

Clinical Report

Multiple Macrodontic Multituberculism

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Ekman-Westborg and Julin [1974: Oral Surg 38:217–222], described multiple macrodontia and multituberculism affecting the teeth without other anomalies (E-WJ). We describe a Chilean case in a 12-year-old with the typical dental alterations and with histopathologic findings that include absence of predentin layer and prominent reduced enamel epithelium. E-WJ is not a syndrome and we propose “multiple macrodontic multituberculism” as a better name for this anomaly of uncertain etiology affecting only the crowns of the teeth. © 2003 Wiley-Liss, Inc.

KEY WORDS: Ekman-Westborg and Julin disorder of teeth

INTRODUCTION

Ekman-Westborg and Julin [1974] first described characteristic anomalies of posterior teeth in a 14-year-old male. In addition to macrodontia, a number of teeth were impacted, and there was a cross-bite between upper and lower first molars. The parents stated that the deciduous teeth had been small and conical, especially the upper incisors. The scalp hair was noted to be very coarse and curly, compared with that of other members of the family. A canine, upper and lower second premolars, and mandibular second molars failed to erupt. One of the maxillary canines had a peak-shaped cusp with a step on the mesial side, suggesting a separate mesial cusp. The second premolars had enlarged multitubercular crowns with elevated nodules in the middle of the occlusal surface. The crowns were larger than normal, measuring 10.7×11.2 mm in

width (7.4 mm, SD 0.47 mm). The crowns of the molars were enlarged and had several small extra cusps.

Radiographic examination showed the third molars to be more than twice normal size with a multicusp form resembling a walnut kernel. There were no signs of roots. The second molars had large multitubercular crowns and single conical roots. Lower second molars were impacted. The first molars all showed root resorptions. The second premolars were impacted and exhibited multituberculism. The second premolar had a central cusp. The first premolars had irregularities in the palatal part of the roots, resembling pulp invaginations.

In summary, multiple macrodontia with especially giant third molars was combined with multituberculism, single conical molar roots, pulp invaginations and evaginations in premolars, and peak-shaped canines.

CASE REPORT

A 12-year-old Chilean girl has been treated for 2 years in the Oral Surgery Service of Consalud, Santiago, Chile and, during the year 2000, was noted to have malocclusion with maxillary protrusion and large molars by one of us (RF). According to information provided by the mother, she was delivered by cesarean section, born the first of two children of normal healthy parents. The second child, also a girl, was examined and had normal dentition and maxillary relationships. The mother was of short stature with normal teeth. The father was not available for examination but was stated to be normal. Consanguinity was denied. General physical examination revealed normal height and weight. During clinical examination, all incisors (both upper and lower) were noted to be shovel-shaped (Fig. 1). The premolars had evaginations and multiple tubercles. The molars had large crowns also with multiple tubercles and thin roots.

Radiographic examination showed a radiolucent area, well delimited to the left mandibular third molar zone, between the second and third molars, measuring approximately 10 mm in diameter. This was biopsied and demonstrated myxomatous tissue with some odontogenic epithelial rests. The second upper and lower molars presented single conical roots. The first molars,

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Fig. 1. All incisors had shovel-shaped form. The premolar teeth had evaginations and multiple tubercles.

upper left and lower right, exhibited extensive carious lesions and were extracted as well as the right lower third molar (Fig. 2). These teeth were sent for histopathologic analysis.

HISTOPATHOLOGY

The soft tissues demonstrated myxomatous changes with small vessels and inactive odontogenic epithelial rests that represented hyperplastic dental follicles.

The molars presented similar images. Both were calcified. On the crown were demonstrated islands of reduced enamel epithelium with a prominent inner layer with large waves in the enamel–dentin junction (Fig. 3). The predentin area in the roof of the pulp chamber was absent and with flattened odontoblastic layer (Fig. 4), but in the radicular area and the lateral aspects of the pulp chamber the layer of predentin was present with normal odontoblasts.

DISCUSSION

Reichart and Treadan [1977] briefly reported a similar finding in a 15-year-old male patient. This descrip-

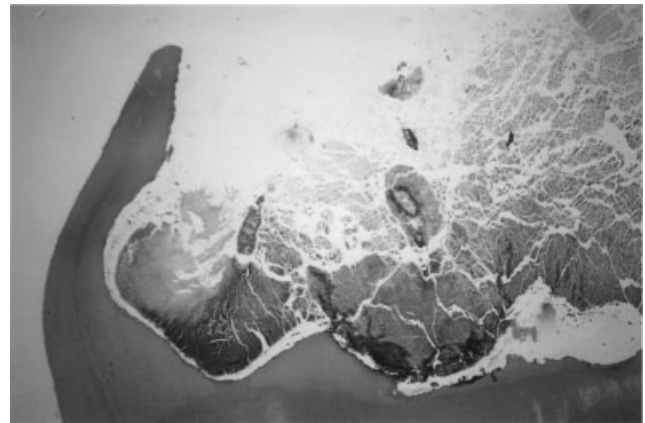


Fig. 3. Photomicrograph showing islands of reduced enamel epithelium with prominent inner layer.

tion was expanded upon the following year [Reichart et al., 1978]. Their patient was mentally retarded. The mother stated that the primary dentition had been normal. The mother and father and three sisters of the propositus were without dental anomalies. Radiologically, the findings noted in the Ekman-Westborg and Julin [1974] case were extremely similar to those reported by Reichart et al. [1978]. Mann et al. [1990] reported similar anomalies in the deciduous teeth of 5-year-old child from the 15th century. The findings were unearthed in 1968 in burial pits in Charles City County, Virginia. It was ascertained that the skeleton was from a Chickahominy family. The Chickahominy was an agricultural tribe of the Algonquian-speaking peoples inhabiting the Eastern Tidewaters region of Virginia. The most striking dental findings were the macrodontic, crenulated (wrinkled) first permanent and second deciduous molars. There were also disproportionate buccal–lingual anterior and posterior occlusal crown measurements; large, extremely shovel-shaped maxillary adult incisors; three-rooted mandibular deciduous second molars; dens invaginatus; agenesis of adult maxillary canines; and poorly devel-



Fig. 2. Radiograph showing huge third molars, radiolucent area mesial to third molar, second molars with single conical roots.

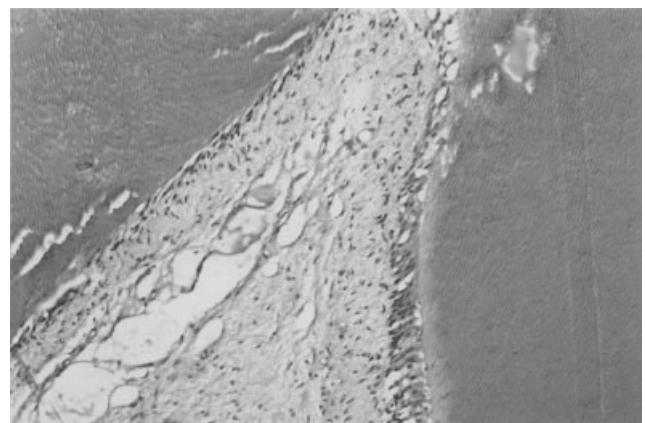


Fig. 4. Predentin area in roof of pulp chamber is absent, odontoblastic layer flattened. In radicular area and in lateral pulp chambers, the layer of predentin is present with normal odontoblasts.

oped enamel of deciduous first molars. This child was described as having a variant of the Ekman-Westborg and Julin condition.

Nagakawa et al. [1997] reported the case of a 15-year-old male with findings like those of Ekman-Westborg and Julin. In addition to general macrodontia, there were gigantic mandibular third molars. Other dental anomalies such as peak-shaped canines, central occlusal cusp, dens invaginatus, multituberculism, and single conical molar roots were present. The boy manifested anterior open bite and crowding. There was no contributory family history. He appeared to have multiple lentiginos on the left side of his chest and bilateral horizontal ocular flutter. Mild mental retardation was noted. No mention was made of involvement of deciduous teeth.

Ritzau et al. [1997] described another example in a 16-year-old Danish boy. The proband was short (10th centile) and bone age was approximately 2 years delayed. His face was noted to be overly long with a severely prognathic mandible and increased gonial angle, the midface being somewhat retruded. The palpebral fissures sloped downward laterally, and the ears were short, protruding, and posteriorly angulated. Mild contractures were noted at the proximal interphalangeal joints of the second, third, and fourth fingers of both hands. It had been noted in the history that when the child was approximately 4 years of age, the deciduous molars were stated to be somewhat larger than normal. The mother and son described by Yoda et al. [1998] cannot be accepted as examples of molar macrodontic multituberculism. The son had gemination of the right and left central incisors. All incisors exhibited extremely shovel-shaped crowns and molars. The premolars had multidiverticular occlusal surfaces. The mesial-distal widths of all teeth were large, and both mother and son had a complex odontoma. Less certain are the cases reported by Noriko et al. [2001]. Both the unrelated 11- and 13-year-old males had macrodontia and enamel hypoplasia of second deciduous molars and permanent successors. The incisors were markedly shovel-shaped. The deciduous and perma-

nent molars had underdeveloped cusps and appeared to have crenated occlusal surfaces. Delayed bone age was noted.

There does not appear to be a genetic aspect to this dental anomaly. All are isolated cases and there is no parental consanguinity. Various non-dental findings appear to be aleatory. Until our report, all patients were males but, in view of our female patient, this appears likely due to chance.

The histopathologic changes of flattened odontoblastic layer in the roof of the pulp, and the absence of predentin in this area, according to the best of our knowledge, has not been noted before. The incisors may be shovel-shaped, and the molars and premolars have large crowns with multiple tubercles and very thin roots. Due to the morphological dental changes, the patients usually present open-bite as in our case and in that of Reichart et al. [1978] or cross-bite [Ekman-Westborg and Julin, 1974], and mandibular prognathism [Ritzau et al., 1997].

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