE 3:** Intraoperative image showing the surgical field and gap in the area after removal of the lesion by extraoral approach.



**FIGURE 4:** Image of the lesion removed from the area.



**FIGURE 5:** Irregularly shaped immature bone trabeculae without osteoblastic rim in fibrotic stroma composed of spindle cells. **A):** Hematoxylin staining, x20 **B):** Hematoxylin staining, x40.



**FIGURE 6:** The three-dimensional model was obtained from follow-up cone-beam computed tomography at year 8. The image shows good remodeling in the lesion area and condyle and coronoid processes.

were encountered during the follow-up period, and the patient reported satisfaction with both aesthetic and functional outcomes. Eight years later, further clinical and radiological evaluations demonstrated no evidence of lesion recurrence, indicating lesion stabilization.

## DISCUSSION

FD constitutes 7% of benign bone tumors, with facial bones involved in 10%-27% of MFD cases.<sup>1-3</sup> Studies present differing views on FD distribution between the maxilla and mandible. In different studies, there is different information that MFD is seen more frequently in favor of the maxilla or mandible.<sup>7-10</sup> FD's differential diagnosis encompasses bone disorders such as bone cysts, non-ossifying fibroma, osseous-fibrous dysplasia, adamantinoma, intramedullary osteosarcoma, Paget's disease, MAS, and MS.<sup>1,8,10</sup>

Panoramic radiographs offer insights, yet CBCT is the gold standard, yielding 3D models and comprehensive lesion data. CBCT images are crucial for accurate lesion delineation.<sup>7</sup> In this case, CBCT was selected as the preferred modality for assessing the condylar damage, ascertaining the necessity for grafting, and elucidating the lesion's relationship with anatomical structures in greater detail.

Treatment options comprise conservative surgery, radical excision, and bisphosphonate therapy.<sup>8,9,11-14</sup> Conservative surgery removes poorly mineralized bone, aiding histological confirmation. The surgical approach considers patient age, as FD can recur with growth maturation.<sup>3,13</sup> Kruse et al. proposes successful modeling osteotomy for MFD.<sup>9</sup> Valentini et al. reports MFD (76%), PFD (22%), and MAS (2%) in 95 craniomaxillofacial FD cases, with 68 receiving surgeries.<sup>13</sup> On the other hand, some avoid surgery despite its preventive potential, opting for bisphosphonates to alleviate bone pain. Bisphosphonates can improve clinical status, but long-term use poses an osteonecrosis risk.<sup>12,14,15</sup> In the present case, a young patient's diagnosis relies on clinical, radiological, and histological data. Given the age, a conservative surgical approach is prudent, avoiding

facial growth disturbance. The drawbacks associated with the extraoral approach include its heightened surgical invasiveness, necessitating a greater level of expertise compared to alternative treatment methods. Additionally, the extraoral approach has the potential to induce variable degrees of scarring in the treated area, thereby giving rise to aesthetic concerns. After eight years, the case exhibited no recurrence, with notable reconstruction of mandibular structures. Clinical stability and aesthetic satisfaction persist.

The present case report emphasizes the significance of a comprehensive approach to managing MFD, particularly in pediatric patients. The diagnostic complexities inherent in this condition warrant thorough evaluation and collaboration among various medical specialties to avoid misdiagnosis and prevent unnecessary aggressive surgeries. Our experience highlights the importance of considering conservative treatment options, especially in young patients, to minimize the potential risks of radical surgical interventions. Our approach serves as an exemplary alternative for similar cases. It underscores the value of patient-centered care in achieving optimal outcomes.

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### Conflict of Interest

*No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.*

### Authorship Contributions

**Idea/Concept:** Behçet Erol; **Design:** Sercan Küçükkurt; **Control/Supervision:** Behçet Erol; **Data Collection and/or Processing:** Sercan Küçükkurt; **Analysis and/or Interpretation:** Behçet Erol; **Literature Review:** Sercan Küçükkurt; **Writing the Article:** Sercan Küçükkurt; **Critical Review:** Behçet Erol; **References and Fundings:** Behçet Erol, Sercan Küçükkurt; **Materials:** Behçet Erol, Sercan Küçükkurt.

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