

# Uncommon Cone-Beam Computerized Tomography Findings in McCune-Albright Syndrome in an Implant Candidate Patient: A Case Report

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**Introduction:** McCune-Albright syndrome is a rare disease, characterized by triad of café-au-lait spots, endocrinopathies and fibrous dysplasia. These bone lesions are usually revealed during the first decade of life, together with pain, pathological fractures and secondary deformities.

**Case Presentation:** A 40-year-old female patient presented an opaque lesion at the left mandibular side of face, in a cone-beam computerized tomography (CBCT) view, during the implant placement evaluations. The patient had experienced precocious puberty and had undergone hysterectomy. Unilateral café-au-lait spots were present on patient's left side of the face. There was no expansion in intraoral examination. The oral mucosa was also normal. No asymmetry was detected. The analysis of sample histopathology confirmed fibrous dysplasia.

**Discussion:** In this patient we preferred following up. Afterwards, total surgical lesion resection can be performed. After a long-term follow-up, the area may receive an implant.

**Keywords:** Cone-Beam CT; Dental Implant; McCune-Albright Syndrome

## 1. Introduction

McCune-Albright syndrome is a rare disease, characterized by triad of café-au-lait pigmentation, endocrinopathies and fibrous dysplasia (1). The café-au-lait pigmentation is the presence of large irregular macules with hypermelanosis, occurring mainly on the anterior and posterior area of the neck, buttocks, thorax, back, shoulder and pelvis and rarely on the face, lips and buccal mucosa (2). It occurs in 10-20% of normal individuals and about 35% of patients with McCune-Albright syndrome (1). Fibrous dysplasia is more common in children and young adults. It usually ceases at the age of skeletal maturity and is most common at the maxillary posterior region (3). These bone lesions usually seen during the first decade of life, presenting with pain, pathological fractures and secondary deformities (1). Somatic mutations, during early developmental phases, cause mosaicism of normal & mutant-bearing cells, therefore, different clinical presentations can be detected in each patient (1). Post-zygotic activating mutation in GNAS (the gene encoding  $\alpha$ -subunit of G protein, the adenylate cyclase stimulator) is the cause of this syndrome (4). Post-zygotic

activating mutation in GNAS, cause increased production of recurrent AMP and leads to hyperfunctional target tissue cells (4). Bone lesions are usually asymptomatic, with slow and progressive growth, resulting in asymmetry (5). Due to the ceased growth of the bone lesion, the patient discussed did not have apparent asymmetry fibrous dysplasia (FD) of the craniofacial bones can cause visual disturbances, proptosis, nasal obstructions, facial asymmetry and hearing abnormalities (6).

## 2. Case Presentation

A 40-year-old female patient was referred for dental implant placement, having a panoramic view of her teeth. There were no unusual findings detected in the panoramic image or during the clinical examinations (Figure 1). CBCT view showed an opaque lesion at the left mandibular side. The opacity was about 3 cm in length, from the second premolar, up to the third molar region (Figure 2). The patient had experienced precocious puberty and had undergone hysterectomy. She presented unilateral café-au-lait pigmentation on her left side of the face (on the frontal and lateral parts of eye and cheek).

### Implication for health policy/practice/research/medical education:

This study would be helpful in diagnosis of McCune-Albright syndrome.

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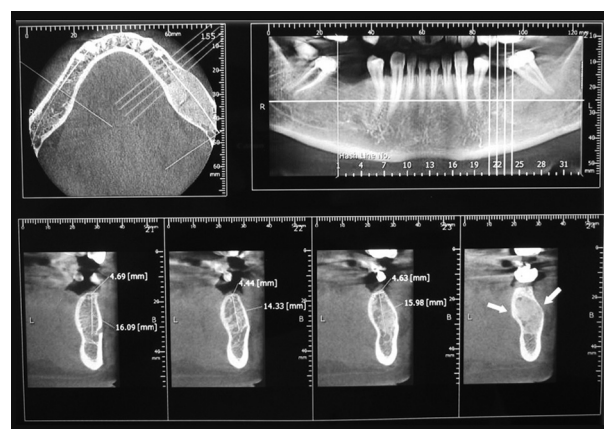
The patient had records of using medications for reducing pigmentation of the cafe-au-lait macules, for 28 years. There was no expansion detected during clinical examinations. The oral mucosa was normal. No asymmetry was seen (Figure 3). There was no complaints of pain and no tenderness was detected. A bone sample was taken by a surgical procedure, using a trephine bur (Figure 3). The microscopic analysis confirmed fibrous dysplasia, having an irregular trabecular pattern of immature bone, in a hyper cellular fibrous connective tissue. The trabecular bone was surrounded by linear osteoblasts. There was no capsule surrounding the bonny lesion, which was mixed with the normal bone tissue. Clastic cells were surrounding the trabecular bone tissue and occupying inside the bone lacunae (Figure 4). The bone lesion was not well demarcated from the surrounding bone, as detected during the surgery, for the specimen biopsy. The CBCT showed radiopaque areas with ground-glass appearance, fusiform expansion pattern, and thinning of the mandible cortical margin, without the radiolucent rim, suggestive of the fibrous dysplasia. The teeth are not displaced (Figure 2).

### 3. Discussion

In this patient the differentiating the fibrous dysplasia from ossifying fibroma was performed based on the fusiform pattern of expansion, self-limited bony lesion and woven trabecular pattern, present in the fibrous dysplasia (2). Hart et al. found that lesions in the craniofacial region were established earliest. Our patient had a low grade type of the lesion that started from the craniofacial region and stopped quickly, therefore the lesion could not be detected in clinical examinations. They also mentioned that late appearance of new lesions is not common in patients with FD. Due to the asymptomatic growth of the lesion in our patient, the diagnosis was made later. (7). Xavier et al. (2) reported a similar lesion in the mandibular region, in a middle aged woman, although there was no expansion in the case of our patient's and this is a rare finding. This shows the importance of both clinical and radiographic examinations. Bone fibrous dysplasia occurs in association with hyper pigmented skin lesions (1). In the case presented here, fibrous dysplasia and cafe-au-lait pigmentation were both on the left side of the patient's face, whereas Laditan (8) reported swelling of

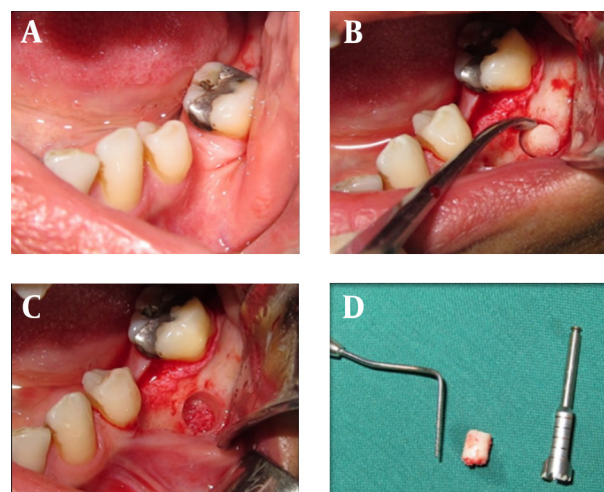


**Figure 1.** Panoramic View of Patient

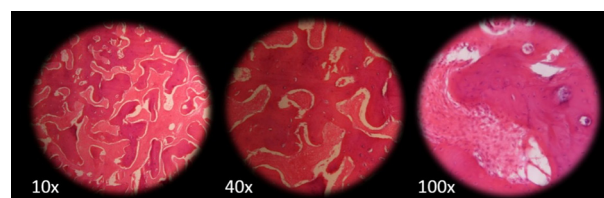


**Figure 2.** The Cone-Beam Computerized Tomography of the Patient, Showing Blending Opacity and Expansion

**Figure 3.** Biopsy Procedure



A: Clinical appearance of the lesion, B: Luxation of the biopsy sample by a periosteal elevator, C: Appearance of the remained bone after biopsy removal, D: Biopsy sample removed by a trephine bur.



**Figure 4.** Histological View of the Immature Bone Irregular Trabecular Pattern in a Fibro-Cellular Matrix

both right maxilla and the occipital bones and two areas of pigmentation on the right side of the trunk. In the panoramic view of this patient, no evidence of the lesion was present, in contrast with the findings of Xavier et al. (2), which emphasizes on the importance of performing detailed various examination methods be-

fore treatment. The CBCT showed radiopaque areas with ground-glass appearance, fusiform expansion pattern, and thinning of the mandible cortical margin, blending with the surrounding bone, without the radiolucent rim, suggestive of fibrous dysplasia (3). The analysis of the histopathology view confirmed fibrous dysplasia, consisting of irregular immature trabecular bone tissue in hyper cellular fibrous connective tissue. There was no capsule surrounding the bony lesion mixing with the normal bone tissue (2). These findings were also present in cases reported by in Xavier et al. (2). Some data suggest that the cancer incidence in adulthood increases in patients with FD (10). There is no specific treatment for McCune-Albright syndrome and due to the great variety of lesions in this syndrome, the treatments are almost specific to each patient. Generally, the disease stabilizes with the skeletal maturity and for this reason, surgical interventions can be delayed until the skeletal maturity occurs in the adult age (7). Surgery is recommended when there is a probability of a functional cranial nerve deficit, to prevent or limit the permanent loss of function (8). The decrease in the bony tissue and the increase in the fibrous tissue, decrease the rigid fixation of the implant and make the area more susceptible to local infection processes (3, 11). McCune-Albright syndrome is a rare syndrome and we diagnosed an occult form of it in the mandible bone of a female patient's face. The important point is that the patient was symptom free during clinical examinations and panoramic radiography did not reveal any abnormalities either, therefore the trigger for more evaluations was the CBCT for dental implant placement. This reveals the importance of comprehensive and meticulous examinations, before any dental treatment (7, 8). In this patient we would prefer following up. Later, total surgical resection of the lesion can be performed. After a long-term follow-up, the area may receive an implant (10, 11).

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