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Parry–Romberg syndrome: a report of the dental findings in a child followed up for 9 years

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Summary. *Introduction.* The incidence of Parry–Romberg syndrome (PRS) is very rare.
Case Report. A case report is presented highlighting the main dental aspects that include delayed eruption, root resorption, dilacerations, and a reduction in the height and width of the ramus and body of the mandible on the affected side.

Introduction

Parry–Romberg syndrome (PRS; OMIM %141300) is a rare disorder characterized by slowly progressive self-limited degeneration of the soft tissues of half of the face (hemifacial atrophy) including dermis, subcutaneous tissue, fat, cartilage, and, sometimes, bone [1–3,4]. Bilateral manifestation has been reported in 5 to 10% of cases [5].

Incidence is approximately one or two cases per million people with 772 cases reported before 1964 [5,6]. The disorder has a male to female incidence of 3:2 [6].

Parry–Romberg syndrome was described first by Parry [7] and then Romberg (1846) [8]. Histologically, an increase in collagen fibres is observed.

Aetiology is not well understood. Suggested theories include trauma, infections, genetic factors, peripheral and trigeminal neuritis, lymphocytic neurovasculitis, localized scleroderma, endocrine disturbance, autoimmunity, and heredity. None of these has been proven [9,10,11]. The occurrence of PRS in one of two identical twins makes the possibility that hereditary factors are involved in its aetiology unlikely [12].

Symptoms and physical findings associated with PRS usually become apparent during the first decade or early during the second decade of life [13]. In some cases, the disorder is apparent at birth. The degree of facial deformity is usually more severe if atrophy begins in the first decade, as growth is rapid during this time.

Extraoral manifestations are facial asymmetry resulting in facial deformation and difficulty with mastication [14,15]. The skin overlying affected areas may become hyperpigmented with patches of vitiligo. The wasting process extends from months to years during which time the skin becomes dry and thin but freely movable. Atrophy of underlying muscles, bones, and cartilage is responsible for the typically aged appearance of the patient. Blanching of the hair or bald patches on the scalp and loss of eyelashes and eyebrows may occur [16]. The ear can be misshapen, smaller than normal, or bat-eared as a result of tissue atrophy [17]. Anterior to posterior growth can be altered by deviation of the entire middle and lower third of the face to the affected side, carrying the nose and chin with it [18]. The mouth may be drawn upward because of skin and subcutaneous tissue atrophy and often deviates to the affected side. When the lips are involved, they can be showing a unilateral dentition.

Intraoral manifestations: soft tissues (atrophy of the upper lip and tongue) [18]. The soft palate may also be deficient on the affected side [19]. Intraoral soft tissues and muscles of mastication can be affected, but they usually function normally. A decreased depth and width of the retromolar region of the pharynx may occur.

Hard tissues: delayed eruption, missing teeth, deficient root development, or resorption of the roots of teeth on the affected side have been reported [15,20–22]. The teeth appear clinically normal, have regular enamel, dentin, cementum and pulp, and test vital [18]. The mandible and alveolar ridge may be

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smaller on the affected side also [19,23]. The mandibular body may be shorter than normal, the ramus can be deficient vertically, and there can be a delay of mandibular angle development.

Spontaneous fracture of the mandible has also been reported [18,24,25]. The jaw disturbances can result in a unilateral malocclusion on the affected side and deviation of the facial and dental midlines [18,25].

Some individuals may also experience associated neurological abnormalities, which may last for extended periods of time.

There are less than six reports on the dental effects of PRS from countries such as America, New Zealand, and India. Following a review of the literature, we did not find any reports of dilacerations in patients with PRS.

Case report

A 7-year-old female patient presented to the University Dental School and Hospital, Cork, Ireland. Her mother was concerned about an area of skin depigmentation and the reduced height of the left side of her face and the unerupted 26 and 36. She had no relevant medical history and was not on any medications. There was no significant family history.

The initial extraoral examination at age 7 years revealed a reduced height of the left side of her mandible and an area of skin depigmentation on the left side of her face. An intraoral examination revealed some atrophy of the left angle of her mouth and left side of the tongue. She had delayed eruption of her upper and lower permanent first molars on the left side. A three-dimensional CAT scan was performed that showed a short mandible on the left side but no evidence of hemiatrophy at this time. She had a restoration placed on 85 and fissure sealants placed on 16 and 46. An Ortho Pantomo Gram (OPG) was taken that showed the presence of all her primary teeth and the development of all her permanent teeth except for 18, 28, 38, and 48. The mandible was short on the left side and there was a reduced height of the left ramus compared to the right side (Fig. 1).

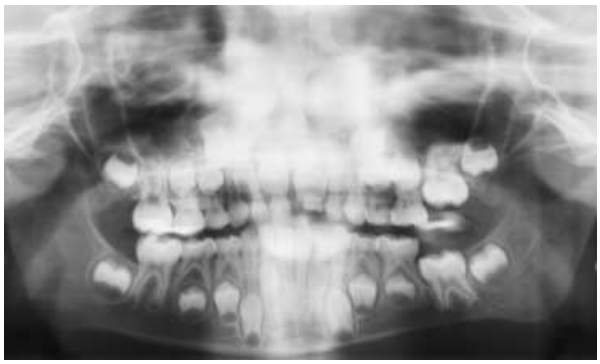

Fig. 1. Patient is 7 years old. OPG shows unerupted upper and lower left first molar teeth, whereas those on the right are fully erupted. The height of the ramus is reduced on the left side compared to the right.

She was regularly reviewed in the Dental Hospital in Cork, where she attended the Paediatric, Orthodontic, and Maxillofacial clinics to monitor her dental and skeletal development and her condition. Her general well-being and ability to cope with her condition was monitored. This patient and her parents were offered psychological support as required to enable them to cope with and understand PRS.

At 9 years of age, she had a full orthodontic assessment by a consultant orthodontist who was satisfied with her dentofacial development at that time. She was reviewed every 6–12 months.

At 10 years of age, extraoral examination revealed marked progression of her facial atrophy. The skin was thin and the subcutaneous tissues, fat, and cartilage were reduced on the left side of her face. On intraoral examination 26 and 36 erupted at 9·5 years and were fissure sealed at aged 10 years. An upper anterior occlusal and OPG were taken to locate 13 and 23 that confirmed the position of 13 to be buccal and 23 to be palatal. It was planned to extract 53 and 63; the latter was extracted at 10 years of age but the patient was reluctant to have 53 extracted. Both upper canines erupted at 11·5 years without further treatment. The OPG at age 10 years showed a marked reduction in the vertical height and width of the ramus and a reduced width and length of the body of the mandible. There were marked dilacerations of the roots of 24 and 34 and the root of 35 appeared very short (Fig. 2).

The patient undertook a full programme of psychological and social support beginning at 10 years of age and continuing until 12 years.

She was reviewed recently at age 15 years. An extraoral examination showed extensive atrophy of the skin, subcutaneous tissues, fat, and cartilage on the left side of her face (Fig. 3). An intraoral examination revealed unerupted upper and lower left second

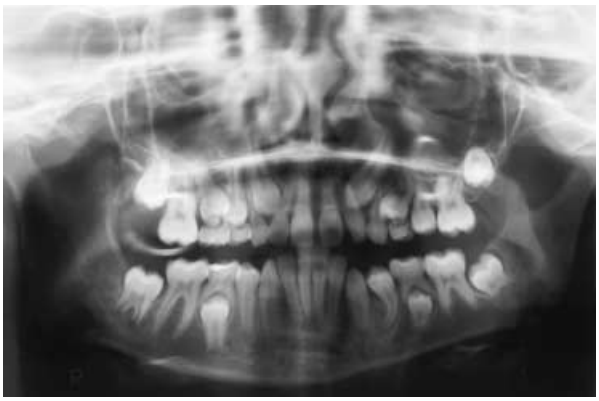

Fig. 2. Patient is 10 years old. OPG shows marked dilacerations of the roots of the upper and especially the lower left first premolars and the root of the lower left second premolar is short. The vertical height and width of the ramus are considerably reduced on the left side. There is also a marked reduction in the vertical height of the body of the mandible.

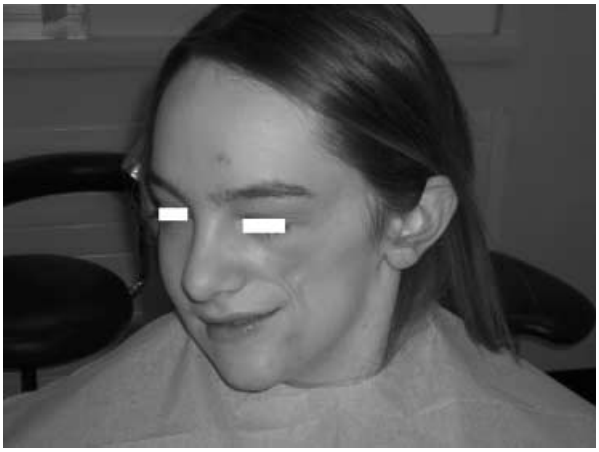

Fig. 3. Fifteen-year-old patient with Parry–Romberg syndrome. Note the atrophy of the soft tissues of the left side of the face. The right side of her face is normal.

molars, whereas those on the right were fully erupted. An OPG taken that showed resorption of the roots of the upper left first and second premolars and first and second molars and also of the lower left second premolar and first and second molars (Fig. 4). The roots of the teeth on the right side are normal in comparison. There is a gross reduction in the vertical height and width of the ramus on the left side and a reduction in the height of the body of the mandible. There is no evidence of development of a lower left wisdom tooth on the OPG.

She is now 15 years of age and her condition appears to have stabilized.

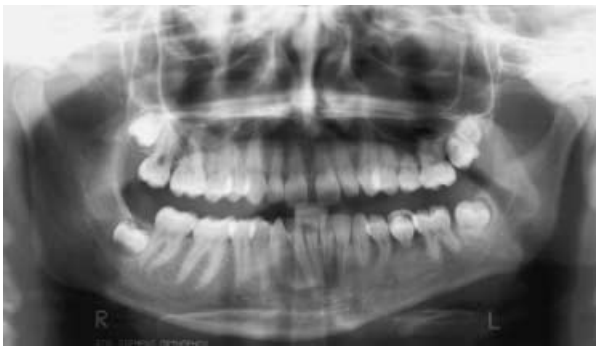

Fig. 4. Patient is 15 years old. OPG shows dilacerations of the roots of the upper and lower left first premolars and marked root resorption of the upper and lower left first and second premolars and the first and second molars. Note also unerupted upper and lower left second molar teeth. The lower left wisdom tooth appears to be absent. There is a significant reduction in the vertical height and width of the ramus and body of the mandible on the left side.

The patient previously described was referred to a plastic and maxillofacial surgeon in the USA where she underwent microvascular free tissue transfer from her left back and flank to the left side of her face in August 2005 and will be reviewed for revision as necessary.

Discussion

This case has several dental similarities when compared with other reports. Extraorally, there is significant atrophy of the skin, subcutaneous tissues, fat, and cartilage on the left side of her face, which was first noticed at age 6 years and now seems to have burned out following her pubertal growth spurt.

The left ramus and body of the mandible are reduced in height and width, and the left angle of the mandible is developmentally delayed. This may be due to pressure from muscles and soft tissues.

There was evidence of retarded eruption of the upper and lower first and second molars. The upper and lower left first molars did not erupt until 9·5 years of age and the upper and lower second molars have not yet erupted; the patient is now 15·5 years old. The erupted teeth on the left side had normal enamel and dentine clinically and there was no evidence of ankylosis on percussion.

There was moderate root resorption of the roots of the upper and lower premolars and molars on the left side, whereas the roots of all the teeth on the right side were of normal length. The aetiology of root

resorption includes trauma, excessive mechanical forces, periapical inflammation, tooth impaction, tumours and cysts, and idiopathic resorptions [24,26]. This patient, however, had no history of any of the above causes of resorption. It can be seen on the OPG that the periodontal ligament is intact, and also there was no evidence of ankylosis when the teeth were tested using the percussion test. Root resorption has been reported in other case reports of patients with Parry–Romberg syndrome [27]. It is possible that the cementum formed around the roots of the teeth may be abnormal and thus reduces the resistance of these teeth to resorption [28–30]. This process may be exacerbated when local factors such as gingival inflammation exist. It is noteworthy that the active stages of hemifacial atrophy coincide with the period of root formation of permanent teeth.

Interestingly, there is marked dilacerations of the roots of the upper and lower left first premolars. It is our opinion that these dilacerations may be caused by pressure from the muscles and soft tissues on the alveolar bone during root development. The most common cause of dilaceration is trauma such as a fall leading to intrusion of primary teeth or extraction of a primary tooth causing damage to the follicle of the permanent successor [31,32]. Other causes include a cyst, tumour, odontogenic hamartoma, or an idiopathic cause. There is no history of any dental trauma in this patient or of any of the above conditions. As can be seen from the OPG, the primary molars were present until they exfoliated. We could not find any other reports of dilacerations in patients with PRS following our review of the literature.

This patient has had intermittent episodes of severe migraine-type headaches, which are typical in patients with PRS. She does not have any other neurological symptoms, however, which include visual abnormalities, enopthalmus, nausea and vomiting, epilepsy, and trigeminal neuralgia. The range and severity of associated symptoms vary from case to case [9,33–35].

Treatment options include orthodontics to attempt to minimize the progressive atrophy and to attempt to maintain parallelism of the facial planes [36], mandibular distraction [37], and maxillofacial and plastic surgery [38].

She was referred to a plastic and maxillofacial surgeon in the USA for assessment. She underwent microvascular free tissue transfer from her left back and flank to the left side of her face in August 2005 and will be reviewed for revision as necessary.

Patients with PRS may have social or psychological difficulties. They may require referral to a medical social worker or psychologist for assessment and a programme of support. These professionals can work with patients and parents and help them deal with any questioning from their peers and other adults by using role-play techniques. This will help to build on their self-esteem. Patients may have such sessions on their own and some with their parents present. These sessions provide great benefits and encourage their development and help them become confident and outgoing people. It is also important to ensure that parental support is maintained.

What this paper adds

- Dilaceration of roots may be associated with PRS which has not been previously reported in the literature.

Why this paper is important to paediatric dentists

- It is important for paediatric dentists to recognise early signs of PRS and refer appropriately.
- Multidisciplinary support is required.
- Optimal dental care needs to be achieved.

Conclusion

It is evident from this case that a multidisciplinary team approach is required to treat these patients. The team may include a general dental practitioner, paediatric dentist, orthodontist, medical social worker, psychologist, general medical practitioner, paediatrician, maxillofacial surgeon, and plastic surgeon.

It is important that she would have regular dental reviews to monitor dental development and for evidence of progression of root resorption. It is necessary that optimum oral hygiene be maintained to preserve her dentition. Should resorption progress, the left upper and lower premolars may be lost eventually. If this occurs, implants may be considered if sufficient bone is present or bone grafting in conjunction with implants. It is of paramount importance that these patients are provided with psychological support when enormous facial changes are occurring to enable them to cope emotionally.

We would like to suggest that patients with Parry–Romberg syndrome should be assessed for root resorption and dilacerations. There has not been any reference to dilacerations in previous literature and this may be where patients have not had a dental assessment.

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