Suspected Endodontic Failure in a Patient with Cleidocranial Dysplasia: A Case Report

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ABSTRACT

A patient's medical history and related dental manifestations can significantly contribute to confounding signs and symptoms leading to a diagnostic challenge. An 18-year-old female patient presented with persistent radiographic radiolucency associated with the apex of a previously treated tooth (tooth 9); asymptomatic apical periodontitis and endodontic failure were suspected. This report presents how a patient's condition of cleidocranial dysplasia had a profound effect on her dental history, which included the presence of multiple supernumerary teeth. Extensive surgical intervention during the patient's childhood was required to remove the supernumerary teeth, which resulted in an endodontic misdiagnosis in her adult life. After clinical and radiographic examination, the patient was diagnosed with a periapical scar. Periapical fibrous scars have a prevalence of between 2.5% and 12% and are a rare healing process with fibrous tissue after surgical and nonsurgical interventions. This report describes the diagnosis and pathophysiology of fibrous scars, including their risk factors and long-term monitoring approaches. *(J Endod 2023;49:445–449.)*

KEY WORDS

Apical scar; case report; diagnosis; pathology

SIGNIFICANCE

This report discusses the importance of a holistic approach and the relevance of a patient's medical and dental history when faced with a diagnostic dilemma. Knowledge of historic radiographic findings and details of previous treatment provided can provide vital information to prevent misdiagnosis.

Cleidocranial dysplasia or dysostosis is an autosomal dominant genetic condition affecting tooth and skeletal development with a prevalence of 1 in 1 million[1]. It was first identified by Martin in 1765[2] and has been reported to affect intramembranous ossification caused by a mutation, insertion, or deletion of the *CBFA1* gene[3].

Typical presentation involves underdeveloped or absent clavicles, shorter stature, and skull abnormalities including frontal and parietal bossing. A hypoplastic maxilla and delayed fusion of cranial sutures, which lead to a protruding mandible and midface retrusion, are also commonly present[4]. Dental abnormalities include delayed exfoliation of primary teeth, delayed eruption of primary and permanent teeth, occlusal abnormalities, and supernumerary teeth, which can lead to crowding and resorption[4]. Patients may complain of functional occlusal problems and poor aesthetics of their teeth, with some presenting with enamel hypoplasia, taurodontia, and dentigerous cysts[5].

The aim of this report is to showcase how a patient's complex medical history can have a profound effect on dental history and result in a diagnostic challenge. Diagnostic approaches and variable management strategies are discussed in this study.

CASE REPORT

An 18-year-old female patient was referred to the endodontic department at Liverpool University Dental Hospital because of a suspected failed root canal treatment on the maxillary left central incisor (tooth #9). The patient was previously seen at the pediatric department 2 years prior presenting with a discolored tooth #9 that did not respond positively to sensibility tests. After a diagnosis of pulpal necrosis and asymptomatic apical periodontitis, primary endodontic treatment was completed. The tooth has been asymptomatic since. The patient's medical history includes cleidocranial dysplasia with no known family history of the condition. Her dental history includes multiple supernumerary tooth extractions under general anesthesia at age 10.

The patient reported no signs or symptoms. The clinical examination revealed normal responses to sensibility tests of the adjacent teeth and no tenderness to palpation/percussion testing of all maxillary anterior teeth. There was no evidence of sinus, discoloration, deep periodontal pocketing, mobility, or

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coronal leakage. The new referral supplied a periapical radiograph (Fig. 1), which showed well-condensed obturation with the presence of radiolucency at the apex of the tooth consistent with the appearance of periapical periodontitis. At this stage, the main differential diagnosis was asymptomatic apical periodontitis of the previously treated tooth (tooth #9).

A cone-beam computed tomographic (CBCT) radiograph was taken (Fig. 2), which indicated that tooth #9 was well obturated and had an associated periapical radiolucency. The images were interpreted by an oral and maxillofacial radiologist who reported a low-density apical region measuring approximately 5.6 mm in the mesiodistal direction and extending approximately 2.7 mm superior to the apex of tooth #9. The periapical area was reported to extend through the buccal and palatal cortices adjacent to the apex of tooth 9 and through the adjacent floor of the nose.

Because of the patient's previous history of supernumerary tooth extractions, previous CBCT radiographs were requested and analyzed to determine the number and locations of the supernumerary teeth. Upon further assessment of the CBCT records from 8 years prior, the presence of a supernumerary tooth was noted in the anterior maxillary region (Fig. 3). The position of the current observed radiographic radiolucency was closely related to the supernumerary tooth that was previously surgically extracted from the maxillary anterior area. Furthermore, CBCT records from 2 years ago, before the start of the initial endodontic therapy, showed no changes in size of the radiolucency before and after the completion of the endodontic treatment of tooth #9 (Figs. 2 and 4).

The patient was diagnosed with an apical scar located in the anterior maxilla associated with the apex of tooth 9 that was likely caused by the surgical extraction of the supernumerary tooth in the anterior maxillary region. Because of the absence of evidence of active disease and no changes in radiographic appearance, no treatment was deemed necessary.

DISCUSSION

Despite its rare prevalence, cleidocranial dysplasia can have a significant impact on patients' oral health and development. The mutation of the *CBFA1* (also known as *RUNX2*) gene affects the function of osteoblasts, chondrocyte differentiation, and processes of skeletal morphogenesis. Tooth morphogenesis and histodifferentiation of the epithelial enamel organ are also regulated by the *RUNX2* gene through epithelial-mesenchymal interactions[6]. In patients with cleidocranial dysplasia, these processes are disturbed, leading to supernumerary teeth that may form because of the lack of resorption and the presence of excess dental laminae[5]. Various cranial abnormalities are also reported in the literature, including frontal bossing and delayed ossification of skull sutures, facial bones, and the cranial base[6,7].

Because of the complexity of the treatment required, a multidisciplinary treatment approach is usually undertaken[8]. In most cases, this includes a combination of orthodontics with oral and maxillofacial surgery, aiming to remove the supernumerary and deciduous teeth combined with exposure of the permanent teeth to facilitate their eruption. The timing and delivery of the surgical management can vary depending on the type of modality used, such as the Toronto-Melbourne approach or the Belfast-Hamburg approach[6]. Orthodontic intervention is then delivered to guide the eruption and alignment of the permanent teeth with the option of orthognathic surgery to improve the skeletal relationship if required[4]. Depending on the case complexity, the treatment time can take multiple years, starting at 5 to 6 years old with the Toronto-Melbourne approach and finishing in adulthood with the delivery of orthognathic surgery.

Bone resorption in the periapical region, which can be recognized as radiographic radiolucency, is a vital diagnostic feature of apical periodontitis[9]. After successful root canal treatment, evidence of osseous regeneration is manifested by radiographic reduction and resolution of the periapical lesion[10]. However, it is well reported in the literature that periapical healing can be a slow process with about 50% of cases showing evidence of radiographic reduction in the size of the lesion at a 6-month interval[11], with 15% of cases taking up to 48 months[12]. Although failure to completely disinfect the root canal system leading to persistent intraradicular infection has been shown as the most common cause for the persistent presence of periapical radiolucency, there are multiple other causes that need to be considered[13]. These include the presence of extraradicular infection because of periapical actinomycosis, a foreign body reaction to materials such as gutta-percha, the formation of true or pocket cysts, the collection of cholesterol crystals, and periapical scars[9,11,13]. Therefore, relying solely on radiographic appearance to determine the success of endodontic treatment can be misleading. Diagnosing an endodontic failure is a complex process combining historic radiographic examination, clinical signs, and the patient's symptoms. Additionally, as shown in this case, the patient's medical and

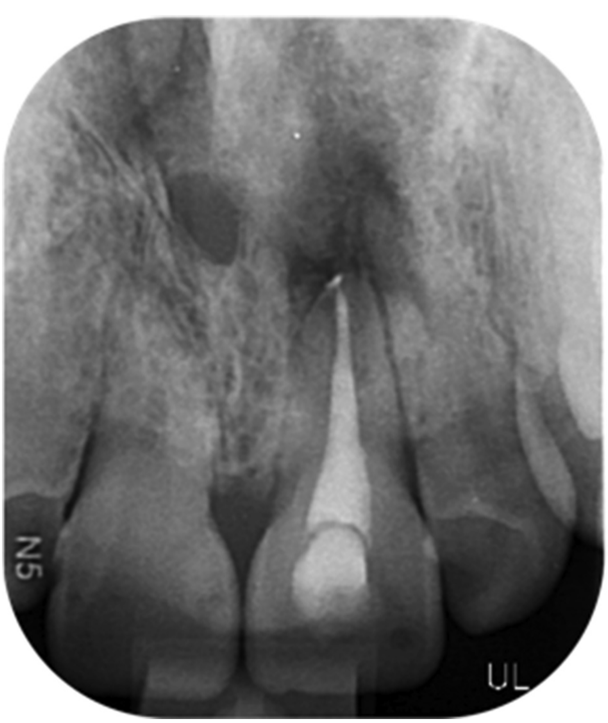

FIGURE 1 – The periapical radiograph supplied with the original referral showing a large periapical radiolucency associated with the apex of tooth #9.

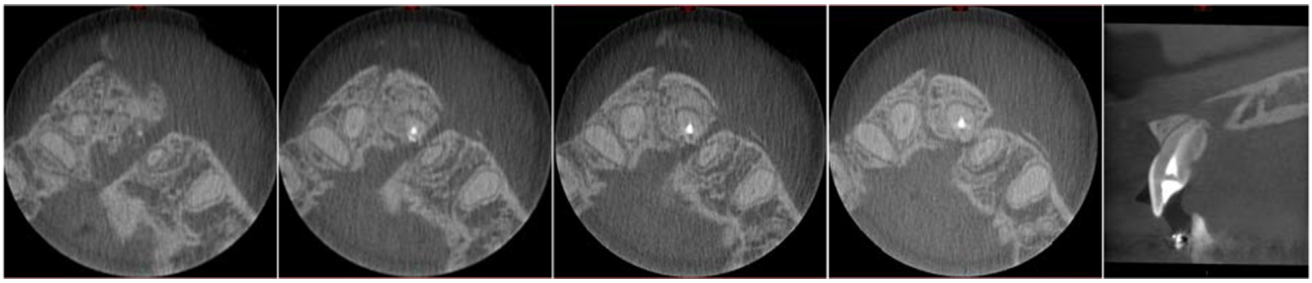

FIGURE 2 – The latest CBCT radiograph showcasing a well-obturated tooth 9 with a large radiolucent lesion affecting both the palatal and buccal cortices.

dental records can provide crucial information to aid in the diagnosis.

Periapical scars or postendodontic periapical fibrous scars are a rare healing process with collagenous fibrous tissue after endodontic therapy[14]. Their reported prevalence is between 2.5%[15] and 12%[13]. Histologically, they are composed of dense collagenous fibrous tissue with fibroblasts, which in the majority of the reported cases is free of inflammation. Lee et al[14] reported a periapical scar lesion that showed the presence of chronic inflammation; however, this was speculated to be caused by the presence of the embedded amalgam particles within the lesion. After successful completion of the root canal treatment, the bone regeneration process is started by the migration of osteoblasts or undifferentiated mesenchymal stem cells with the presence of the appropriate induction or bone growth factors[14]. The bone-forming cells are usually recruited from the nearby periosteum or endosteum. One of the reported risk factors associated with periapical scar formation is the absence and destruction of nearby cortical plates[11], which lead to the inability to recruit the required bone-forming cells. Instead, connective tissue-forming fibroblasts move into the area, and the healing process by the formation of scar tissue occurs[9]. Other factors such as the overproduction of type III collagen and disturbance to the remodeling phase of the wound healing process have also been reported[14].

Clinically, the stable periapical scar lesions are asymptomatic in nature, show good periodontal health, and have no evidence of root fracture[14]. However, some cases reported the presence of swelling and fistulation, which may appear if reinfection occurs[16].

Radiographically, teeth with associated periapical scars show evidence of high-quality endodontic treatment/retreatment, and the size of the radiolucent periapical scar lesion remains stable over time[14]. Certain radiographic features such as the presence of the lamina dura separating the apex of the tooth from the periapical scar lesion were also reported[11]. CBCT imaging has been used in these cases to monitor the radiographic size of the periapical scar because of its 3-dimensional imaging capabilities, high resolution, accuracy, and availability. However, its disadvantages compared with 2-dimensional traditional radiography include increased radiation exposure to the patient, a prolonged scanning time, and a higher cost. Other suggested monitoring methods include the use of magnetic resonance imaging and real-time ultrasonography (US)[11]. Magnetic resonance imaging does not use ionizing radiation and provides excellent soft tissue contrast, which has been shown to effectively monitor and identify the presence of scar tissue[17]. US is a different nonionizing alternative to CBCT imaging that has been shown to provide reliable diagnostic and monitoring capabilities of periapical lesions[18]. One of the main limitations of US is the requirement of the absence of a cortical plate to allow the passage of the ultrasound waves into the lesion. However, because of the nature of the formation of the periapical scar lesion, cortical plates would normally be missing, which may allow for this method to be used in the monitoring of these cases.

Because a diagnosis of a periapical scar is of histologic nature, a biopsy of the lesion enables a confirmation of the diagnosis. The limitations of this approach when used to obtain periapical lesions include its invasive nature, the risk of distortion of the specimen, and procedural errors during sectioning of the lesion resulting in a potential misdiagnosis. In fact, Simon et al[19] reported that CBCT imaging may be a more accurate and useful method to diagnose large periapical lesions compared with a biopsy. However, this study did not include a diagnosis of a periapical scar; therefore, the relevance of the results needs to be interpreted with caution. Alternatively, a novel approach by Zmener et al[20] has been suggested using a core bone biopsy needle to obtain the histologic sample of the periapical scar in a noninvasive manner. The decision of whether a biopsy of the lesion should be undertaken is determined by a discussion of the potential risks and benefits with the patient. Monitoring the suspected periapical scar area nonsurgically is less invasive to patients, and invasive interventions can be used if signs and symptoms develop.

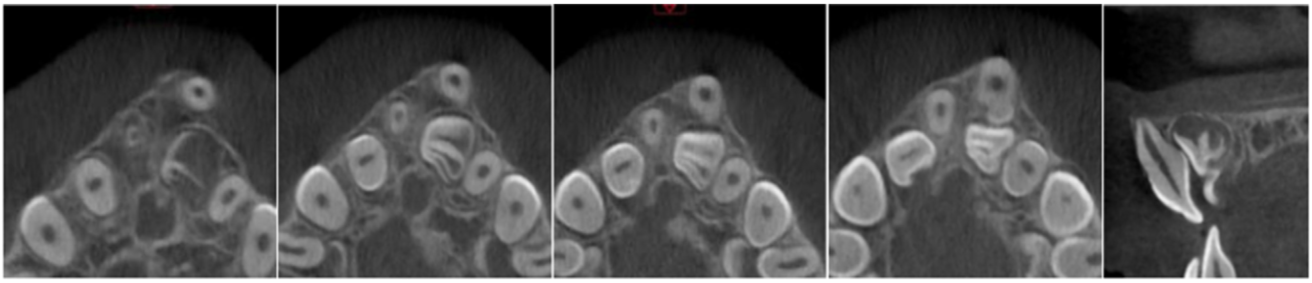

FIGURE 3 – A CBCT radiograph taken when the patient was 10 years old showcasing a supernumerary tooth in the anterior maxilla in close proximity to the erupting tooth #9. This radiograph was taken to assess the position of the supernumerary teeth in the maxilla before their surgical removal,

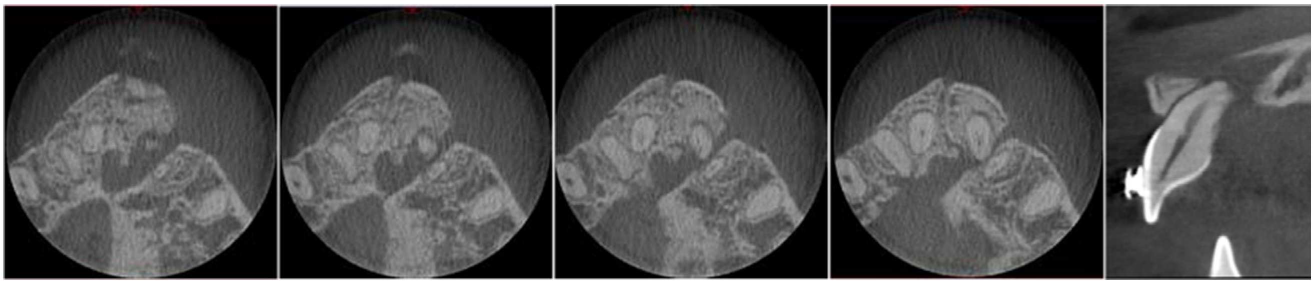

FIGURE 4 – A CBCT radiograph taken when the patient was 16 years old showcasing a large radiolucent bony cleft extending through both the buccal and palatal cortices adjacent to the root apex of tooth #9. This radiograph was taken to assess the anatomy of tooth #9 and the associated periapical radiolucency before the nonsurgical primary endodontic treatment of tooth #9.

The aim of this case report was to highlight the importance of thorough history taking, especially medical history, which in this case included cleidocranial dysplasia and the associated supernumerary teeth. The authors do not attempt to link a direct causation of the formation of apical scars to cleidocranial dysplasia. However, there is evidence to suggest that the presence of supernumerary teeth and their subsequent surgical removal contributed to the formation of the periapical scar. In this case, the clinical and radiographic findings were consistent with a diagnosis of a periapical scar; therefore, no histologic investigation was undertaken. The radiographic radiolucency has been stable in size over at least the last 2 years and will be regularly monitored clinically and radiographically in the future. Because of the rare prevalence of periapical scar lesions, patients with diagnosed apical scars should be given a letter or a dental passport summarizing the diagnoses and results of previous investigations. This should be given to their current and/or future dentist to prevent misdiagnosis and unnecessary treatment of the affected tooth.

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The authors deny any conflicts of interest related to this study.

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