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A case report of Parry Romberg Syndrome initially presenting as periodontitis.

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Complete List of Authors:	Taylor, Greig; Newcastle University, School of Dental Sciences Culshaw, Shauna; University of Glasgow Dental School, Periodontology Armas, Jose; University of Glasgow Dental School, Periodontology Savarrio, Lee; University of Glasgow Dental School, Restorative Dentistry Goodall, Christine; University of Glasgow Dental School, Oral Surgery
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Title: A case report of Parry Romberg Syndrome initially presenting as periodontitis.

Running Title: Case Report: Parry Romberg Syndrome

Keywords: *Parry Romberg Syndrome; Asymmetry; Hemi-facial atrophy; periodontitis*

Greig Taylor[1, 2], Shauna Culshaw[1], Jose Armas[1], Lee Savarrio[1] and Christine Goodall[1]

1. Glasgow Dental Hospital & School, College of Medical, Veterinary and Life Sciences, 378 Sauchiehall Street, University of Glasgow, Glasgow, G2 3JZ
2. Current address: School of Dental Sciences, Newcastle Dental Hospital, Richardson Road, Newcastle Upon Tyne

Mr Greig D Taylor

Academic Clinical Fellow/StR in Paediatric Dentistry, School of Dental Sciences, Newcastle Dental Hospital, Richardson Road, Newcastle UponTyne

Phone: 0191 233 6161

Email: Greig.taylor@ncl.ac.uk

Dr Shauna Culshaw

Senior Clinical Research Fellow/Hon Consultant in Periodontology, University of Glasgow

Dr Jose Armas

Consultant in Periodontology/Hon Senior Clinical Lecturer, University of Glasgow

Mr Lee Savarrio

Consultant in Restorative Dentistry/Hon Senior Clinical Lecturer, University of Glasgow

Dr Christine Goodall

Senior Clinical Lecturer/Hon Consultant in Oral Surgery, University of Glasgow

Abstract

Parry Romberg Syndrome (PRS) is a rare disorder of progressive hemi-facial atrophy, involving soft tissues, fat and occasionally bone. It can co-exist with presentations of Morphea. We describe an unusual case of persistent periodontal and alveolar destruction associated with PRS. A 56-year-old African female initially presented with persistent periodontal destruction, which showed minimal response to conventional periodontal treatment. Following non-surgical treatment, surgical debridement and extraction of the two right maxillary incisor teeth was required to halt the periodontal destruction. Atrophy was not limited to the periodontal tissues. Multi-disciplinary care and extensive investigations were required to diagnose PRS. Once the PRS has stabilised, adipose tissue transplants will be required to improve the facial appearance. We highlight the need for extensive investigations and a multi-disciplinary approach to diagnose rare systemic causes for recalcitrant periodontal disease.

Case Report

A 56 year old female of African origin presented with a persistent periodontal defect, which showed minimal response to non-surgical interventions and systemic antibiotic therapy.

The patient's medical history revealed nothing of relevance and she was generally fit and well. The patient was a non-smoker and consumed alcohol within recommended weekly limits.

Extra-oral examination highlighted a prominent right zygomatic arch **(Fig. 1)** with a fibrous scar-like lesion on the upper lip. On intra-oral examination there was a mucosal defect, approximately 1 cm in diameter adjacent to teeth 11 and 12, with full thickness gingival erythema and necrosis of the interdental papilla **(Fig. 2a & Fig. 2b).** Gingival recession in the area and a 9 mm periodontal pocket with bleeding and discomfort on probing was noted**.** Radiographic examination revealed a moderate angular bony defect associated with teeth 11 and 12 **(Fig. 3).**

There was no improvement following conventional non-surgical debridement. A swab was sent for microbiological analysis, which showed a mixed growth of *Streptococcus mitis, Streptococcus sanquis, Streptococcus constellatus* and *Gemella haemolysans*. Sensitivity to Amoxicillin of the these cultured strains were noted and a 5 day course of 500 milligrams, three times daily, was prescribed prior to any further non-surgical or surgical treatment. Upon completion of the systemic antibiotic treatment, a subsequent course of non-surgical periodontal therapy was ineffective in managing the periodontal defect. The site was surgically explored and necrotic bone and affected gingivae were biopsied. Primary closure with a coronally advanced flap was carried out. The biopsy results were non-specific with no

evidence of dysplasia or malignancy in either sample. Further deterioration over a six-week period merited further microbiological investigations, which revealed a mixed growth of *Streptococcus constellatus, Anaerococcus prevotii*, *Veillonella*, *Streptococcus oralis, Streptococcus sanguinis, Gemella amorbillorum* and *Lactococcus cremoris*. No further antibiotics were prescribed at this time.

Upon review, three months later, the fibrous scar-like lesion in the midline of her upper lip was still prominent **(Fig. 4).** The patient did not report any history of trauma or a cause for this prominence. It was felt that this lesion may be a feature of *lichen sclerosis et atrophicus* which is a chronic inflammatory dermatosis that results in white plaques with epidermal scarring and atrophy; however, a biopsy revealed non-specific keratosis. Recession and exposure of necrotic bone intra-orally continued to worsen **(Fig. 5).** The right maxillary incisor teeth were extracted along with further debridement of surrounding necrotic bone. Histopathological examination reported dense connective tissue with active inflammatory cell infiltrate; the incisor teeth were surrounded by necrotic bone colonised with abundant bacterial colonies, mainly of a filamentous nature, and a few fungal forms. No evidence of malignancy was noted. A full blood count, haematinics (Serum Folate, Vitamin B12, and Serum Ferritin) and blood glucose tests were normal. An anti-neutrophil cytoplasmic antibodies (ANCA) test was negative. A referral to Infectious Diseases and Tropical Medicine ruled out HIV and TB.

The patient was referred for assessment to the Maxillofacial Unit with a provisional diagnosis of chronic osteomyelitis. A CT scan of the head was normal and further surgical debridement of the affected area was carried out under general anaesthesia with primary closure. At follow up the area was healing well and histopathology reports indicated an area of chronic inflammation and was suggestive of osteomyelitis. The patient was placed on a long-term course of antibiotics and reviewed regularly. At subsequent reviews, intra-oral healing was

noted; however, there was evidence of progressive facial asymmetry affecting the right side **(Fig. 6).** Six months later the facial asymmetry had worsened and this was felt to represent a form of hemifacial atrophy or of localised scleroderma. A Rheumatology assessment was requested. The rheumatologist noted the *"*patient presented with right hemifacial atrophy, with and patches of Morphea on her back and right arm, but without overt skin change. The lesion on her upper lip may be of similar aetiology. No neurological abnormalities were detected clinically or on reviewing the CT scan. There were no strong immunological markers at present. The weakly positive antinuclear antibody (ANA) is non-specific and it is therefore difficult to reach a firm conclusion."

A diagnosis of Mixed Type Parry Romberg Syndrome with Morphea was made jointly by OMFS and Rheumatology based on the clinical findings observed in this case. Reconstructive surgery was not indicated as the patient was still in the active phase of this disease process.

. Adipose tissue transplants may be required in the future once the disease process stabilises. Similarly, fixed prosthetic options to replace teeth 11 and 12 will be delayed until the disease progress has halted. The patient was provided with a removable prosthesis **(Fig. 7).** The patient has been kept under regular review and has declined the option of reconstructive surgery.

Discussion

Parry Romberg Syndrome (PRS) is a rare disorder of progressive hemi-facial atrophy[1,2] (1,2) of soft tissue[1], fat[1,3] and, on occasion, bone[1,3-6] which was first described by Parry (1825) and then Romberg (1846) and known as a *trophoneurosis facialis*[7]. Presenting features of PRSare shown in **Table 1.**

PRS and localised scleroderma/*en coup de sabre* Morphea can be diagnosed as isolated disease processes. Morphea is an uncommon persistent dermatological condition where there are areas of thickened skin, occasionally involving underlying tissues, which may or may not have altered pigmentation. However, they have been known to co-exist, although the rationale for this co-existence is unclear within the literature[2,5-9]. In a small retrospective analysis of 54 patients, twenty-six patients (48%) had isolated *en coup de sabre* Morphea, thirteen patients (24%) had PRS while fifteen patients (28%) had both[9]. PRS has been described as a form of scleroderma that only affects the head and neck region[7, 10].

PRS is often associated with various systemic manifestations, particularly neurological, ophthalmological and orofacial[9]. PRS can often affect the central nervous system and be associated with epilepsy[9], trigeminal neuralgia, facial nerve palsy, mental disorders, learning difficulties and severe migraine headaches[1, 9]. Ophthalmic involvement occurs in 10% to 35% of cases[8] with progressive enophthalmos a frequently reported feature[11]. Less commonly reported ophthalmic features are pupillary disturbances, heterochromia, uveitis, pigmentary disturbances of the ocular fundus, and restrictive strabismus[11]. Orofacial changes usually affect the upper maxillary and nasolabial fold regions. Subsequent progression to the angle of the mouth, areas around the eye, the brow, and the neck are then usually observed[8]. The ear is often misshapen and smaller than normal in size[8].

Relatively few oral findings have been reported in the literature[8] with the salient Oral and Maxillofacial features of PRS being shown in **Table 2**:

PRS treatment aims to augment atrophied areas for aesthetic rehabilitation[6, 8,9,14] and provide symptomatic treatment for any concurrent neurological disorder[7]. The augmentation process is usually deferred until stabilisation of the disease process[6-8, 15] Augmentation is commonly

carried out with silicone and fat injections, to the atrophied area, to restore facial symmetry[6-8, 15]. Facial asymmetry can potentially be improved during the craniofacial growth period by stimulating those growth centres that are not directly involved in the wasting process with functional orthodontic appliance therapy in an attempt to maintain mandibular symmetry[7]. Prosthetic dental rehabilitation is often required for PRS patients following craniofacial augmentation. The timing of fixed prosthetic treatment is often delayed until the disease process has arrested with temporary measures being put in place during the active phase.

Conclusion

Parry Romberg Syndrome (PRS) is a rare disorder of progressive hemi-facial atrophy which affects soft tissues, adipose and osseous structures. To the best of our knowledge, this appears to be the first case reported of periodontal destruction as an initial presentation of PRS. PRS should be considered as one of the varied systemic causes for recalcitrant periodontal lesions.

Conflict of Interest

The authors confirm they have no conflicts of interest.

Figure Legends

Figure 1: Initial examination shows a prominent right zygomatic arch (*Published with the patient's consent)*

Figure 2a/b: Mucosal defect associated with upper right central and lateral incisor teeth and 12. Full thickness gingival erythema and associated necrosis of the interdental papilla was noted. Gingival recession in the area and a 9mm periodontal pocket. *(Published with the patient's consent)*

Figure 3: Periapical radiographs taken on presentation showing moderate angular defect associated with upper right central and upper right lateral incisor. *(Published with the patient's consent)*

Figure 4: The affected area on three month review showing the increased prominence of a scar-like fibrous band in the upper lip *(Published with the patient's consent)*

Figure 5: Review three months: Post–surgical review of the non-healing defect with exposed interdental bone exposure around tooth 11 and 12 *(Published with the patient's consent)*

Figure 6: Review nine months: Progression of facial asymmetry on the right hand side, particularly in the labial region. *(Published with the patient's consent)*

Figure 7: Provision of removable prosthesis to replace teeth 11(UR1) and 12 (UR2) *(Published with the patient's consent)*

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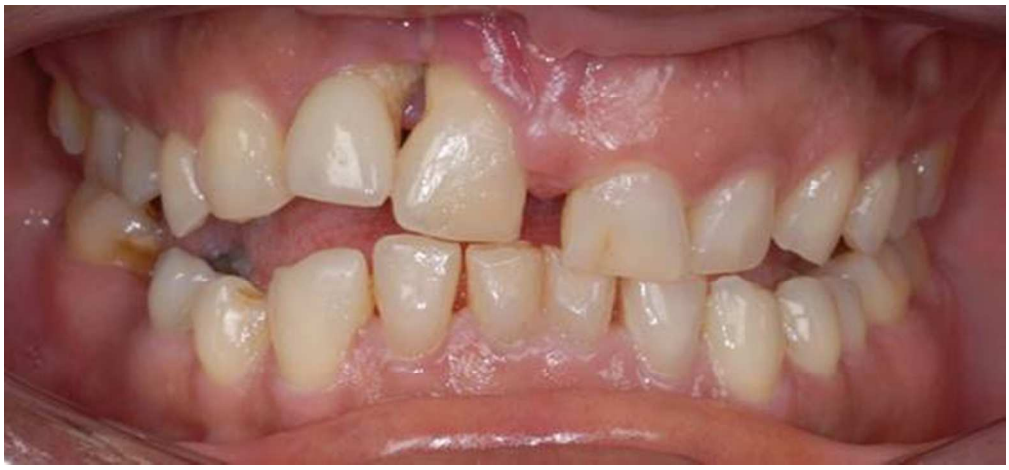
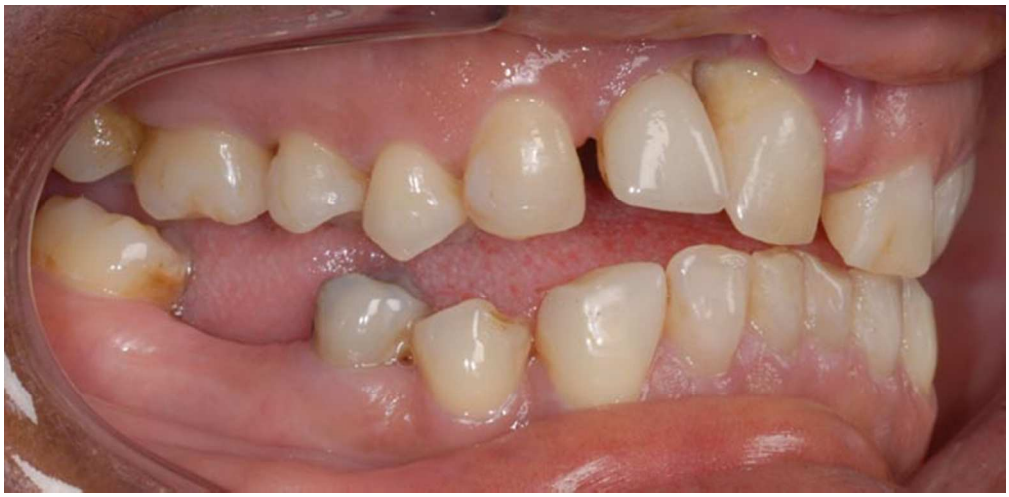
Initial examination shows a prominent right zygomatic arch (Published with the patient's consent)

69x103mm (96 x 96 DPI)



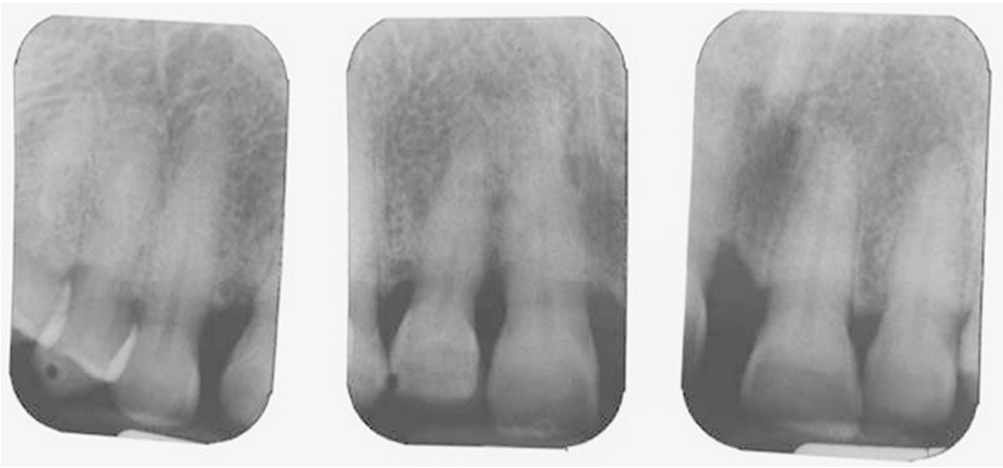
Mucosal defect associated with upper right central and lateral incisor teeth and 12. Full thickness gingival erythema and associated necrosis of the interdental papilla was noted. Gingival recession in the area and a 9mm periodontal pocket. (Published with the patient's consent)

160x73mm (96 x 96 DPI)



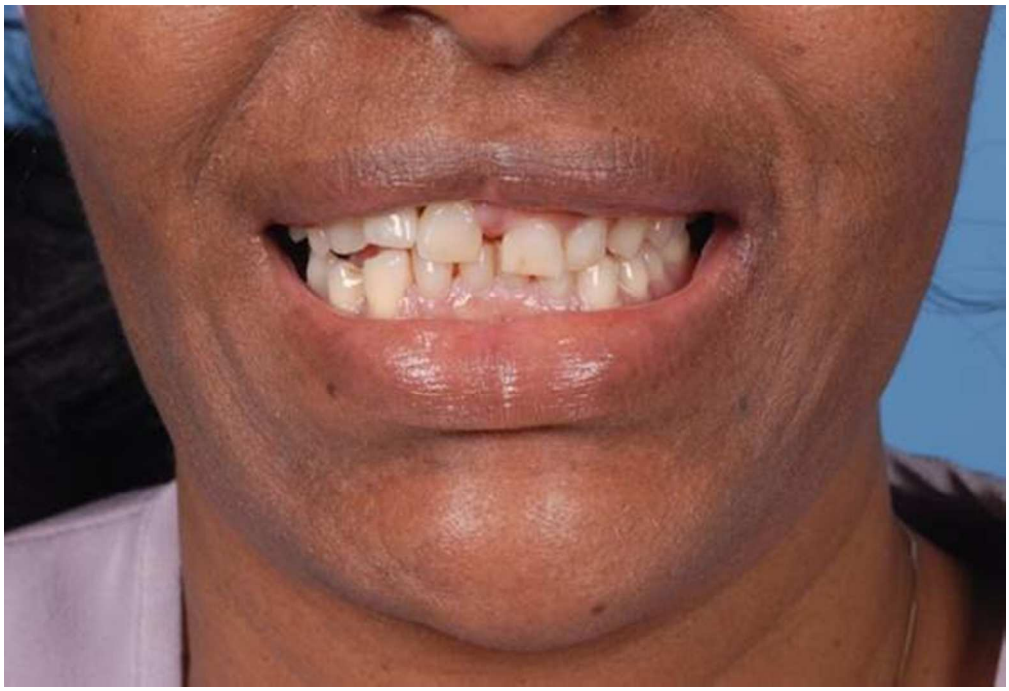
Mucosal defect associated with upper right central and lateral incisor teeth and 12. Full thickness gingival erythema and associated necrosis of the interdental papilla was noted. Gingival recession in the area and a 9mm periodontal pocket. (Published with the patient's consent)

59x28mm (300 x 300 DPI)



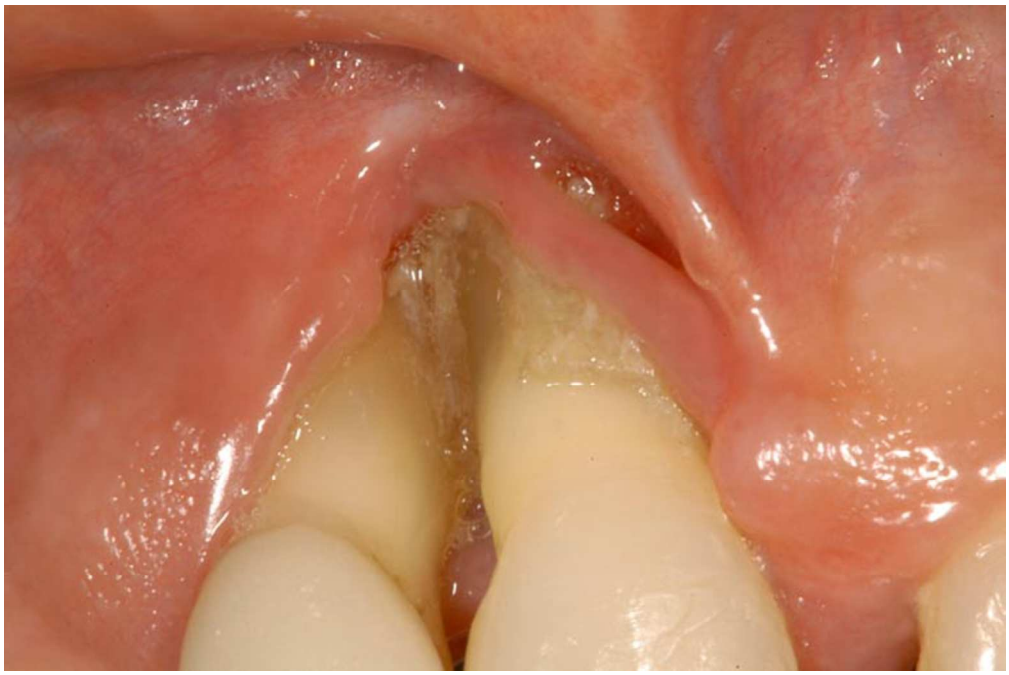
Periapical radiographs taken on presentation showing moderate angular defect associated with upper right central and upper right lateral incisor. (Published with the patient's consent)

159x73mm (96 x 96 DPI)



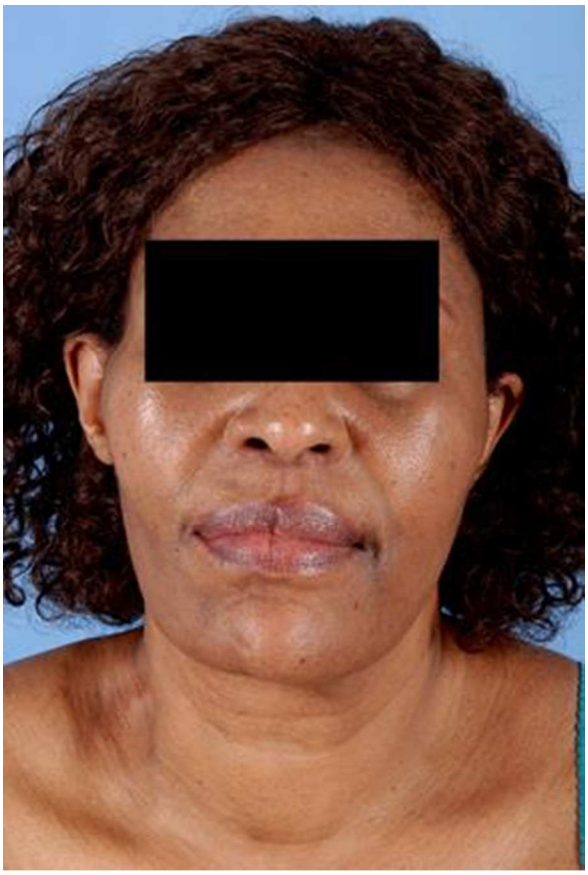
The affected area on three month review showing the increased prominence of a scar-like fibrous band in the upper lip (Published with the patient's consent)

157x107mm (96 x 96 DPI)



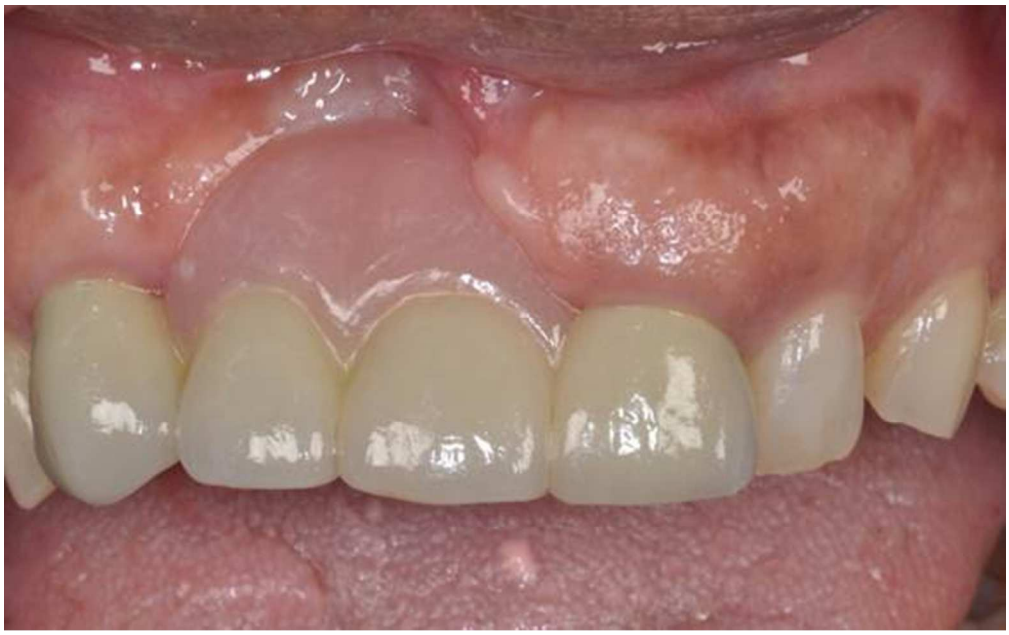
Review three months: Post–surgical review of the non-healing defect with exposed interdental bone exposure around tooth 11 and 12 (Published with the patient's consent)

59x39mm (300 x 300 DPI)



Review nine months: Progression of facial asymmetry on the right hand side, particularly in the labial region. (Published with the patient's consent)

76x114mm (96 x 96 DPI)



Provision of removable prosthesis to replace teeth 11(UR1) and 12 (UR2) (Published with the patient's consent)

153x95mm (96 x 96 DPI)

Table 1

Table 1: Presenting features of PRS
Of unknown aetiology, with a slow period of progression, ranging from 2 – 20 years before stabilising[6]
Often noted in the first decade of life with late onset having been reported[6]
More common in females[6,8]
No racial discrimination is noted[6]
Often occurs sporadically, though some familial cases have been reported[6]
Usually unilateral, often following the sensory innervations of one or all three branches of the trigeminal nerve[8]. However, it has been reported as bilateral in 5% -10% of cases[8].
May extend beyond the head and neck region to rest of the body on the ipsilateral side; however this is infrequently reported[9]
Often associated with dry, pigmented skin[9]

Table 2

Table 2: Oral and maxillofacial features of PRS
Deviation of the normal position of the oral cavity[8]
Unilateral malocclusion and deviation of facial and dental midline[6,8,9]
Atrophied Muscles of mastication; not usually impairing function[8]
Changes in tooth size on the affected side[11,12]
Delayed tooth eruption[6,8]
Crown and root dilacerations[6,8]
Root resorption[6,13]
Arrested tooth formation[6,8]
Odontogenic Cyst[6]
Odontome formation[6]
Facial concavity appearance due to deficiencies of intra-oral structures[8]
Poor growth of facial bones[2,6,8,9] which may lead to pathological fractures[8,14]