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Case Report

Multiple supernumerary teeth in a child with Rubinstein-Taybi syndrome: a rare feature

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Abstract Rubinstein-Taybi syndrome is a multiple anomalies congenital disorder characterised by broad thumb and halluces, facial dysmorphism with mental and growth retardation. Oral features include small mouth, retro and micronagthic jaws, highly arched and narrow palate. Dental anomalies such as teeth with talon cusps and screwdriver shaped permanent incisors together with crowded teeth are common features in these patients. Although hyperdontia is said to be one of the features of this condition, nevertheless, presence of multiple supernumerary teeth has never been documented. This report highlighted a case of an eleven-year-old boy with Rubinstein-Taybi syndrome referred for unerupted permanent incisors who exhibited multiple supernumerary teeth radiographically.

Keywords: Dental anomalies, hyperdontia, Rubinstein-Taybi syndrome, supernumerary teeth, unerupted teeth.

Introduction

Rubinstein-Taybi syndrome (RTS), Online Mendelian Inheritance in Man (OMIM 180849), is a sporadically occurring congenital disorder first described in 1963 by Rubenstein and Taybi (Hennekam *et al.*, 1990). Its birth prevalence has been estimated at about one in 100,000–125,000 live births (Hennekam *et al.*, 1990). Occurrence of the syndrome in both sexes is reported to be similar. However, its presence among non-Caucasians is low, probably due to underreporting (Hennekam, 2010). Deletion or mutation of the CREB-binding protein (CBP) gene or that of the p300 gene has been linked with this condition in 55% of the cases reported (Hennekam, 2006).

Generally, the diagnosis of RTS is based on its clinical features such as delayed mental development, beaked nose with low hanging septum, grimacing smile, highly arched eyebrows, down slanting palpebral fissures, broad thumbs and big toes (Hennekam, 2006). Some of the reported oral findings in RTS patients include teeth with talon cusps, screwdriver shaped permanent incisors, high arched and narrow palate, dental crowding, anomalies in tooth numbers, crossbite, enamel hypoplasia and tooth wear (Hennekam, 2010; Bloch-Zupan *et al.*, 2007).

This case report highlighted the presence of multiple supernumeraries in a RTS patient that prevented the eruption of the permanent teeth. To the best of our knowledge this feature has never been reported before amongst the RTS patients.

Case history

An eleven-year-old Chinese boy presented to the Paediatric Dental Clinic, Faculty of Dentistry, UKM, with a chief complaint of unerupted upper front teeth. He was diagnosed with RTS. Clinically he appeared to be reserve and shy. His mother claimed he can converse normally despite of his learning difficulties. He is second of three siblings.

Physical examination revealed presence of broad angulated halluces and thumbs (Figs. 1 and 2). Facial features were characteristic of RTS, with broad forehead, broad nasal bridge, hypertelorism and downward slanting palpebral fissures (Fig. 3). He also has bilateral middle ear effusion

and was treated with T-tube insertion. Intra oral examination revealed, highly arched and narrow palatal vault, unerupted multiple permanent teeth especially the upper four incisors with evidence of healing sockets of recently extracted upper primary anterior teeth (Figs. 4 and 5). Some of his primary teeth were extracted by a private dentist to facilitate eruption of the unerupted permanent teeth prior to referral.

A panoramic radiograph (OPG) taken at the private dental clinic also revealed presence of multiple supernumerary teeth in all four quadrants. Six teeth were noted in the premolar regions, one each in the upper quadrants, three in the lower left quadrant and another in the lower right quadrant. In the anterior maxillary region another two supernumerary teeth were observed. All the supernumerary teeth were of supplemental type, with the exception of those found in the anterior region which appeared to be of tuberculate form. These teeth seem to impede the eruption of the actual permanent successor teeth (Fig. 6).

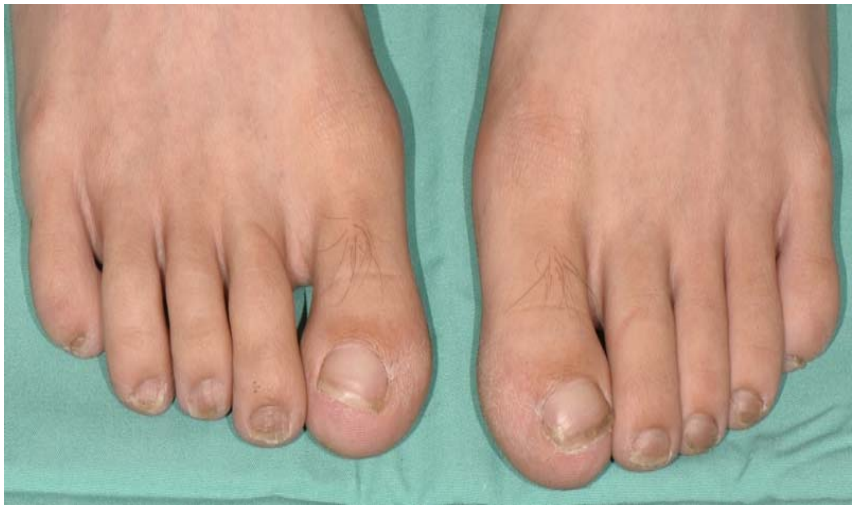

Fig. 1 Broad and angulated halluces.

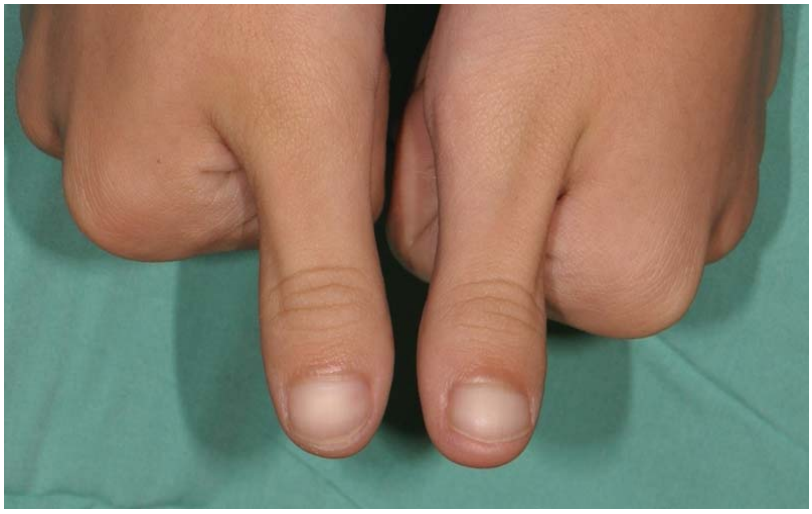

Fig. 2 Broad thumbs.

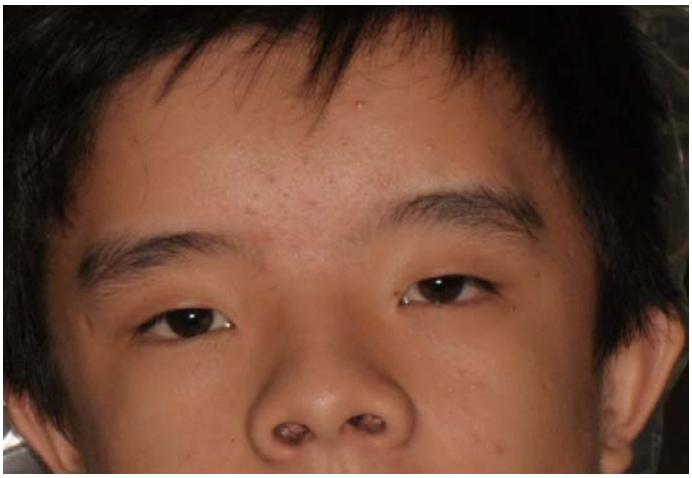

Fig. 3 Broad base nasal bridge with hypertelorism and downward slanting palpebral fissures.

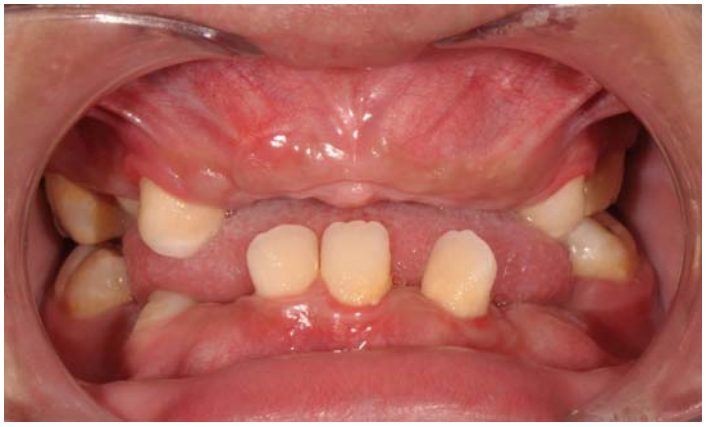

Fig. 4 Anterior view showing clinically missing multiple permanent teeth and malpositioned teeth.

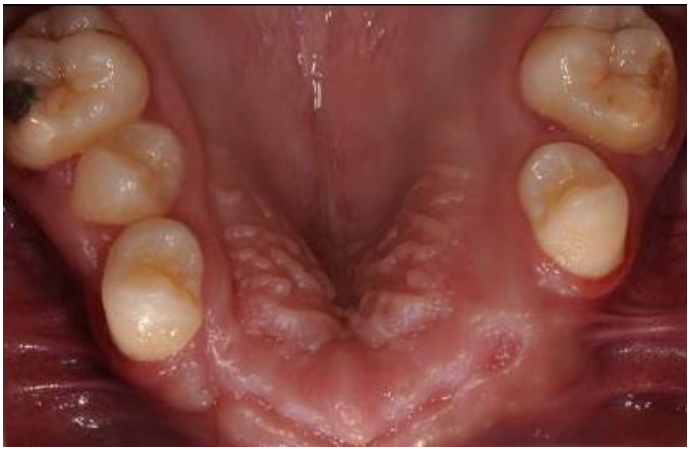

Fig. 5 The highly arched and narrow palatal vault unerupted upper anterior teeth.

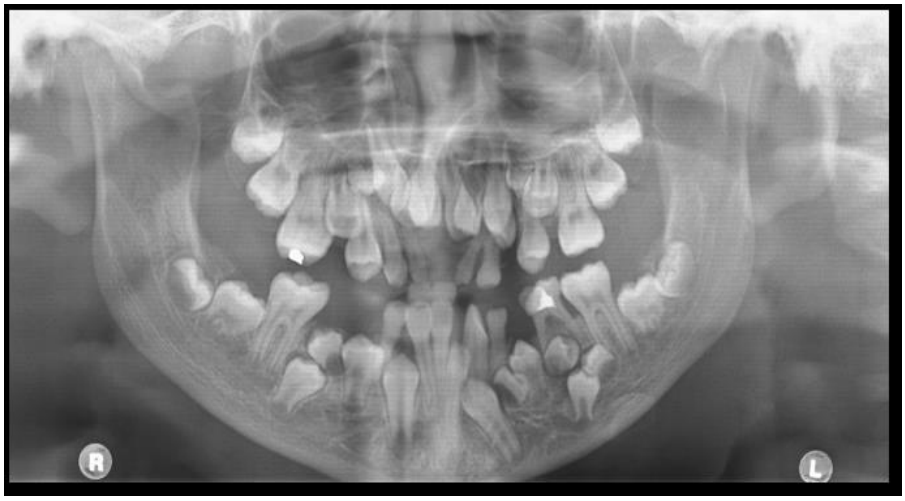

Fig. 6 A panoramic radiograph showing multiple supernumeraries at the premolar region of all quadrants, and between 23 and 11 region.

Discussion

The most important dental feature reported in Rubinstein-Taybi syndrome (RTS) is the high incidence of teeth with talon cusps in the permanent dentition (92%), which is often used as one of the diagnostic features for RTS patients. Talon cusps are referred to as accessory cusp-like structures that are often located on the lingual aspect of the incisors. Their presence sometimes can be detected in the jaws by panoramic radiographs (Hennekam, 2010). Despite the high incidence of talon cusps reported among the RTS patients, there was no evidence of such teeth either noted in the OPG or seen clinically in the present case.

It was also reported that hypodontia, hyperdontia, and natal teeth can manifest in RTS patients (Hennekam, 2010). Presence of supernumerary teeth has been reported in about 15% of cases of RTS patients (Hennekam *et al.*, 1990). Nevertheless, in most of the reported cases, patients had fewer supernumerary teeth (one or two) with common predilection towards the upper anterior region of the palate (Stalin *et al.*, 2006). Multiple supernumerary teeth (more than 5 teeth) are commonly associated with some syndromic disorders such cleidocranial dysplasia and Gardner's syndrome (Shah *et al.*, 2008). However, this feature has never been reported amongst the RTS patients before.

In the present case, the patient presented with a total of eight supernumerary teeth, of which, six were found in the premolar region of all quadrants and are similar in shape to the actual premolars. Often premolar region is one of the commonest sites for occurrence of multiple supernumerary teeth (Moore *et al.*, 2002). The other two supernumerary teeth in this patient were tuberculate in shape and were located at the upper anterior region of the palate.

Presence of supernumerary teeth may give rise to number of problems such as delay in the eruption of permanent teeth, displacement and rotation of teeth, dental crowding and other less common complications (Shah *et al.*, 2008; Rajab *et al.*, 2002). In the discussed case, presence of the supernumerary teeth had led to the delay in the eruption of the permanent teeth especially the upper right central and lateral incisors and left canine. Other possible reason for the delay in eruption of the permanent teeth may have been attributed to the presence of highly arched and narrow palatal vault. However, Hennekam (2010) reported that timing of the eruption of deciduous

and permanent dentition was normal in Rubinstein-Taybi syndrome patients.

Another feature seen in the radiograph is the developmental stage of the supernumerary premolars. Only the calcified crowns of these teeth were noted. Some of the actual permanent premolars were displaced due to the presence of these supernumerary teeth. It appeared that development of some of the supernumerary teeth might have taken placed much later than the actual permanent premolars. This was more obvious in the mandible, where the location of the supernumerary premolars were more coronally placed than the actual premolars which are unerupted. In view of this, the probability of late embryological occurrence of newer supernumerary teeth in future can't be ruled out. Therefore, there is a need to follow-up this patient periodically.

To the best of our knowledge, multiple supernumeraries with delayed eruption of permanent successors have never been reported in Rubinstein-Taybi patients thus far, making this patient a unique one. The present case highlighted the need for careful assessment of multiple supernumerary teeth in patients with Rubinstein-Taybi syndrome. Often, the commonest complication associated with multiple supernumerary teeth is failure of eruption of permanent teeth (Açikgöz *et al.*, 2006). Thus, early diagnosis with routine radiographs and intervention is important in minimizing aesthetic and functional complications later in patients with RTS.

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