

Fibrous Dysplasia: A Case Report

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Abstract

Fibrous dysplasia is a rare benign bone tumor. It has the potential to demonstrate malignant transformation. Fibrous dysplasia occurs when normal bone tissue is replaced by fibrous tissue. The most common locations in the maxillofacial region are the maxilla and mandible. Fibrous dysplasia is divided into monostotic and polyostotic forms. If involvement is observed in more than one bone, it is called polyostotic fibrous dysplasia, if involvement is observed in a single bone, it is called monostotic fibrous dysplasia. Monostotic type fibrous dysplasia accounts for 75% of cases. There are many syndromes in which polyostotic fibrous dysplasia is seen. The best known of these are McCune Albright syndrome, Jaffe-Lichtenstein syndrome and Mazabraud syndrome.

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Introduction

Fibrous dysplasia was first defined by Lichtenstein in 1938 as polyostotic fibrous dysplasia, but this term was changed to fibrous dysplasia because skeletal changes were divided into monostotic and polyostotic forms (1). Fibrous dysplasia is a sporadic disease of genetic origin that occurs in the bone. It occurs as a result of mutation in the gene code in the α subunit of the Gs protein of the GNAS complex locus on chromosome 20q13 (2,3).

Fibroosseous lesions describe processes characterized by the replacement of normal bone by fibrous tissue containing a newly formed mineralized product. Fibro-osseous lesions are fibrous dysplasia, cemento-osseous dysplasia, and ossified fibroma. Fibrous dysplasia is characterized by tumor tissue that develops from the replacement of normal bone tissue with a mixture of irregular trabeculae and cellular fibrous connective tissue (10).

Fibrous dysplasia is a rare skeletal disorder that can cause bone fractures, deformities, pain, and impairment in function. In fibrous dysplasia, bone is replaced by fibrous and osseous tissue, which are structurally weaker. Although it is

localized and benign, it can show malignant transformation (4,5).

Fibrous dysplasia is divided into two types, monostotic and polyostotic. In the literature, 70% of fibrous dysplasia cases are monostotic. In a large multicenter clinicopathological study conducted in Europe, it was determined that the most common site of involvement in patients with monostotic fibrous dysplasia disease was the femur. Other common sites of involvement are ribs and craniofacial bones. Polyostotic type fibrous dysplasia is characterized by the presence of multiple foci in many bones (6,7).

McCune-Albright syndrome (MAS), which consists of the triad of fibrous dysplasia, cafe au lait spots, and endocrine disorders, was described in 1937 by Donovan McCune and Fuller Albright. The most common endocrine disorder is related to sex hormones. While surgical treatment was used for treatment in the past, anti-hormonal medical treatments are preferred today. Bone lesions, cafe-au-lait spots, and affected gonads are typically on the same side of the body. Endocrine disorders can be any one of hypothyroidism, adrenal disorders, diabetes, hypopituitarism and hypercalcemia (8,9).

Magnetic resonance imaging, conventional x-rays, computed tomography, histopathology and

scintigraphy may be preferred in the diagnosis of fibrous dysplasia. Computed tomography is very important for the diagnosis and treatment of fibrous dysplasia. In the literature, it has been stated that it is the most effective method used in the diagnosis of fibrous dysplasia. Radiological appearance changes according to the development of the bone matrix. While radiolucent appearance is dominant in the early stage of the lesion, radiopaque appearance is more common in advanced stages. The radiological appearance of the fibrous dysplasia lesion is expressed as 'ground glass' or 'orange peel' (3,5,11,12).

In the early stages of fibrous dysplasia, the lytic zone can be well or poorly circumscribed, unilocular or multilocular. The 'ground glass' appearance seen on the radiograph is one of the most important criteria for diagnosis. In the later stages of the lesion, the lamina dura of the teeth become indistinct in the periapical films taken from the affected area. It has been reported that the canal can change its location in the mandibular canal region (11,13).

In fibrous dysplasia cases, asymptomatic cases are followed or conservative treatment is preferred. Due to the possibility of recurrence, long-term follow-up of the treated cases is required (14).

Case report

A 21-year-old female patient was admitted to the Department of Oral and Maxillofacial Surgery of the Faculty of Dentistry of Harran University with complaints of swelling in the right maxillary region and facial asymmetry. In the anamnesis, it was learned that the patient had previously undergone surgery due to a case of fibrous dysplasia in the same region. It was determined that the patient had been complaining of swelling in the same area for the past year and had difficulty breathing due to this swelling. No systemic disease was detected in the patient.

Extraoral examination revealed an expansive area in the right maxillary region of the patient, extending to the inferior edge of the orbit, causing facial asymmetry. Intraoral examination revealed a painless swelling resembling a bony prominence in the right maxilla. (Figure 1)

Orthopantomograph film was taken for radiological examination. In the examination, bone expansion and ground glass appearance were observed in the right maxillary region. (Figure 2)



Figure 1. Preoperative intraoral view of the case.



Figure 2. Panoramic radiograph of the lesion in the right maxilla

Computed tomography sections were examined in order to more clearly monitor the borders of the lesion. In computed tomography sections, it was observed that the lesion completely filled the inside of the maxillary sinus and caused a deviation in the nasal septum. (Figure 3) Expansion was detected in the right maxillary palatal and vestibule regions. (Figure 4) It was learned that the patient had difficulty in breathing due to the expansion caused by the lesion. In the three-dimensional film of the patient, it was seen that the lesion included the right posterior region of the maxilla. (Figure 5)

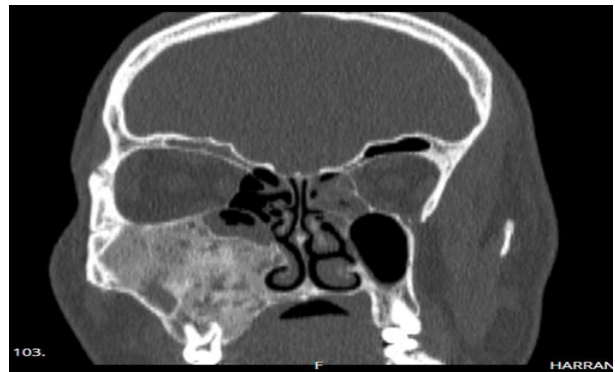


Figure 3. Coronal section computed tomography image of the lesion located in the posterior region of the maxilla.

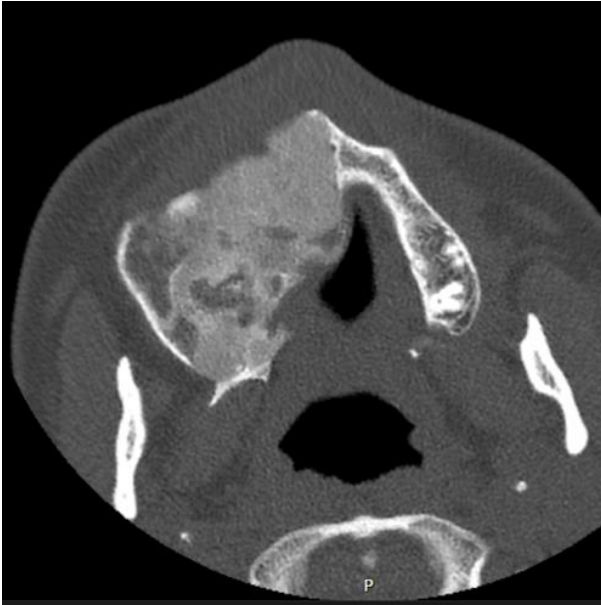


Figure 4. Axial section computed tomography image of the lesion located in the maxilla posterior region.



Figure 6. Parts removed from the lesion during the operation.



Figure 5. Computed tomography 3D view of the case.

As a result of clinical and radiological examinations, it was decided to carry out a biopsy from the lesion suggestive of fibrous dysplasia. It was deemed appropriate to operate the patient with general anesthesia. After nasal intubation, vertical incisions were added to expose the lesion borders in the right maxillary posterior region and the flap was removed. Bone contour adjustment was made in order to eliminate the cosmetic problem that bothered the patient in the vestibular region. The flap was sutured with 3-0 silk suture. Pieces taken from the lesion were sent for histopathological evaluation. (Figure 6)



Figure 7. Postoperative radiological image of the patient.

Discussion

Fibrous dysplasia is a non-neoplastic developmental hamartomatous disease of bone characterized by a mixture of fibrous and osseous elements in the region. It is a rare disease with an incidence of 1:4000-1:10,000. It accounts for approximately 2.5% of all bone lesions and approximately 7% of all benign bone tumors. It has three subtypes: polyostotic, monostotic, and craniofacial. Craniofacial fibrous dysplasia describes the lesion that occurs at the junctions of bones in the craniofacial skeleton. Most cases of craniofacial fibrous dysplasia cannot be classified as monostotic because it may involve many bones in the cranium. They are not polyostotic because the outer cortex of the craniofacial complex is preserved. Craniofacial fibrous dysplasia tends to be more common in women. Since it coincides with the maturation time of the bones of the patients, its incidence is high in the thirties (15, 16, 17).

Fibrous dysplasia is generally seen with equal frequency in both sexes. The lesion, which is more common in the maxilla than in the mandible, usually grows slowly. Displacement of teeth, facial asymmetry and malocclusion are common findings of fibrous dysplasia. The lesion may cause expansion in the vestibule region. The mucosa is usually normal in color. Depending on the lesion stages, it may have various radiographic appearances. While there is a multilocular or unilocular radiolucent appearance in the early stage, radiopaque foci are added on the radiolucent area in advanced stages. As the lesion matures, the radiopaque area becomes dominant. Radiological definitions used for fibrous dysplasia; fingerprints, orange peel and frosted glass (18, 19). Petrikowski et al. reported that the only lesion that moves the mandibular canal superiorly is fibrous dysplasia. This finding is used to distinguish fibrous dysplasia from other lesions (20).

Differential diagnosis of fibrous dysplasia; It is performed with Paget's disease, giant cell tumor, Brown tumor seen in hyperthyroidism, ossifying fibroma and central giant cell granuloma. At the same time, the radiological appearance of chronic osteomyelitis and the radiological appearance of fibrous dysplasia are similar. Purulent discharge, pain and tenderness of inflammation are seen in osteomyelitis but not in fibrous dysplasia. Fibrous dysplasia is usually characterized by painless, slow-growing swelling. These features distinguish it from malignant tumors (18,19).

McCune-Albright syndrome is a subform of fibrous dysplasia characterized by endocrine disorders and brown patches on the skin described as café-au-lait spots. Findings such as hyperthyroidism, acromegaly and Cushing's syndrome can also be seen in this syndrome. It is more common in the female gender. Stains are usually seen on the dorsum, sacrum and hips (11,12).

Treatment of fibrous dysplasia is mostly surgical. The most effective treatment is total resection of the bone involved by the lesion. However, this leads to serious cosmetic and functional problems. It can also cause serious

complications in the future. Therefore, conservative surgical treatment is preferred. With conservative treatment, it is aimed to correct the asymmetry and cosmetic problem caused by the lesion. Patients with fibrous dysplasia need long-term follow-up because of the risk of malignant transformation (21, 22).

Medical treatment is not often preferred in the treatment of fibrous dysplasia. Recognition of the pathogenesis of fibrous dysplasia brought to mind the use of bisphosphonates in the treatment of this disease. Agents with bisphosphonate active ingredients inhibit the osteoclastic activity of the bone and prevent the destruction of the bone. This stabilizes the bone and reduces the patient's pain. At the same time, another medical agent to be used for the treatment of fibrous dysplasia is pamidronate. This drug is also a second generation agent that inhibits bone resorption. Since serum calcium is low in these patients, vitamin D and calcium supplementation are also recommended (16).

Because of the possibility of malignant transformation and recurrence, it should be followed for a long time. Fibrous dysplasia is a rare lesion. The radiological and clinical features differ in cases (1).

In the case we presented, fibrous dysplasia was located in the posterior region of the right maxilla. A ground glass appearance was observed radiologically in the relevant region. Clinically, it caused swelling in the vestibule and caused asymmetry. With conservative surgical treatment, the bone contour was corrected and the cosmetic problem was resolved. The patient is followed in our clinic.

Conclusion

As a result; fibrous dysplasia is a disease diagnosed by radiological, clinical and histopathological correlations. It causes asymmetry and cosmetic problems in the craniofacial region. Ground glass appearance is characteristic in radiology. Its treatment is total resection or conservative surgical approach. Expected results from medical treatments could not be obtained. Fibrous dysplasia because of the risk of transformation into malignant tumors such as osteosarcoma, it should be under continuous follow-up.

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