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Case report

Parry–Romberg syndrome with rare maxillofacial deformities: A report on two cases

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ABSTRACT

Parry–Romberg syndrome, also known as progressive hemifacial atrophy, is a rare developmental disorder characterized by progressing unilateral facial atrophy slowly, which may affect the skin, fat, muscle and bone. It can also be associated with different systemic manifestations and deformities. In this article, we present the two cases with Parry–Romberg syndrome. Of them, one has additional and rare facial deformity with rare facial cleft and the other has a special tongue feature.

1. Introduction

Parry–Romberg syndrome, first reported by Parry in 1825 (Parry, 1825) and subsequently described as a syndrome by Romberg in 1846 (Romberg, 1846), is a rare disorder characterized by slowly progressing unilateral facial atrophy (Moore et al., 1993; Longo et al., 2011). However, it has been reported that 5%–10% of cases present the bilateral progressive atrophy (Lakhani and David, 1984; Miller et al., 1987; Tollefson and Witman, 2007). The etiology and causes of its incidence remain unclear thus far. This syndrome generally occurs in the first or early second decade of life. Female-to-male incidence is approximately 3:2 (Miller et al., 1987). The deformities associated with Parry–Romberg syndrome vary widely, such as age of onset, duration, and the systems involved.

Common clinical manifestations of Parry–Romberg syndrome include atrophy of the skin, soft tissues, muscles, and underlying bony structures. In the craniofacial region, it may involve the paramedian forehead and scalp, cheek, and lateral aspect of the chin. It can result in ipsilateral enophthalmos with lower lid atrophy and deviation of the mouth and nose toward the affected side (Miller and Spencer, 1995). Deficiencies of the soft and hard palates in all dimensions, shortness and deficiency of the mandibular body and ramus, hemiatrophy of the tongue, delayed tooth eruption, root atrophy, and retarded tooth formation may also be observed in patients with Parry–Romberg syndrome (Foster, 1979; Glass, 1964). In addition, it has been reported that Parry–Romberg syndrome is occasionally associated with homolateral segmental vitiligo (Creus et al., 1994), hyperpigmentation (Pinheiro et al., 2006), port-wine stain, lupus profundus, morphea and linear scleroderma en coup de sabre (Menni et al., 1997). In our department we encountered two unusual cases of Parry–Romberg syndrome not previously described in the literature. One presented with a rare facial cleft and the other with special tongue manifestations. The Ethics Committee of our hospital approved the study, and written informed consent was obtained from the patients' parents.

2. Case reports

2.1. Case 1

In 2009, a well-behaved 10-year-old boy with normal mental development arrived at our clinic for the evaluation of facial abnormality. There was no consanguinity between the parents and there were no congenital abnormalities in family members. Although his mother's pregnancy was full-term and she had a normal delivery, he was affected by facial clefting. When he was 3 years old, his maxillary deciduous lateral incisor on the right side was lost for no apparent reason. Soon after, atrophy of the homolateral upper lip began, followed by gradual atrophy of other ipsilateral facial areas, including soft and hard tissues. The entire atrophy process lasted almost 6 years, until he reached the age of 9 years. The diagnosis of Parry–Romberg syndrome with a Tessier No. 3 facial cleft was made. At the time of consultation, the atrophy process had been stable for almost one year. Physical examination revealed that the upper lip on the right side was atrophied just apart from the medial line and the ipsilateral corner of the mouth was drawn upward. The skin and subcutaneous tissues on the right part of the chin were affected. The

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ala base on the right side was involved and superiorly displaced. The right side of the face was smaller than the left side (Fig. 1A). Intraoral examination showed that the cleft palate had been repaired but the alveolar cleft on the right side remained (Fig. 1B). CT scan and 3D reconstructive CT imaging showed skeletal deformities including an alveolar cleft, a defect of the ipsilateral hard palate, cleft between the maxilla and zygoma, and cleft between the nasal bone and nasal process of the maxilla (Fig. 1C). Three-dimensional images showed dysplasia in the homolateral bodies of the maxilla, zygoma, and maxillary sinus (Fig. 1D). The mandibular body and ramus were smaller on the affected side than on the normal side. There was a deep sigmoid notch on the affected side.

2.2. Case 2

In August 2011, a 10-year-old boy presented to our clinic with facial abnormality and severe right facial atrophy. His parents recalled no special problems, except that the right side of his face exhibited some redness at birth. The pregnancy and delivery had been normal. He was the first child of his healthy parents with no congenital abnormalities. At 4 years of age, the atrophy began on the right side of his face, gradually becoming severe. Based on clinical and radiologic findings, the diagnosis of progressive hemifacial atrophy was made when the patient was 6 years of age. When he was 9 years old, the progress of dystrophy was nearly stable. His parents reported that no additional atrophy had occurred for almost one year. In 2011, our first physical examination revealed severe unilateral facial atrophy, including the skin, soft tissues, muscles, and underlying bony structures, as well as hyperpigmentation on the buccal area on the same side, ipsilateral enophthalmos, and deviation of the mouth and nose toward the affected side (Fig. 2A). Unfortunately, the initial consultation, intraoral examination was not conducted and no treatment was administered because of parental concerns.

In September 2012, the patient presented again for consultation and treatment. There had been no further change in his face (Fig. 2B). However, an intraoral examination revealed delayed tooth eruption, ipsilateral reduction in height and width of the maxilla and mandible, and malocclusion on the affected side. He also presented with atypical tongue atrophy, which was only located on the posterior part of the tongue on the affected side. There were deep, distinguishing grooves between unaffected and affected parts of the tongue. The tongue mucosa of the atrophy-affected part was white, and fungiform papilla had caused congestion and swelling, giving the appearance of a “strawberry tongue” (Fig. 2C). After obtaining parental consent, we prepared a step-by-step treatment plan, including autologous fat transplantation for the atrophied areas to improve the soft tissue situation until he was 18 years old, at which time orthognathic surgery and orthodontics would be used to correct his skeletal deformities and malocclusion. Thus, at this time, the first autologous fat transplantation was performed on his forehead and right buccal regions.

In January 2013, he returned for a follow-up. The transplanted fat survived well, and the facial contour was improved (Fig. 2D). In addition, we observed that the entire tongue had atrophied on the affected side (Fig. 2E). No further treatment was administered,

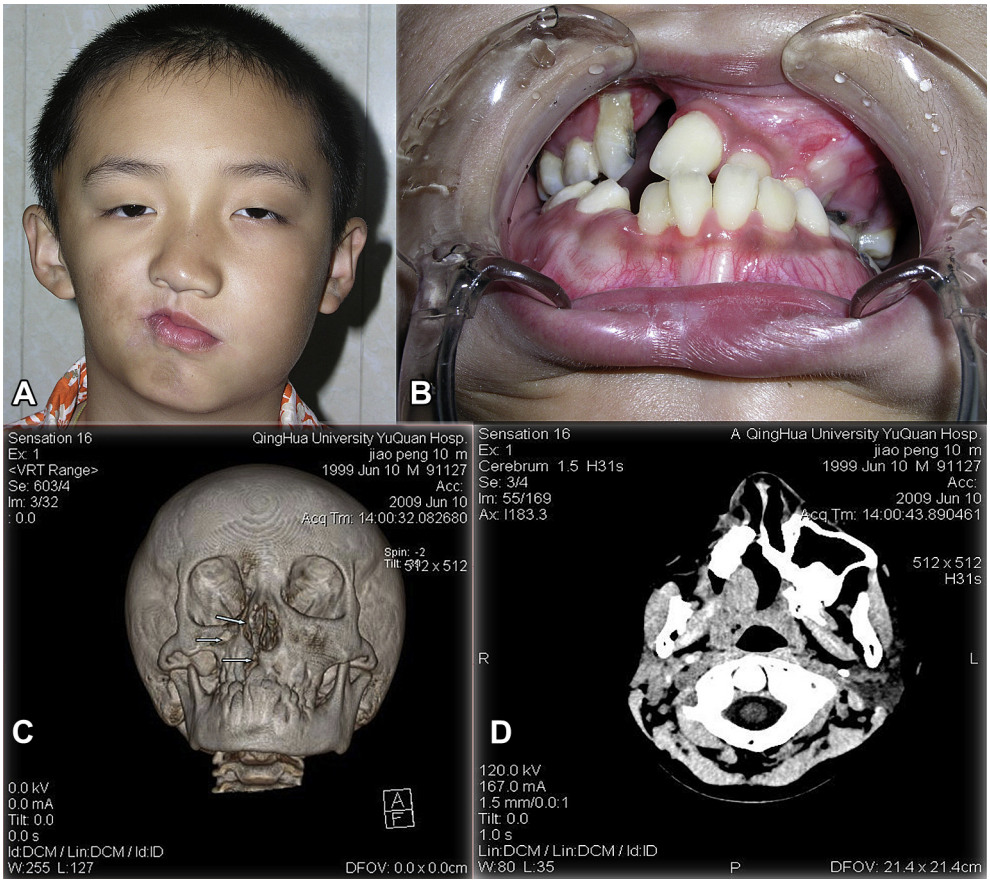

Fig. 1. Case 1: A 10-year-old boy suffering from Parry–Romberg syndrome with Tessier No. 3 facial cleft. A: Front view; B: Intraoral examination showing cleft alveolar and occlusal disturbance; C: Skeleton deformities shown by 3D-CT, including alveolar cleft, defect of the ipsilateral hard palate, cleft between the maxilla and zygoma, cleft between the nasal bone and nasal process of the maxilla (white arrows). The bodies of maxilla, zygoma and maxillary sinus were dysplasia in the three dimensions; D: CT scan showing dysplasia of the body of maxilla and maxillary sinus with palate defect.