Photoclinic

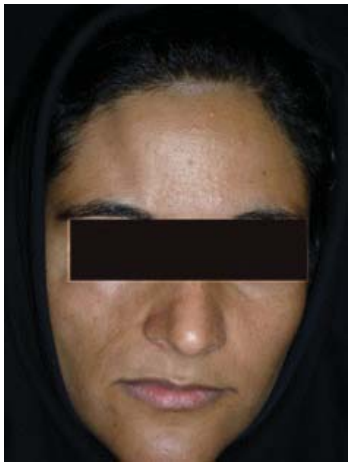

Figure 1. Rt. hemifacial atrophy

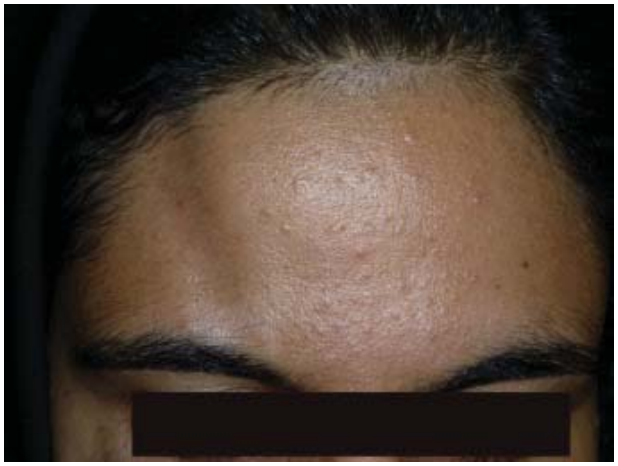

Figure 2. Rt. frontal atrophy and depression

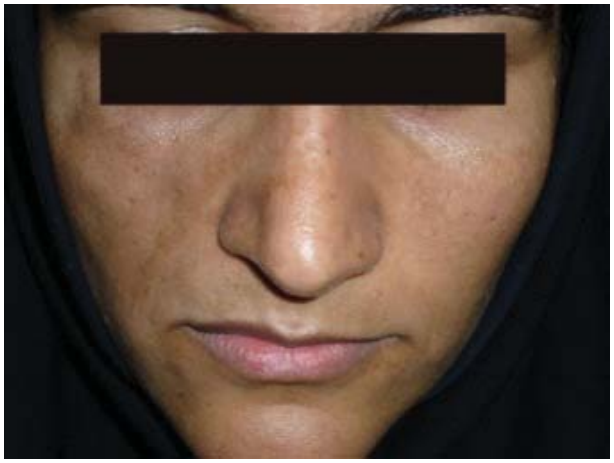

Figure 3. Rt. zygomatic atrophy leading to Rt. cheek indentation

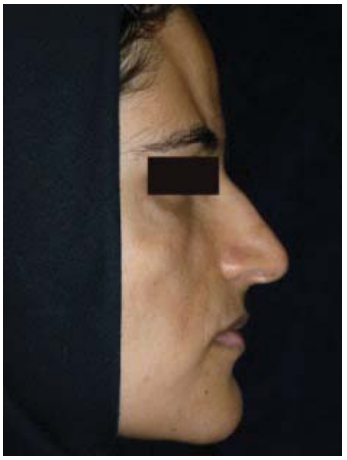

Figure 4. Rt. frontal and zygoma indentations, disfiguring patient's face (lateral view)

Cite this article as: Paydary K, Esmaeeli Sh, Jabbari H. Photoclinic. *Arch Iran Med*. 2013; **16(9):** 565 – 566

The presenting case, a 27-year-old single young woman from Mashhad, Iran, came to our hospital Outpatient Department with the chief complaint of asymmetric facial appearance. The patient was totally healthy until two years prior to admission when she observed a dark colored line over the central section of her forehead originating from the medial corner of the left eyebrow. Two months later, she noticed a painless dimple over the right side of her forehead with no burning sensation, pain, or itching. At six months after the onset of symptoms patient's father passed away. As she claims, this caused worsening of her symptoms over the subsequent two months. She referred to several physicians, including an ear, nose, and throat specialist. The patient was prescribed prednisolone, 35 mg qd for ten months without any obvious improvement. At 18 months of starting symptoms, she detected a similar abnormality over her right cheek, which became stationary after few months. On physical examination, asymmetry and atrophy of the right side of face were observed. There was a depression in the right frontal region, right zygoma, and right side of the nose from midline along with irregularity of the underlying right zygomatic bone, which denoted an underlying bone involvement, but with no tenderness over the affected areas. (Figures 1 – 4) The patient's past medical history was unremarkable for any severe or significant medical problem, accident or any surgical intervention. There was no family history of bone, joint or metabolic disease or any other underlying bone or soft tissue problem. On laboratory examination, her complete blood count, urinalysis and anti-nuclear antibody were within normal limits. CT scan of the skull was performed to evaluate the extent of involvement.

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What is your diagnosis?
See the next page

Photoclinic Diagnosis: **Parry-Romberg Syndrome**

Parry-Romberg syndrome (PRS) is a rare disorder characterized by progressive hemiatrophy of the skin and adipose tissue, with occasional involvement of the muscles, cartilage, and underlying bony structures.[1,2] This syndrome mostly occurs in a sporadic fashion and has an unknown etiology. It usually begins in childhood and mostly involves the upper face. The principle features are atrophy of the soft tissues and sometimes the bone, on both or one side of the face or forehead.[3] It may start with hyper- or hypo-pigmentation, followed by slowly progressive atrophy that usually remains limited to the distribution of the trigeminal nerve or may involve the entire face.[4] There may be various ophthalmologic complications such as loss of eyebrows or eyelashes, iris atrophy, ocular muscle palsies and uveitis. There may be neurological complications such as migraines, headaches, hemiparesis, trigeminal neuralgia and unilateral epilepsy.[3,5] New studies report that bilateral disease is becoming more common.[6] Trauma, viral infection, endocrine disturbances, and heredity factors are believed to be associated with this disease.[7]

The treatment goal is to improve the physical appearance. The offered treatment for PRS patients is based on the replacement of tissue that was lost due to atrophy; there is preference for auto grafts due to the lower risk of tissue rejection along with less local and systemic inflammatory reaction, which could cause graft loss or other co-morbidities. According to the literature, clinical treatment of the disease with immunosuppressive drugs or other medications used for scleroderma such as chloroquine and calcipotriol have been discussed however until now, there is inadequate scientific evidence for their benefit.[8] Our patient's plan is watchful waiting, followed by plastic surgery after disease stabilization.

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