Tooth abnormalities in congenital infiltrating lipomatosis of the face

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Objective. The aim of this study was to present a literature review and case series report of tooth abnormalities in congenital infiltrating lipomatosis of the face (CIL-F).
Methods. Four typical cases of CIL-F are presented. Tooth abnormalities in CIL-F documented in the English literature are also reviewed. The clinical and radiological features of tooth abnormalities are summarized.
Results. In total, 21 cases with tooth abnormalities in CIL-F were retrieved for analysis. Accelerated tooth formation and eruption (17 cases), macrodontia (9 cases), and root hypoplasia (8 cases) were observed in CIL-F.
Conclusion. Tooth abnormalities including accelerated tooth formation or eruption, macrodontia, and root hypoplasia are common in CIL-F. (Oral Surg Oral Med Oral Pathol Oral Radiol 2013;115:e52-e62)

Lipomatosis refers to a diffuse overgrowth or accumulation of mature adipose tissue, which can occur in various anatomical regions of the body including the trunk, extremities, head and neck, abdomen, pelvis, or intestinal tract.[1] Congenital infiltrating lipomatosis of the face (CIL-F) was first described by Slavin et al.[2] in 1983 with the following main characteristics: a nonencapsulated mass containing mature adipocytes; fat infiltration in muscles and adjacent soft tissue; absence of malignant characteristics; absence of lipoblasts; presence of fibrous elements and increased number of vessels and nerves; and adjacent bone hypertrophy or exophytic overgrowth.[3]

Facial structures from the inferior orbital rim to the angle of the mandible are regularly involved in this disease.[4] The parotid gland, tongue, cheek, lip, soft palate, masticatory muscles, and jaw bones tend to be frequently affected by diffuse fat tissue infiltration.[4-6] Oral mucosa neuromas also frequently occur in this disease.[4] Cosmetic deformities, oral malfunction, speech and sleep disorders, and subsequent psychological problems can occur.[4,6,7]

Although radiological manifestations of this disease have been documented in the literature, related tooth developmental disorders have not been systematically reviewed. Various tooth developmental abnormalities including accelerated tooth eruption, macrodontia, abnormal root shape, and early loss of deciduous or permanent teeth have been documented.[4-8] In this article, we report 4 additional typical cases and present a review of associated tooth developmental abnormalities in this disease.

Congenital unilateral enlargement of hemifacial soft and bone structures, together with dental malformations, has also been documented in hemifacial hyperplasia. The diagnostic distinction between these 2 congenital disorders has been imprecisely documented. Some authors have reported that hemifacial hyperplasia is related to increased fat cells in the soft tissue.[9,10] Therefore, we also present our opinions on these 2 disorders.

CASE REPORTS

Case 1

A girl with a congenital diffuse left facial mass was referred to our hospital at 5 years and 9 months of age. The patient had undergone a partial resection of the mass at 8 months of age through a preauricular and submandibular incision. Abundant subcutaneous fat tissue was found and pathologic examination showed the mass to be composed of mature fat cells. A diagnosis of diffuse lipoma was made at that time. The mass increased proportionally in size as the patient grew. Her parents reported that the permanent maxillary and mandibular left first molars erupted when she was 4 years old. Family history and blood tests revealed no unusual findings. Physical examination revealed a soft, nontender, noncompressible and ill-defined mass on the left face (Figure 1, A). Ipsilateral hemimacroglossia was detected and several small nodules were seen at the tip of the tongue (Figure 1, B). The left soft palate was also thickened with an irregular soft mass. Early eruption of numerous permanent teeth was noted, including the permanent maxillary left first premolar and first molar and the permanent mandibular left first molar. No hypoplasia of the enamel was noted in existing primary or

Supported by the National Natural Science Foundation of China (30901680).
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Received for publication Apr 17, 2012; returned for revision June 28, 2012; accepted for publication July 9, 2012.

2212-4403/$ - see front matter
http://dx.doi.org/10.1016/j.oooo.2012.07.433

Fig. 1. Clinical photograph shows hemifacial enlargement on the left side (A). Intraoral view of the patient shows hyperplasia of the left hemitongue (B).

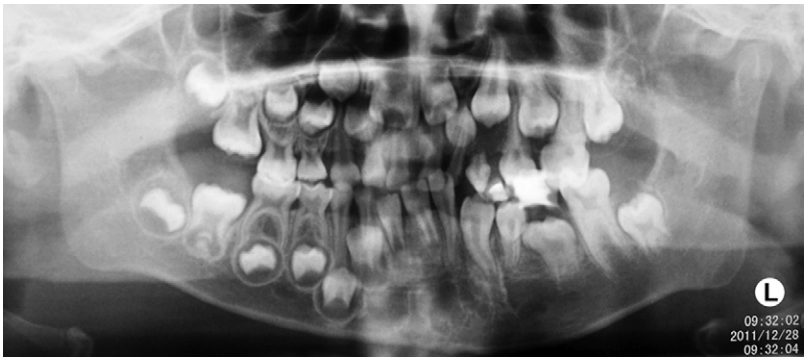

Fig. 2. Panoramic radiograph shows generally accelerated root formation (23-27, 31-37) and eruption (24, 26, 31-37) on the left side. Early calcification of 28 could be observed, whereas 18 is not seen. Macrodontia could be observed in 26 and 36, in which the roots are elongated and the apices have almost completely formed. Age-appropriate primary dentition is observed on the right side.

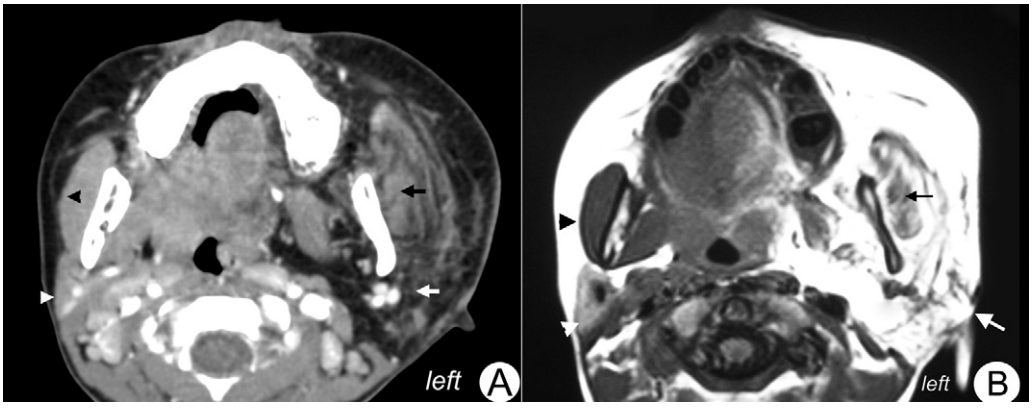

Fig. 3. Axial computed tomography (CT; A) and magnetic resonance imaging (MRI; B) show abundant fat tissue infiltrating the left masseter (black arrow), parotid gland (white arrow), pterygomandibular, and parapharyngeal spaces. Also note the normal appearance of the masseter (black arrowhead) and parotid gland (white arrowhead) on the right side. The fat tissue could be determined by its specific CT attenuation (−100 and −80 Hu before and after enhancement (A). Fat tissue infiltration observed on MRI shows high signal intensity on the T1-weighted image (B).