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# Florid cemento osseous dysplasia and dentygerous cyst in a patient with apert syndrome: A case report

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## ABSTRACT

**Florid cemento osseous dysplasia and dentygerous cyst in a patient with apert syndrome: A case report**

Apert syndrome is a rare congenital malformation characterized by craniocinocytosis, craniofacial anomalies and symmetric syndactyly of the feet and hands. Oral manifestations of Apert syndrome usually represents bifid uvula, malposition of the teeth, severe open bite, tooth decay and periodontal diseases. Fluorid cemento-osseous dysplasia is usually asymptomatic slow-growing non-neoplastic fibro-osseous lesions. Lesions are detected by routine radiographic examination. The aim of this case report is to present dentigerous cyst and florid cemento-osseous displasia in a patient with Apert syndrome. A 38-year-old female patient with a history of Apert syndrome referred to Marmara University, Faculty of Dentistry, Clinic of Oral and Dentomaxillofacial Radiology due to pain and swelling. On panoramic radiography, unilocular, hyperdense lesion with regular borders was observed in the anterior region of the mandible. Cone-beam computed tomography (CBCT) was performed for further examination of the lesion and perforation of buccal bone cortex was seen. Additionally, a regular monolocular hypodense lesion was observed in the anterior region of maxilla. A biopsy was performed to examine the lesion histopathologically. Histopathologic examination was performed to evaluate the lesions and the lesion in the mandible was diagnosed as cemento-osseous dysplasia. Because the lesion was multifocal, it was considered and compatible with fluoride cemento-osseous dysplasia. The lesion in the maxilla was diagnosed as dentigerous cyst on histopathologic examination. The diagnosis of fluoride cemento-osseous dysplasia is established by definite radiological and histopathological evaluation. In asymptomatic cases of fluorid cemento-osseous dysplasia, treatment is not required but patients should be followed up regularly. Practitioners should take into consideration the oral and dental findings in patients with Apert syndrome which rarely appear.

## KEYWORDS

**Apert Syndrome, florid cementoosseous dysplasia, dentigerous cyst**

## ÖZ

**Apert sendromlu bir hastada florid semento osseoz displazi ve dentigeröz kist: Olgu sunumu**

Apert sendromu kraniyosinositoz, kraniyofasiyel anomaliler ve ellerin ayakların simetrik sindaktili ile karakterize nadir görülen bir konjenital malformasyondur. Apert sendromunda oral belirtiler genellikle bifid uvula, dişlerin malpozisyonu, şiddetli open-bite, diş çürüğü ve periodontal hastalıkları olarak gözükür. Florid semento-osseoz displazi genellikle asemptomatik yavaş büyüyen non-neoplastik fibro-osseoz bir lezyondur. Lezyonlar rutin radyografik muayene ile tespit edilirler. Lokalizasyonları dişli bölgelerdedir, dağılımları simetriktir. Bu vaka sunumunun amacı Apert sendromlu hastada florid semento-osseoz displazi ile birlikte dentigeröz kist olgusunu sunmaktır. Anamnezinde Apert sendromlu olduğu öğrenilen 38 yaşındaki kadın hasta ağrı ve şişlik sebebiyle Marmara Üniversitesi Diş Hekimliği Fakültesi Oral Diagnoz ve Radyoloji Kliniğine başvurmuştur. Hastadan alınan panaromik radyografide mandibula anterior bölgede uniloküler, sınırları düzgün, hiperdens yapıda lezyon izlenmiştir. Daha detaylı inceleme için alınan konik ışınlı bilgisayarlı tomografi (KIBT) görüntülerinde lezyonun bukkal kemik korteksinde perforasyona neden olduğu gözlenmiştir. Ayrıca, maksillada horizontal konumda gömük olan 23 numaralı diş kuronunu çevreleyen sınırları düzenli monoloküler hipodens lezyon izlenmiştir. Histopatolojik değerlendirme sonucu mandibuladaki lezyona semento-osseoz displazi tanısı konulmuştur. Olguda izlenen lezyonlar multifokal olduğu için florid semento-oseoz displazi ile uyumludur. Maksilladaki lezyonun ise histopatolojik değerlendirmesi sonucu dentigeröz kist tanısı konulmuştur. Florid semento-osseoz displazinin tanısı doğru klinik ve radyografik değerlendirme ile yapılır. Florid semento-osseoz displazinin asemptomatik olgularında tedavi gerekmez ancak hastanın düzenli takibi yapılmalıdır. Ancak bu olgu semptomatik olduğu için cerrahi müdahele gerekmiştir. Diş hekimleri nadir görülen Apert sendromlu hastalarda dental bulguları dikkate almalıdır.

## ANAHTAR KELİMELER

**Apert Sendromu, florid sementooseöz displazi, dentijeröz kist**

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## INTRODUCTION

Apert syndrome (AS), also known as acrocephalosyndactyly, is one of the rarest and most severe cranio-synostosis syndromes, accounting for about 4.5% of all craniosynostosis cases.[1] AS was first clinically described by Baumgartner in 1842 and by Wheaton in 1894; later, it was reviewed extensively by Eugene Apert, a French Pediatrician, who published a series of nine cases in 1906.[2] Prevalence of the syndrome has been estimated to be between 1/65 000 and 1/200 000 newborns, without predilection by gender.[2,3] AS has an autosomal dominant pattern of inheritance, associated with advanced paternal age, maternal infections, maternal drug consumption, and cranial inflammatory processes.[4] Investigations reported in the literature on AS are usually related to genetics or surgical management, with little emphasis on the oral aspects. Early fusion of the skull bones affects the development of the brain, which can disrupt intellectual development. Cognitive abilities in people with AS range from normal to mild or moderate intellectual disability. Individuals with Apert syndrome have webbed or fused fingers and toes. The severity of the fusion varies; at a minimum, three digits on each hand and foot are fused together. In the most severe cases, all of the fingers and toes are fused. Less commonly, people with this condition may have extra fingers or toes (polydactyly). Additional signs and symptoms of AS can include hearing loss, unusually heavy sweating (hyperhidrosis), oily skin with severe acne, patches of missing hair in the eyebrows, fusion of spinal bones in the neck (cervical vertebrae), and recurrent ear infections that may be associated with an opening in the roof of the mouth (a cleft palate). Oral and dental characteristics of AS were documented as; skeletal anterior open bite, soft palate cleft, bifid uvula, narrower dimensions of dental arches with severe crowding, bilateral swellings of the palatine processes, bilateral posterior crossbite, gingivo/periodontal diseases, hypotonic lip, impaired speech, tooth agenesis, supernumerary teeth, dental fusion, dental delay in maturation and eruption. [2,3,5-7] Florid cemento-osseous dysplasia (FCOD) is more commonly seen in middle-aged black women, although it also may occur in Caucasians and Asians. In some cases, a familial trend can be observed .[8, 9] The process may be totally asymptomatic and, in such cases, the lesion is detected when radiographs are taken for some other purposes. Symptoms such as dull pain or drainage are almost always associated with exposure of sclerotic calcified masses in the oral cavity. This may occur as the result of progressive alveolar atrophy under a denture or after extraction of teeth in the affected area.[9] This aim of this case report is to present a patient with Apert Syndrome diagnosed with florid cemento-osseous dysplasia and dentigerous cyst on the basis of clinical, radiographic and histological findings.

## CASE REPORT

A 38-year-old female patient with a history of Apert Syndrome referred to Marmara University, Faculty of Dentistry, Clinic of Oral and Dentomaxillofacial Radiology with chief complaint of pain on extra-oral examination, swelling was noted in anterior mandible. Panoramic radiograph was obtained and unilocular, sclerotic mass with regular radiolucent borders was observed in the anterior region of the mandible. Bilateral styloid process elongation was also noted. Cone-beam computed tomography (CBCT) was performed for further examination of the lesion. Coronal, sagittal and axial cross-sectional images demonstrated hyperdense lesion, with 21x18x14 mm in size and regular borders in the anterior region of mandible. The lesion formed expansion and perforation on the buccal

cortex. Another hyperdense lesion, 14x18x19 mm in size, was observed on base of the right nasal cavity with regular borders. The lesion caused perforation in the buccal bone cortex. Additionally, a regular monolocular hypodense lesion with unerupted tooth no 23 was observed in the anterior region of maxilla (Figure 1). Based on the patient's clinical and CBCT features, preliminary diagnosis was florid cemento-osseous dysplasia with differential diagnosis of complex odontoma/cementoblastoma. The lesion in the maxilla anterior region was thought to be a dentigerous cyst. Biopsy was planned (Figure 2) and histopathological examination revealed the formations of dense sclerotic calcified cementum-like masses. Sections from soft tissue showed fibrous to fibrocellular stroma with dilated blood vessel and chronic inflammatory cell infiltration. The periphery of the lesion showed globular or ovoid structures of cementoid appearance involved by thin fibrous tissue (Figure 3).

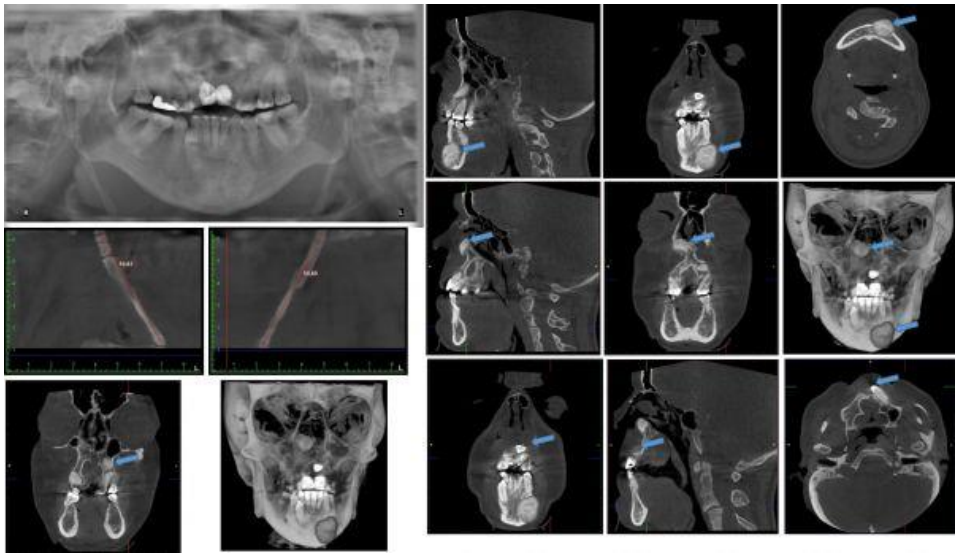

**Figure 1.** Hyperdens lesions in the maxilla and mandibula and cystic formation in the anterior region of the maxilla with bilateral sthyloid process elongation on panoramic radiograph and CBCT images

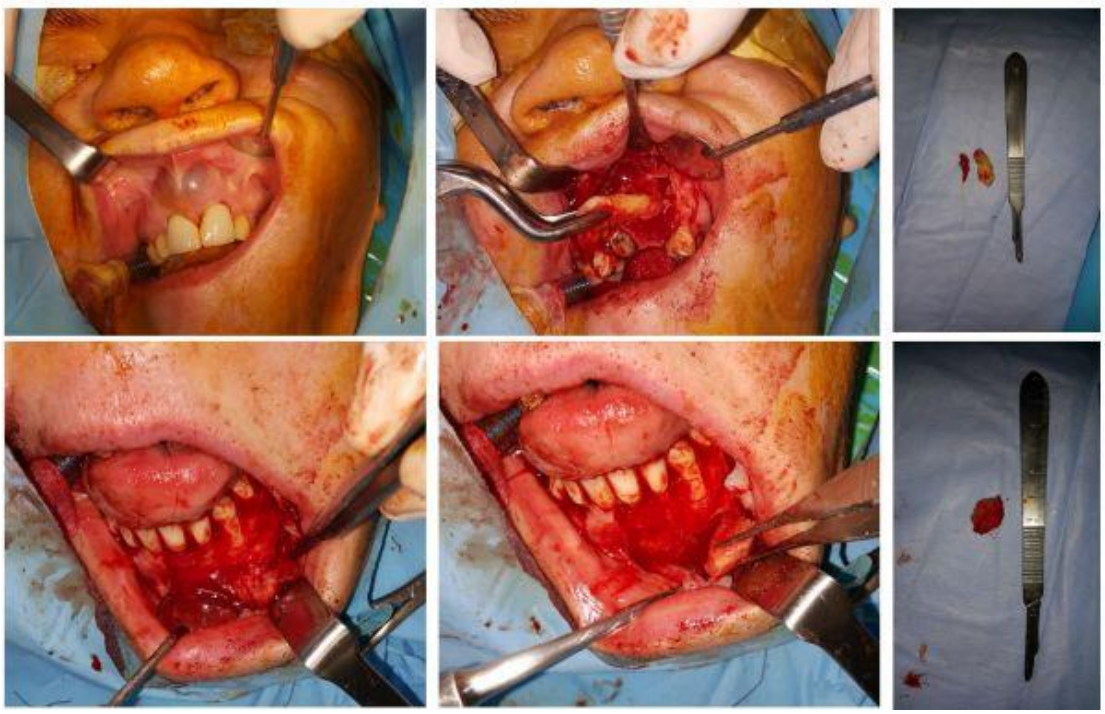

**Figure 2.** Biopsy was performed from the monitored lesions

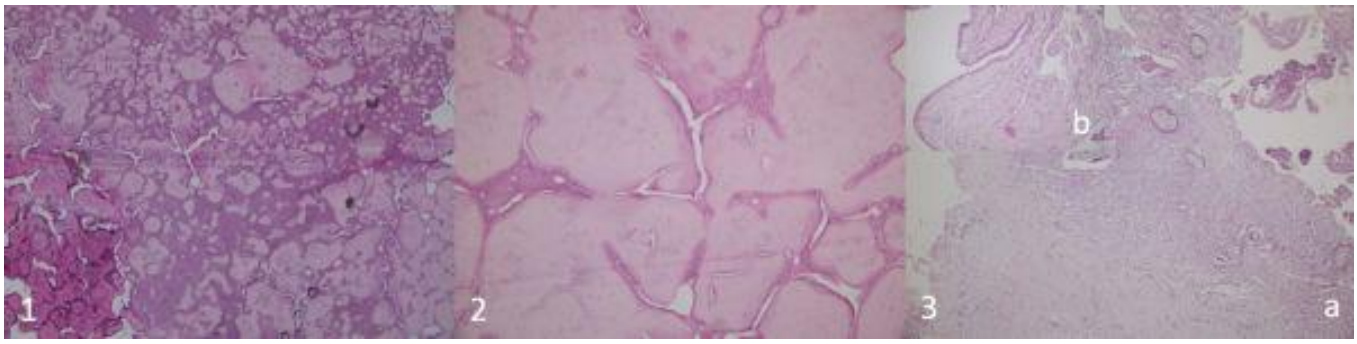


**Figure 3.** 1,2, 3a )Inside the cell-rich lesion composed of spindle cells, mostly round, ovoid structure cementite-osseous calcifications, some of which resemble cement (H&E x40, x100, x100 respectively) 3b) The inner surface of the fibrous wall is covered by a thin epithelium composed of several rows of cubic cells H&EX200

## DISCUSSION

Although FCODs have been reported extensively in the literature, to our knowledge this is the first case representing a unique combination of FCOD with features of Apert Syndrome. FCOD may be familial with an autosomal dominant inheritance pattern, but there are only a few examples in the literature in which the familial pattern has been confirmed.[2,4-7] Radiologic findings of FCOD depends on the degree of maturation of the lesion. Radiographs often demonstrate numerous, irregularly shaped, sclerotic radiopacities admixed with diffuse ill-defined, radiolucent–radiopaque areas. In mature cases, lesions appear as completely radiopaque with radiolucent periphery and surrounded by sclerotic borders same as periapical cemental dysplasia. FCOD affects both jaws bilaterally, mainly mandibular posterior region and occurs above the inferior alveolar nerve canal.[8-10] Few reports commented on the presence or absence of jaw expansion. Yonestu and Nakamura reported mild bony expansion based on occlusal

radiographs or axial Computed Tomography images.[10] On the basis of the clinical and radiographic features of the present case, the lesion was diagnosed as FCOD with associated dentigerous cyst. Studies showed that simple bone cyst is the most seen associated lesion of FCOD. Considering this high rate of oral alterations, patients with Apert syndrome may have large dental needs; thus, dental professionals should be well informed on the oral aspects of these individuals, to be able to provide thorough information to patients and parents. Knowledge on the oral disturbances present in this syndrome is fundamental to establish a treatment plan adequate to their needs, clinical and radiographic follow-up is essential.

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