Observation of supernumerary maxillary molars in a mother and her son diagnosed with Trichorhinophalangeal Syndrome Type I.

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Background

Trichorhinophalangeal syndrome (TRPS) is an autosomal dominant multisystem disorder caused by disruption or changes of TRPS1 gene[1]. Three types of TRPS (I, II and III) have been described, all of which exhibit the common features of hair, craniofacial and skeletal abnormalities[2]. Dental anomalies frequently associated with TRPS type I include presence of supernumerary teeth, congenital absence of teeth, malocclusion, delay in tooth development, abnormal tooth shape and extensive caries[3].

Case 1

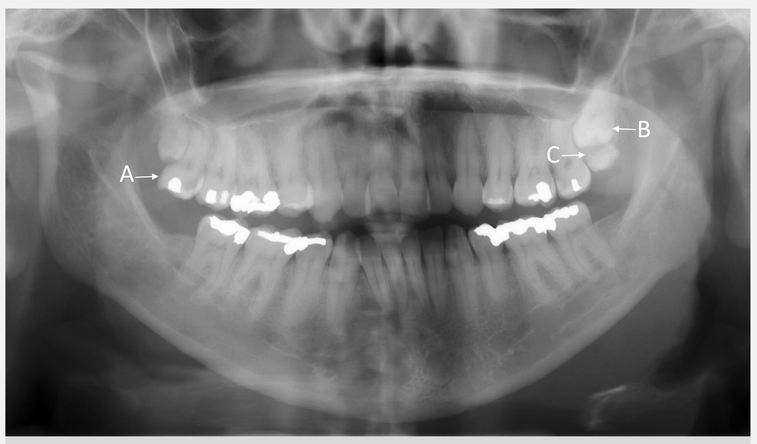

Figure 1: Panoramic radiograph shows a partially impacted supernumerary molar[A] posterior to 18, and a totally impacted supernumerary molar[B] posterior to 28 resulting root resorption[C] on 28. All first mandibular premolars were extracted due to orthodontic treatment.

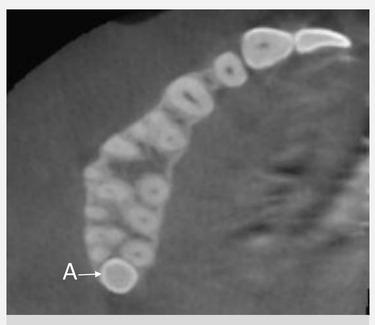

Figure 2: Axial CBCT image shows impacted supernumerary molar[A] on patient's right side with no root resorption on tooth 18.

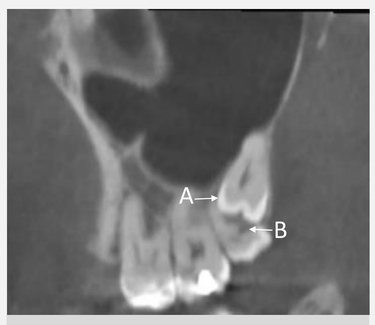

Figure 3: Sagittal image of patient's left side shows impacted tooth 29[A] causing severe root resorption[B] on tooth 28.

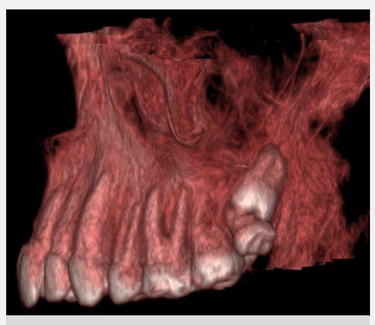

Figure 4: 3D representation of the same situation as in figure 3.

Case 2

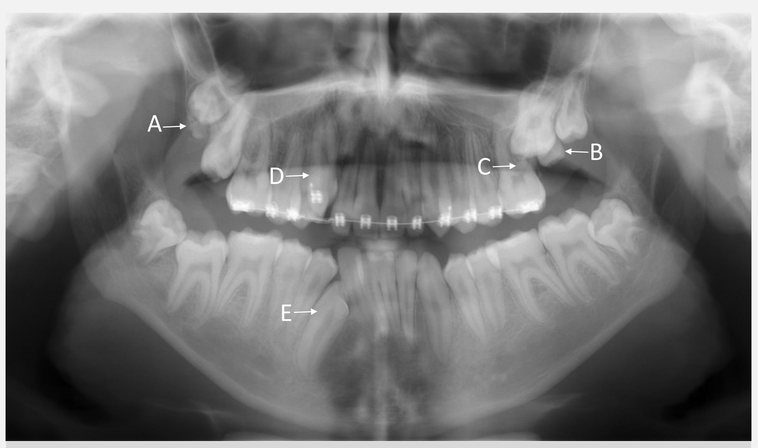

Figure 5: Panoramic radiograph shows a partially developed supernumerary molar[A] adjacent to impacted 18, and an impacted supernumerary molar[B] adjacent to impacted 27 with possible root resorption[C] on erupted tooth 26. It also shows formally impacted right maxillary canine[D] which is under orthodontic treatment and fully impacted right mandibular canine[E].

Goals

The aim of the study is to present dental anomalies observed in two TRPS type I patients, a mother and her son.

Method

We report two cases of TRPS type I patients, a mother and her son. In addition to the clinical features of the patients, the results of panoramic radiographs (both cases) and cone beam computed tomographic (CBCT) examination of the posterior maxilla (case 1) are presented.

Case 1 (Figure 1-4) involved a 41-year-old female with two maxillary supernumerary molars on each side. The one on the right side is partially erupted and the one on the left side is impacted (Figure 1), causing root resorption on 28 (Figure 1, 3, 4). Due to malocclusion, crossbite and midline shift, she received orthodontic treatment during which all four first premolars were removed. Maxillary second molars have advanced carious lesions (Figure 1).

Case 2 (Figure 5) involved her 17-year-old son with two impacted maxillary supernumerary molars on each side, one of which is not yet fully developed. He has severe malocclusion, impacted molars and canines, crossbite, midline shift and abnormal shape in certain teeth. He is currently receiving orthodontic treatment.

The patients in this study did not exhibit congenital absence of teeth. In both patients, surgical interventions might be necessary to remove the impacted molars.

Conclusion

1. Our study highlights the complex nature of dental anomalies in TRPS type I patients and a need for examinations of the oral cavity since certain dental anomalies can be of importance in determining TRPS type I diagnosis.

2. The absence of certain abnormal features emphasizes the significance of studying a number of different patients with the same syndrome to accurately identify specific syndrome-related features.

3. Our findings suggests that dental treatment plan in TRPS type I patients should be established by a multidisciplinary team.

References

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