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CASE REPORT / OLGU SUNUMU

Tumoral Calcinosis: Clinical and Radiological Findings

Tümoral Kalsinozis: Klinik ve Radyografik Bulgular

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ABSTRACT

Tumoral calcinosis (TC) is an infrequent hereditary ailment described by localized ectopic calcification around the joints in affected regions. Reports of TC patients manifesting dental findings are scarce. Dental radiographs of a TC diagnosed patient show pulp stones, obliterated pulp cavities and short roots. In this case report, a 21-year-old female patient with TC was clinically and radiologically evaluated.
Keywords: calcification, calcinosis, dental radiography, pulp calcification, tooth abnormalities

ÖZ

Tümoral kalsinozis (TK), etkilenen bölgelerdeki eklemler etrafında anormal kalsifikasyonla belirgin, nadir görülen kalıtsal bir bozukluktur. TK hastalarının dental bulgularını içeren raporlar sınırlıdır. TK tanısı konmuş bir hastanın radyografisinde kısa kökler, pulpa taşları ve oblitere olmuş pulpa boşlukları belirgindir. Bu olgu sunumunda, 21 yaşındaki TK tanılı kadın hasta klinik ve radyolojik bulgular yönünden değerlendirilmiştir.
Anahtar Kelimeler: dental radyografi, diş anomalileri, kalsifikasyon, kalsinoz, pulpa kalsifikasyonu.

INTRODUCTION

Tumoral calcinosis (TC) is a progressive ailment described by the accumulation of calcium salts and crystals in periarticular soft tissues. TC lesions are predominantly noted on the extensor surfaces of the limbs, shoulder, the trochanter of the hip, the posterior aspect of the elbow, the humeral head, the acromio-clavicular joint, the inferior angle of the scapula, as well as the hands and feet (Witcher et al., 1989). The disease is infrequent, and there is no consensus on its etiological factors and pathogenic mechanisms. It typically presents between the ages of 10 and 30, being extremely rare in early childhood. There have been a total of 16 reported cases during infancy (Hammoud et al., 2005).

The expression TC was introduced by Inclan in 1943, describing slow-growing calcified masses commonly observed in proximity to major joints such as the elbow, shoulder, and hip (Inclan, 1943). Duret provided the first description of TC in 1989 (Duret, 1899). The initial study carried out in Turkey was published by Hacıhanefioğlu in 1978 in the English literature (Hacıhanefioğlu, 1978).

Familial TC cases have been predominantly documented in the Black population, with no observed gender bias (Gal et al., 1994). Familial tumoral calcinosis is more commonly observed in Africa and New Guinea, whereas it is rare in the populations of Europe and North America (Marinho et al., 1999). Also, TC cases are nearly absent in Asian region (Hunter et al., 1973).

TC is divided into two distinct groups: the primary form that occurs without an underlying disease (also known as familial TC), and the secondary form, which is linked to underlying conditions such as hypervitaminosis D, chronic kidney failure, scleroderma, sarcoidosis, primary hyperparathyroidism, milk-alkali syndrome, and malignancy. Within familial TC a subclassification is employed, distinguishing between a normophosphatemic variant described by sadnormal levels of serum calcium and phosphate, and a hyperphosphatemic subtype displaying normal serum calcium levels but elevated phosphate concentrations. (Olsen & Chew, 2006; Polykandriotis et al., 2004).

The clinical symptoms of TC include multiple or single hard, painless swellings in periarticular areas without redness, reducing the patient's capacity for joint movement. These swellings can sometimes lead to ulceration with chalky discharge and secondary infection on the overlying

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skin. Additionally calcified masses that are observable and palpable, in conjunction with dental and ocular abnormalities, may also be detected (Olsen & Chew, 2006; Polykandriotis et al., 2004; Foster et al., 2014).

There are few reports describing dental and oro-facial symptoms in familial TC (Dumitrescu et al., 2009; Krstevska et al., 2012; Naikmasur et al., 2008). Numerous manifestations affecting the facial and intraoral soft tissues have been documented in individuals with TC. These manifestations include angular cheilitis; maculopapular rash, erythematous patches, and calcified masses localized in the facial region. Additionally, there are erythematous lesions evident in diverse regions of the tongue, buccal mucosa, palate and adherent gum with papillary hyperplasia. Also, alterations in periodontal conditions are evident (Gal et al., 1994). Radiographically, most researchers have documented pulp stones, short and bulbous roots, and incomplete or complete obliteration of the pulp cavity in the impacted teeth (Witcher et al., 1989; Naikmasur et al., 2008; Burkes et al., 1991; Krsteyska et al., 2012; Favia et al., 2014). The observations resemble dentin dysplasia, featuring short and bulbous teeth with irregular calcifications (Ramnitz et al., 2016; Krstevska et al., 2021). The existence of abnormal calcification within the pulp space may impede the progress of root canal treatments (Stewart, 1995).

In this case presentation, the detailed clinical and radiological findings of a patient diagnosed with TC, who presented to our clinic with pain and mobility complaints, have been evaluated, and treatment options have been discussed.

CASE REPORT

A 21-year-old female patient presented to the Marmara University Faculty of Dentistry with a complaint of pain and advancing teeth mobility. Before coming to the faculty, the patient had previously visited a private dental clinic due to decayed teeth and pain. At this clinic, the patient underwent a tooth extraction procedure to alleviate the pain. When the patient applied to the faculty, her teeth were missing, decayed and mobile. The initial panoramic radiography, which was taken at the faculty, is shown in Figure 1.

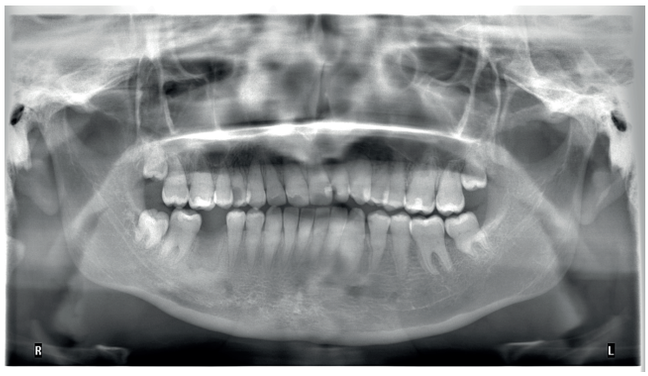

Figure 1: Patient's panoramic image; The X-ray was taken when the patient first applied to the faculty

Considering the diagnosis of TC, an examination was conducted on other family members, but no additional findings were identified. It has been learned that the mother and father are carriers of the TC. The serum concentrations of calcium and phosphorus in the patient were systematically documented. In our patient, the serum concentrations of calcium and phosphorus exceeded the established reference range (serum calcium, 10,6 mg/dl and serum phosphorus, 7,3 mg/dl) (Reference values: Serum calcium 8.6-10 mg/dl, Serum phosphorus 2.5-4.5 mg/dl).

In 2018, an X-ray of the right knee joint, requested by the Department of Orthopedics and Traumatology, observed a calcified mass (Figure 2). The patient underwent knee surgery later on.

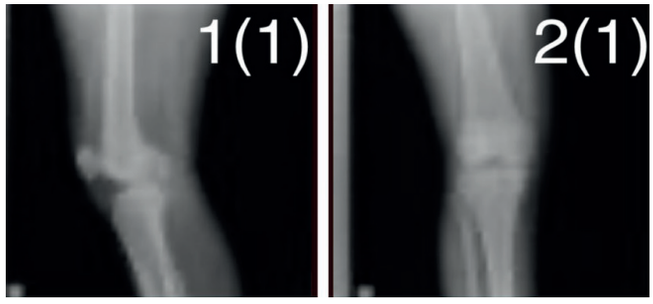

Figure 2: Calcified soft tissue mass at the right knee joint

As a result of the physical examination, no symptoms of TC were identified. During the intraoral examination, no abnormalities in the soft tissues were observed. The mucosa exhibited a normal color and structure. Normal salivary flow was observed from the major salivary glands. Additionally, no intraoral swelling was observed.

Teeth numbered 14, 16, 24, 25, 26, and 46 have been extracted in the patient due to caries or mobility (Figure 3). Teeth numbered 32, 31, 41, and 42 exhibited mobility. Teeth numbered 31 and 41 had severe mobility. The patient has a history of extraction due to mobility in non-carious teeth. The patient indicated in the taken medical history that the teeth crumble and fall apart like powder. In the medical history obtained, the patient mentioned that a previously filled tooth was causing pain and mobility, and they pulled it out painlessly with their own hands.

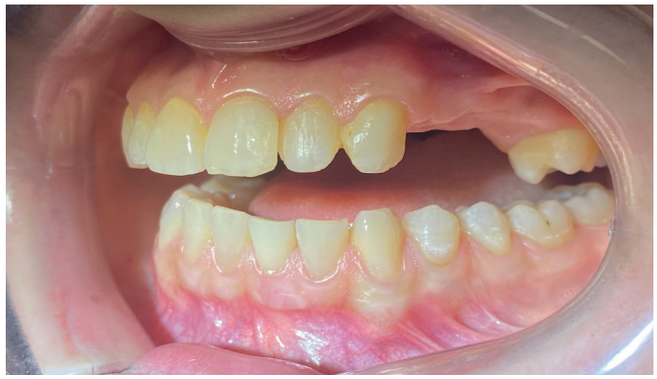

Figure 3: Patient's intraoral image; edentulous space and normal appearing clinical crowns

The patient exhibits general sensitivity to hot and cold, along with mobility in specific teeth. Especially tooth number 31 exhibited severe bone loss and mobility (Figure 4). Another noteworthy observation included the whole obliteration of both pulp chambers and root canals in all teeth. (Figure 5). Irregular areas on the root surfaces of some teeth were detected in periapical radiographs, such as in teeth numbered 33 and 34 (Figure 6).

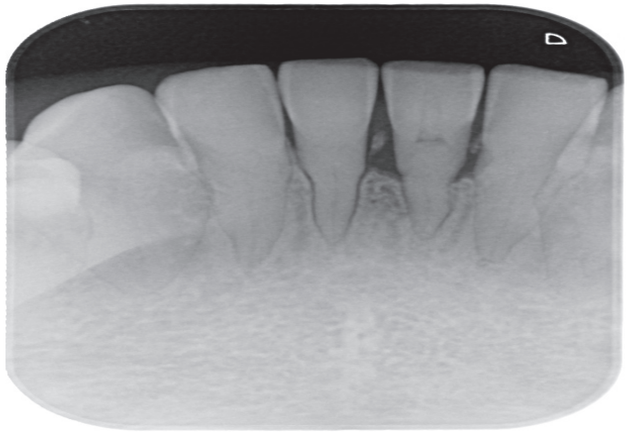

Figure 4: Periapical radiographs of the patient exhibiting bone loss

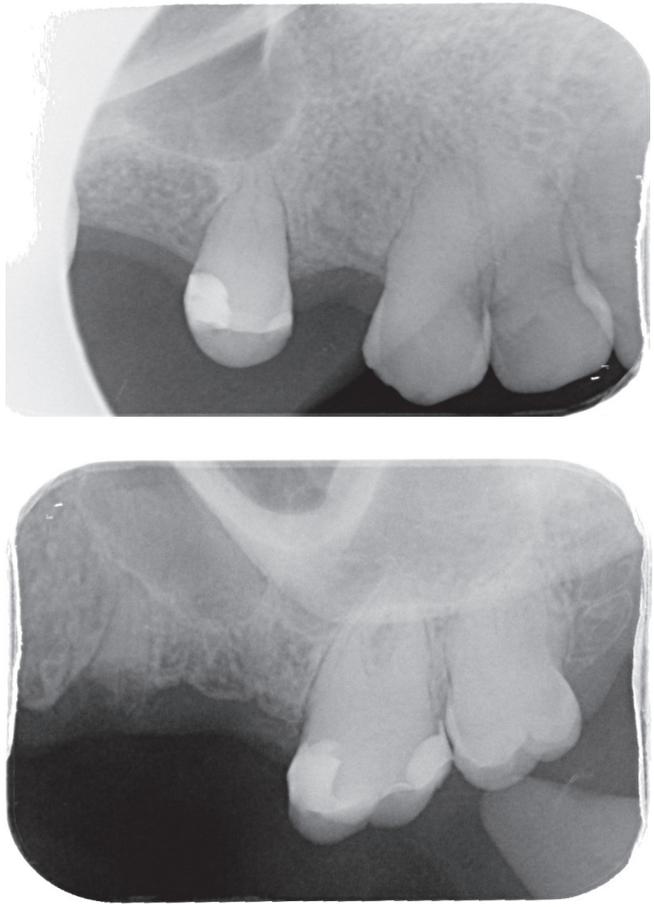

Figure 5: Periapical radiographs showing obliteration of pulp chambers and root canals

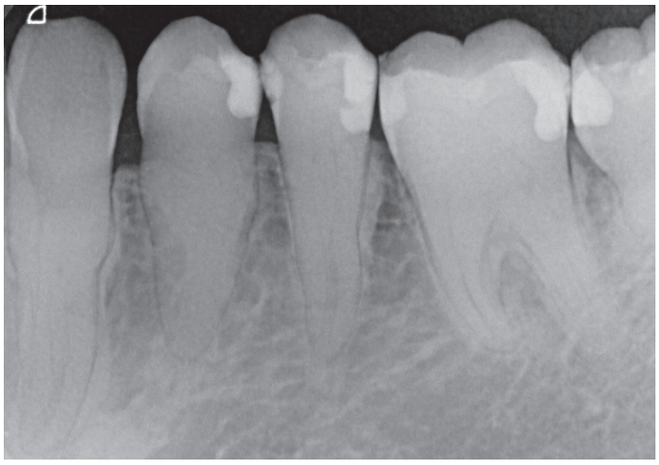

Figure 6: Periapical radiographs showing irregular areas on the root surfaces and nlargement of the coronal third of the roots

Normal appearing clinical crowns (Figure 3), the lengths and shapes of the patient's roots were also normal. There were no observed abnormally short or conical-shaped roots. Accessory roots and root dilacerations were not detected in the teeth.

A detailed medical history was taken from the patient at our faculty, and a clinical and radiological examination was performed. At our faculty, the patient's teeth with excessive mobility and bone loss were extracted. The patient's decayed teeth were filled. The patient's teeth numbered 14, 16, 24, 25, and 26 were extracted, and fillings were performed on teeth numbered 15, 17, 22, 23, 27, 32, 35, 36, 37, 47, and 48. The patient was referred to the prosthetics department for the edentulous spaces.

DISCUSSION

TC is an infrequent hereditary autosomal disorder distinguished by the existence of a cystic mass in the periarticular area containing calcified materials. It is characterized by various clinical manifestations. Possible causes encompass a congenital anomaly in phosphate metabolism, renal osteodystrophy, hypercalcemia due to sarcoidosis, or potentially an idiopathic origin (Davies, 2002). The predominant age group affected is children and adolescents, particularly those between 6 and 25 years old (Yochum & Rowe, 2005). Similarly, the patient mentioned in this case presentation was 21 years old.

Clinical symptoms typically involve periarticular, subcutaneous, and soft tissue calcifications occurring in areas prone to recurrent trauma. In this case, calcified masses were also observed in the right knee joint. An alternative manifestation of TC is dental anomalies. Existing literature, it has been observed that the majority of dental manifestations of TC are documented in individuals with hyperphosphatemia (Dumitrescu et al., 2009; Burkes et al., 1991; Favia et al., 2014; Hunter et al., 1973).

Clinically; enamel hypoplasia (Hunter et al., 1973; Favia et al., 2014), morphological changes in crowns (Burkes et al., 1991; Naikmasur et al., 2008) or normal coronal tooth structure (Witcher et al., 1989) have been identified. In this case, similar to the findings of Witcher et al., normal coronal tooth structure was observed.

Gal et al. conducted an investigation into head and neck symptoms among a cohort of six patients, revealing the presence of calcified masses in varying distributions across facial regions such as the cheeks, nose, jaw, and lips. Notably, one patient exhibited papillary hyperplasia of the lips, and all individuals displayed early periodontal alterations typified by marginal gingivitis. The soft and hard palates of all six patients exhibited erythematous changes, while three patients showed such alterations on the buccal mucosa and five patients on the tongue. It is noteworthy that calcified deposits were uniformly found on the cheeks, nose, jaw, and lips in all six cases, and importantly, none of these patients manifested atypical calcifications in periarticular tissues (Gal et al., 1994). In this case, however, the soft tissues were normal, and the gums were healthy. There were no calcified deposits in the neck and head region. Three cases of TC have been reported in the temporomandibular region (Shirasuna et al., 1991; Noffke et al., 2000; Zanetti et al., 1994) and one case has been reported in the premaxilla (Marinho et al., 1999).

Dental observations encompass thistle-shaped dental pulps, short and bulbous teeth, shortened roots, root dilacerations, obliteration of pulp chambers and root canals, pulp stones and taurodontism (Burkes et al., 1991; Dumitrescu et al., 2009). In this case as well, obliteration of the pulp chamber and root canal is similarly detected but unlike the previous case, there were no root dilacerations.

Yılancı et al. observed short bulbous roots, coronal expansion in the upper third of the root, the presence of oval radiopacities in this region, obliteration of the pulp, periapical radiolucencies in non-carious teeth and root dilacerations. They also noted that the most commonly impacted teeth were incisors and premolars (Yılancı et al., 2017). Burkes et al. recorded that the most notable alterations were discerned in the premolars of both the maxillary and mandibular arches (Burkes et al., 1991).

In the literature, significant bilateral mandibular tori and bone sclerosis of the mandible, maxilla, and skull have been recorded in one patient (Krstevska et al., 2012). Additionally, cases have been recorded in the literature involving progressive enlargement of gingival and alveolar tori, hypoplasia in the maxillary and mandibular regions, skeletal Class II malocclusion, and the presence of a deep bite (Krstevska et al., 2012; Favia et al., 2014). In this case, there were no torus enlargement, and malocclusion was not present.

Favia et al. reported multiple impacted teeth related to TC (Favia et al., 2014). In this case, no impacted teeth were observed. The findings in this case resemble those reported by Polykandriotis et al., as well as the observations in the publications of Burkes et al. (Polykandriotis et al., 2004; Burkes et al., 1991).

Dentists should be cognizant of the distinctive features of pulpal obliteration and the presence of roots resembling thistles, as dental radiographic observations may serve as the initial indication of the disease, especially in patients lacking other systemic manifestations (Yamamoto et al., 2016; Topaz et al., 2004; Ichikawa et al., 2007).

The uncommon occurrence of enamel abnormalities in TC may elevate the risk of caries; meanwhile, more frequently encountered pulp calcifications hinder the endodontic treatment of carious lesions (Favia et al., 2014). As a result, the implementation of dental sealants for the preventation of caries is recommended as a more effective treatment choice.

CONCLUSION

TC is an uncommon disorder of mineral metabolism observed among adolescents and juvenile adults, distinguished by the pathological accumulation of calcific masses surrounding major joints. Laboratory analyses commonly unveil hyperphosphatemia alongside normocalcemia, normal parathyroid hormone levels, and alkaline phosphatase within the standard range. Dental observations bear similarities to dentin dysplasia, encompassing the obliteration of both root canals and coronal pulp chambers to varying degrees. The obliteration of pulp chambers and root canals presents a challenge in endodontic treatment procedures. Dentition may be periodontally affected, leading to early permanent tooth loss. It is crucial for the dentist to be aware of the dental indications of TC when treating a juvenile patient with completely or partial obliteration of pulp chambers or root canals.

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CONFLICTS OF INTEREST

The authors confirm that there are no conflicts of interest.

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