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A FEMALE PATIENT WITH PARRY-ROMBERG SYNDROME: A CASE REPORT[+]

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A 40-year-old woman with left progressive hemifacial atrophy (Parry-Romberg syndrome/PRS) is reported. PRS is an unusual condition of unknown etiology, characterized by spontaneous and slowly progressive atrophy of the subcutaneous tissue and fat.

We present the findings of a patient with PRS who was observed in our clinic for over four years. Ophthalmic and neurologic involvements are reviewed. The patient presented with progression of enophthalmos associated with pseudoptosis that occured secondary to intraorbital fatty atrophy, alopecia, poliosis, iris heterochromia, chronic dacryocystitis and lack of blink reflex.

Key words: *Parry-Romberg syndrome, enophthalmos, blink reflex.*

Bir Kadın Hastada Parry Romberg Sendromu: Olgu Sunumu

PRS'u etyolojisi belli olmayan, yüzün bir yarısında derialtı yağ ve diğer dokuların kendiliğinden yavaş seyirli ilerleyici atrofisi ile karakterize nadir görülen bir tablodur.

Kliniğimizde sol progressif hemifasial atrofi nedeniyle 4 yıldır takip ettiğimiz, 40 yaşında kadın hastada oftalmik ve nörolojik bulgular gözden geçirilmiştir. Hastada, intraorbital yağ dokusu atrofisine ikincil olarak gelişen ve psödoptozisin eşlik ettiği ilerleyici enoftalmus, allopesi, poliozis, irisde heterokromi ve kırpma refleksinde kayıp saptanmıştır.

Anahtar kelimeler: *Parry-Romberg sendromu, enoftalmus, kırpma refleksi.*

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Parry-Romberg syndrome (PRS), also known as progressive facial hemiatrophy, is an uncommon disease with progressive facial asymmetry as the major symptom.[1-3] There is a slight female predominance and the disorder typically begins within the first two decades.[1-3] It is an atrophic process of subcutaneous fatty tissue and its etiology is unknown.[1-2] The etiology of PRS remains controversial, and its relation scleroderma and autonomic nervous system disorders are discussed.[1-2] Skin, muscles and bones can be secondarily affected.[1,4] It most often affects the half of the face and scalp on the involved side.[1,2,5] Hemiatrophy of subcutaneous tissue and fat is usually the presenting sign of the disease.[2,5,6] Involvement of the surrounding soft tissues and the orbit is common, such that progressive unilateral enophthalmos with subsequent changes in the palpebral fissure occurs.[2,5,6]

CASE

A previously healthy 40-year-old woman with progressive hemifacial atrophy involving the left side of the chin is reported (Figure 1). The atrophy at her left face started about ten years ago. She gave a history of an operation for chronic dacryocystitis in another ophthalmology clinic and still had a minimal epiphoria. The visual acuities in both eyes were normal with spectacle correction. She presented with severe enophthalmos associated with eyelid atrophy, loss of cilia, moderate pseudoptosis (3 mm) and heterochromic iris. There was alopecia localized in the vertex and atrophic areas of skin, recognized as scleroderma 'en coup de sabre' (Figure 2). The Hertel exophtalmometer results were 13 mm in the right eye and 9 mm in the left eye. Computed tomography (CT) and magnetic resonance imaging (MRI) of the brain were normal. Orbital CT and MRI findings included the intraorbital fat atrophy and enophthalmos in the involved left side. In the blink reflex test of the left eye, there were electromyography findings related with the involvement of nucleus and proximal part of facial nerve.

Figure 1. This photograph demonstrates the prominent hemifacial atrophy, enophthalmos and pseudoptosis in the involved side of the face.

Figure 2. This photograph shows extensive areas of alopecia in the vertex region.

DISCUSSION

PRS is a poorly undertsood disorder characterized by progressive hemifacial atrophy involving the skin, soft tissue, and bone.[7] The early facial asymmetry progresses over years to a striking atrophy of subcutaneous tissue.[1-3] When the disorder is well developed, the appearance is striking: the affected side of the face is sunken and wrinkled.[1-3,6]

Many authors have proposed an involvement of the sympathetic nervous system.[8,9] Wartenberg believes that the primary disturbance involves an active process in higher centers resulting in an irritative,

unregulated control over lower centers.[10] Secondary involvement of the peripheral nervous system causes local disturbances in fat metabolism and subcutaneous tissues, leading to alopecia and atrophy.[5] Electromyography, blink reflex and trigeminal evoked potential abnormalities indicate that the brain stem may be implicated in the aetiology of the disease.[8] Immunological evidence favoured this possibility and demonstrated possible involvement of the noradrenergic system.[8,9,11] Hyperactivity of the brain stem sympathetic centres, possibly caused by an autoimmune process, may be the primary cause of the cutaneous and subcutaneous atrophy in PRS.[8,9,11] The association of PRS with Horner's syndrome and Fuch's heterochromic iridocyclitis increases the possibility of this mechanism.[2,12,13]

Ophthalmic involvement in PRS is common, such as progressive enophthalmos, pseudoptosis, pupillary disturbances, heterochromia, uveitis and restrictive strabismus.[2,12,13] Although there is no definite therapy of PRS, aesthetic surgical treatments of enophthalmos and skin atrophy may be performed.[14]

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