

## Paediatric Dermatology Column: Case Report

### A teenage girl with facial asymmetry

#### 一名面部不對稱的女孩個案

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A teenage girl with a history of right hemifacial atrophy of unknown cause initially presented with loss of hair. Then she developed a violaceous rash over her waist which was shown by skin biopsy to be morphea. Based on all the clinical features, the diagnosis of Parry-Romberg syndrome, also known as hemifacial atrophy was made.

一名女孩過去有著成因不明的右半側面萎縮，最初因脫髮而求診，及後在其腰間長出紫紅色皮疹，病灶皮膚活檢顯示為硬斑病。綜合以上所有臨床特點，其診斷確立為帕-羅二氏綜合症，又稱半側面萎縮。

**Keywords:** Hemifacial atrophy, morphea, Parry-Romberg syndrome

**關鍵詞：**半側面萎縮、硬斑病、帕-羅二氏綜合症

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#### Case history

A 9-year-old girl was referred for loss of hair for two years. It was non-itchy without any preceding symptoms of infection or inflammation. She did not have any precipitating life events or medications before the hair loss. She had a history of right hemifacial atrophy of unknown cause and short stature since the age of four, and

Hashimoto's thyroiditis on thyroxine supplement with a markedly raised microsomal antibody titre at the age of seven. She had a normal development with no history of seizures or any neurological abnormalities. Plain computed tomography of the brain, X-rays of the skull, mandible and spine were performed and were normal. She was followed-up at the paediatric clinic and genetics clinic.

On physical examination, her face was asymmetrical with an atrophic right cheek and mandible with overlying normal looking skin (Figure 1). There was a patchy non-scarring alopecia over the occiput in a band-like fashion (Figure 2). There was no erythema or any signs of infection over the scalp. There was no loss of eyebrow or eyelashes. The fingernails were normal.

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Topical steroid (0.1% mometasone furoate cream) daily was prescribed without much improvement over the patch of alopecia. Two years later, she complained of a slightly depressed non-itchy violaceous rash over her waist (Figure 3). A 3 mm punch skin biopsy was then done on the waist, which showed an epidermis with mild hyperkeratosis and acanthosis, thickened collagen bundles and a mild superficial perivascular lymphocytic infiltrate in the dermis, and atrophic eccrine glands high in the dermis (Figure 4). The histological features were suggestive of morphea.

In view of the combined presentation of morphea, hemifacial atrophy and band-like alopecia, the diagnosis of Parry-Romberg syndrome was made. Oral methotrexate was suggested but her parent declined and preferred conservative management.



**Figure 1.** Right hemifacial atrophy.

## Discussion

Parry-Romberg syndrome, also known as progressive facial hemiatrophy, was first described by Parry in 1825 and further characterised by Romberg in 1846. It is an uncommon disorder that usually develops in the first and second



**Figure 2.** Band-like non-scarring alopecia.



**Figure 3.** Violaceous rash at the waist.

decades of life. Due to its rarity, its demographics and clinical features are based on case reports and small case series only.

Parry-Romberg syndrome is a slowly progressive unilateral facial atrophy of the skin, soft tissues, muscles and underlying bone structures affecting one or more dermatomes innervated by the trigeminal nerve.

Other than facial asymmetry, patients often have other cutaneous features. Case reports and reviews show patients may have concomitant *en coup de sabre morphoea*, which is a linear scleroderma involving frontoparietal scalp or medial forehead.<sup>1,2</sup> In a case series of 12 patients with Parry-Romberg syndrome, some of them have associated cicatricial alopecia, localised scleroderma of other body parts other than *en coup de sabre*.<sup>3</sup>

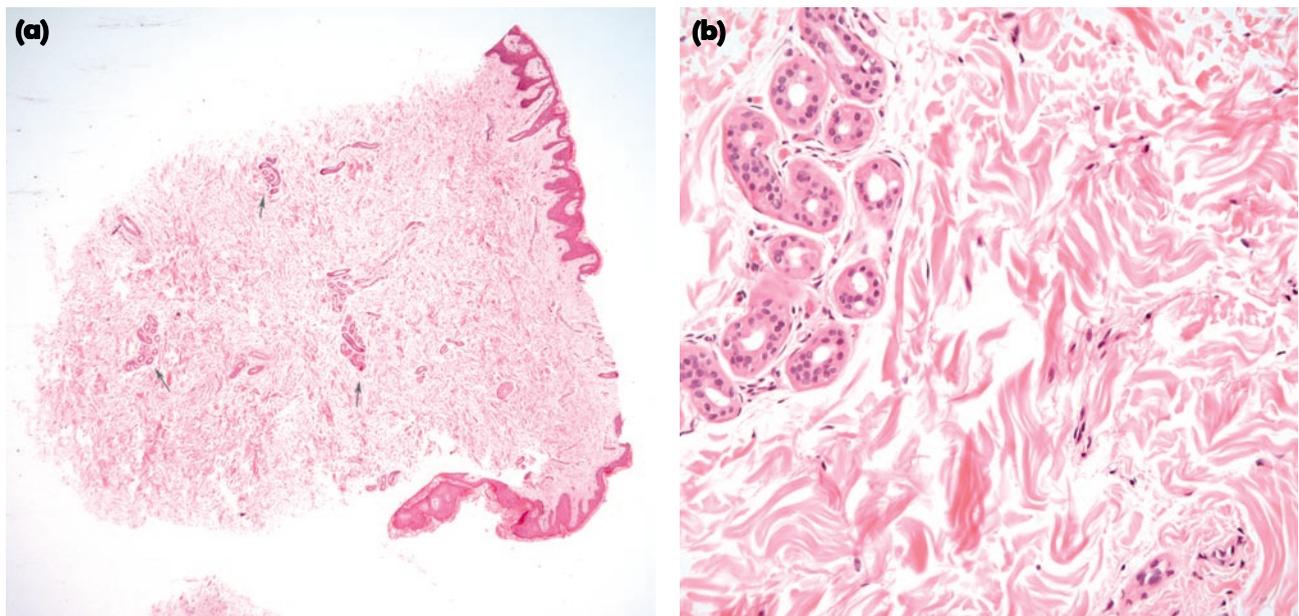
In addition to the dermatological features, neurological involvement has been found in a proportion of patients in reviews and case studies, ranging from 11% to 36%. They include epilepsy,

migraine, trigeminal neuralgia, developmental delay, psychiatric disorder, abnormal computed tomography or magnetic resonance imaging of the brain.<sup>1,3-6</sup>

The pathogenesis of Parry-Romberg syndrome remains uncertain. Autoimmunity has been suggested as a possible cause, but no evidence has been shown to be supportive of this theory.<sup>3</sup> Viral infections, trauma, endocrine disturbance have also been postulated as causative factors but their causality is yet to be proven.<sup>2,7</sup>

When a teenager with hemifacial atrophy is presented, the differential diagnoses other than Parry-Romberg syndrome include lupus panniculitis, acquired partial lipodystrophy and subcutaneous T-cell lymphoma. The diagnosis is difficult to make due to its rarity, but associated features and skin biopsy can often give clues to the diagnosis.

Many authors believe that Parry-Romberg syndrome and *en coup de sabre* are in fact variants of linear scleroderma being in the same spectrum



**Figure 4.** (a) The dermis is thickened and the sweat glands are surrounded by collagen fibres without accompanying fat. (b) Thick collagen fibres in the lower dermis.

of disease, based on the following observations: a large proportion of patients (about 50%) having both conditions at the same time, sclerodermic features as in *en coup de sabre* are often present in Parry-Romberg syndrome, and neurological associations are not uncommon in both conditions.<sup>1,3</sup>

Treatment options for Parry-Romberg syndrome are merely based on literature reviews. Tollefson and Witman pointed out in their review that three out of seven patients showed improvement after treatment with methotrexate for 5 to 48 months.<sup>1</sup> Antimalarials have also been tried for Parry-Romberg syndrome but with a poor outcome.<sup>1</sup> Topical steroids and tetracycline were also mentioned in the review as treatment in some cases but only involved one to two patients. Aesthetic treatment of the hemifacial atrophy is another option after disease stabilisation for facial reconstruction. This includes autologous fat transplantation, revascularised free flaps, lipoinjection.<sup>7-9</sup> However, these procedures could only give momentary cosmetic satisfaction, repeated or further interventions are needed when the effect wears off.

In conclusion, Parry-Romberg syndrome is a rare disease entity characterised by slowly progressive unilateral facial atrophy and may be associated with alopecia, clinical or radiological neurological abnormalities. Treatments commonly used include oral methotrexate and antimalarials but their efficacy is yet to be proven.

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