PROGRESSIVE HEMIFACIAL ATROPHY – A REPORT OF 2 CASES

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ABSTRACT

Idiopathic hemifacial atrophy (Parry-Romberg Syndrome) is characterised by progressive wasting or loss of subcutaneous tissue in half of the face, starting usually in childhood, and often associated with skin changes. Two adult onset cases are described. They did not demonstrate skin changes but one had complicated hemiplegic migraine with headaches always ipsilateral to the facial wasting.

Keywords : Hemifacial Atrophy, Complicated migraine

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INTRODUCTION

The Parry-Romberg Syndrome is a rare condition which produces disfiguring hemifacial wasting first described in 1825[1]. Since then, sporadic case reports and reviews(2-12) have appeared describing associated localised scleroderma, neurological and eye abnormalities. We describe 2 cases with this condition, the first a Chinese female and the second a Sri Lankan female, with hemifacial atrophy but without localised skin changes of scleroderma.

CASE REPORT

Case 1

SAM, a 41-year old Chinese female, first developed subacute painless swelling of the left side of her face in March 1984. This occurred over a few days and improved with medication given by a doctor. She subsequently noted progressive wasting of the affected side of the face. The wasting continued until the left side of her neck became involved towards the end of 1984. The patient sought acupuncture treatment and, although the wasting did not improve, it stayed static until the present.

There were no sensory symptoms and no limb weakness or wasting in association with the facial wasting and she did not notice any asymmetry of her smile or weakness of eye closure. The patient also denied any history of migraine headaches, seizures or hair loss. She had no history of hearing loss or impairment and there was no family history of similar facial wasting.

Clinical examination showed marked wasting of the subcutaneous tissue and muscles of the left side of the face including the temporalis, masseter and facial muscles (Fig. 1a & b). The sternocleidomastoid on the left was also wasted. There were no fasciculations and sensation over the face was intact. The smile was not asymmetrical but eye closure on the left was slightly weak with enophthalmos on the left. There were no pupillary abnormalities or extraocular muscle weakness.

Motor system testing revealed normal tone and full muscle power with symmetrical, brisk tendon reflexes. No alopecia or skin changes of scleroderma was present.

Needle electromyography of the temporalis, masseter and muscles of facial expression on the left revealed no spontaneous activity and normal voluntary motor unit potentials. There was no difference in conduction latency for facial nerves on both sides stimulating at the stylomastoid foramen and recording over the nasalis muscle. Both computed tomographic scan of the brain and an electroencephalgram were normal.

The patient has since not improved but has shown no progression of the wasting. She is otherwise in excellent health and declined referral for cosmetic surgery.

Case 2

SBP, a 32 year old Sri Lankan female, presented with acute left hemiparesis and hemianaesthesia resolving over five days following a severe right sided throbbing headache in December 1987. She gave a history of recurrent right hemicranial headaches associated with nausea but no preceding visual prodrome since adolescence. In addition, she had developed progressive, painless and disfiguring wasting of the right side of her face over the past four years. There was no associated loss of sensation or hearing loss with this wasting. Neither the patient nor her husband noted any asymmetry in her facial expression prior to the onset of the hemiparesis.

In her past history, she had undergone resection of a left cervical fibrous band which had produced a 'cervical rib' syndrome on the left with wasting of the left

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Fig 1a & b
Marked wasting of the temporalis, masseter, facial muscles and the sternomastoid on the left side.

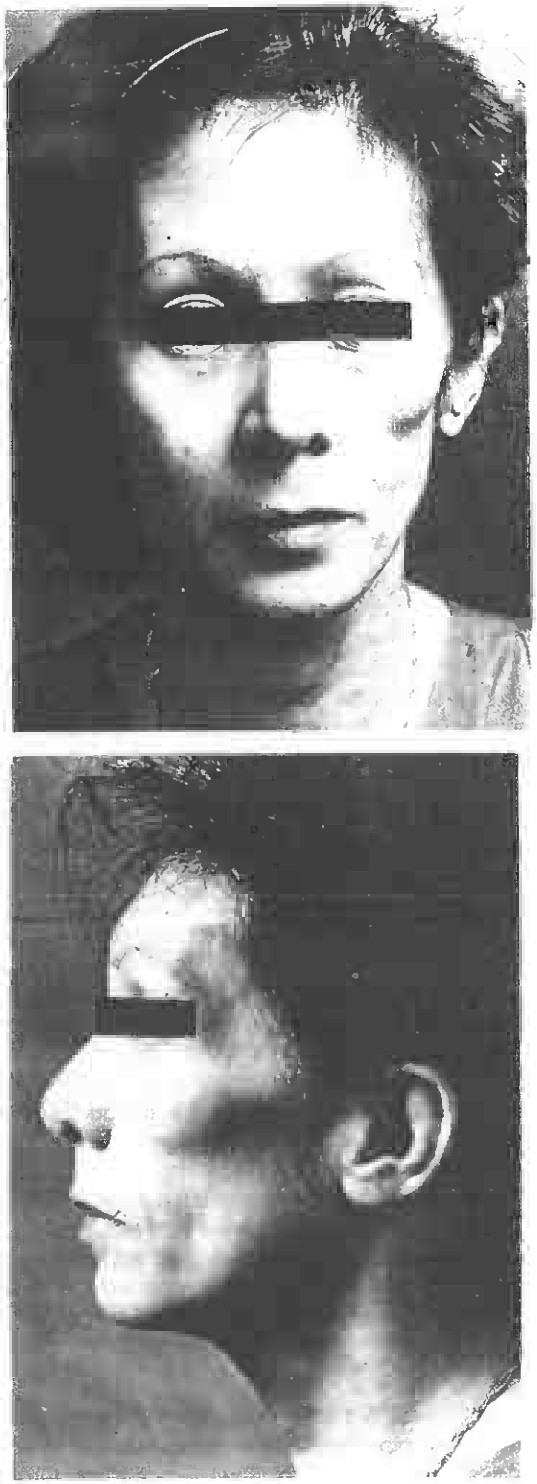

Fig 2 a & b
Wasting of temporalis, masseter and facial muscles on the right side. The sternomastoid was spared.

hypothenar eminence. Both the patient and her husband are also under investigation for primary infertility and the patient was previouly under treatment for depression because of her multiple medical problems. There was no family history of similar facial wasting.

Clinical examination revealed severe wasting of the subcutaneous tissue as well as the temporalis, masseter and facial muscles on the right (Fig. 2a & b). The sternocleidomastoid was spared and sensation over the face was intact. There was no asymmetry of her smile and eye closure was complete bilaterally.

The other cranial nerves were intact with no pupillary abnormalities and no diplopia. No skin changes of scleroderma or alopecia were noted. Motor system examination showed Grade 4+ weakness of her left arm and leg. The deep tendon reflexes were normal. Plantar responses were flexor. No fasciculations were seen over the face. Wasting of the left hypothenar eminence due to the previous cervical fibrous band was the only other finding clinically.

A computed tomographic scan of the brain confirmed a lacunar infarct at the posterior limb of the right internal capsule. Four vessel cerebral digital subtraction angiography showed normal cerebral vessels. Electroencephalogram showed no epileptogenic activity. Needle electromyography of the facial muscles and

temporalis on the right showed no denervation with normal voluntary motor units and recruitment. Needle EMG of the left hypothenar muscles revealed a pattern of denervation with reinervation and nerve conduction velocities for the left median and ulnar nerves were within normal limits. The hematological profile was normal as was her serum creatinine and urea. Her erythrocyte sedimentation rate was not elevated and the anti-nuclear antigen was negative. Diabetes was excluded with a 75g oral glucose tolerance test.

The patient was given aspirin 150mg daily and started on propranolol 20mg bid as prophylaxis for her migraine headaches. Her facial wasting has continued to progress despite improvement in her headache frequency.

DISCUSSION

Progressive Facial Hemiatrophy, first described by Parry[1] and Romberg[2], is also known as Facial Hemiatrophy of Romberg or the Parry-Romberg Syndrome. This condition is rare and has interested neurologists, dermatologists, ophthalmologists and cosmetic surgeons since its early descriptions. Despite this interest, the cause remains unknown. The characteristic feature is progressive wasting or loss of subcutaneous tissue involving half of the face.

This wasting may involve the underlying muscle and bone, occasionally spreading to involve the ipsilateral neck, trunk or even limbs[3]. Interestingly, the function of the wasted muscles is usually unaffected and no abnormality is demonstrable on needle electromyography. This latter point is illustrated in our two cases, confirming that the loss is principally of subcutaneous tissue and fat.

Wartenberg[3] described the condition as usually starting with circumscribed alopecia and the appearance of a depressed linear scar in the skin to scleroderma's "coup de sabre". Most cases also present in childhood, the facial wasting progressing together with the development of the poikiloderma and skin thickening of scleroderma. Onset in adult life is less common but does occur. Rogers[4], in his analysis of 772 cases, noted a slight female preponderance with a female to male ratio of 3:2. He also mentioned a non significant slight predominance of left sided wasting.

The major associated condition is scleroderma[3,5-9], which is usually circumscribed and confined to the face but generalised forms occur. Important neurological accompaniments include epilepsy and migraine, ophthalmic complications being the other significant problems[3,5,10,11]. Characteristically, patients have focal epilepsy or epilepsy with focal onset and secondary generalisation. The epileptogenic focus is usually ipsilateral to the facial wasting. Similarly, the migraine headaches always occur on the side of facial atrophy[3,5,10,11] as is the case with patient 2. Cerebellar ataxia is an uncommon complication and pathological evidence of cerebellar degeneration had been demonstrated[10].

Eye abnormalities are many and include enophthalmos, lid changes, pupillary abnormalities and ophthalmoplegia[10,12]. The enophthalmos probably occurs from loss of subcutaneous tissue around the orbit of the eye and is the commonest eye abnormality encountered. Bilateral ptosis and Horner's Syndrome have all been described. Kuto, Sakaguchi et al[7] described a single case with associate Henoch-Schonlein Nephritis and paroxysmal nocturnal hemoglobinuria.

It is probable that what is often described as the Parry-Romberg Syndrome is a collection of unrelated conditions with the common feature of facial hemiatrophy. There is certainly a distinct subgroup with early onset, a strong association with scleroderma and frequent neurological abnormalities in the ipsilateral cortex.

Unfortunately, the cause of this condition remains unknown and no form of medical intervention appears to show consistent benefit; Case 1 possibly responding to acupuncture notwithstanding. Wartenberg felt that the problem was in the sympathetic innervation to the face[3]. In most instances, the wasting ceases or burns itself out after producing gross disfigurement. Attempts to cosmetically reconstruct the face offer some hope and the vogue appears to be repeated silicone injections[12] into the wasted tissue.

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