What is Parry Romberg Syndrome?

One of the least known autoimmune diseases is Parry Romberg Syndrome (PRS), a disorder that gradually deteriorates one side of the face or head. Also known as hemifacial/cranial atrophy or facial hemiatrophy, PRS will most often present itself by deterioration and asymmetry of the soft tissue and/or bone. The atrophic process can, in varying degrees, affect the brain, skull, hair, skin, eye, ear, nose, mouth, teeth, cheek and/or neck, generally limited to one side. In rare cases, PRS can also be bilateral and/or can affect the trunk as well. PRS has often been referenced with regard to Localized Linear Scleroderma and En Coup de Sabre Morphea.

No two cases of PRS are alike; and because PRS is a slow, insidious process, patients often display symptoms limited to one side of the face or head well before any noticeable or remarkable facial atrophy is present. This makes PRS an often misdiagnosed or underdiagnosed syndrome. Along with a multitude of one-sided symptoms, PRS complaints can include the following:

—Neurological (migraines, seizure, stroke, trigeminal neuralgia, brain lesions, blood vessel and hemibrain atrophy)

—Ophthalmic (uveitis, decreased ocular pressure, global retraction, vision loss, eyelid asymmetry)

—Dermatological (hair loss, pigmentation changes, en coupe de sabrederma, epidermal atrophy)

—Dental/orthodontic (asymmetry of the maxillary structures, lips, tongue and/or gums; root malformation or re-absorption)

—Trunk (atrophy of trunk, arm, leg)

PRS is more prevalent in women, but males and females of all ages and all nationalities are affected. Although onset can be at any age, it often begins in the first two decades of life. It is very common for patients with PRS to have intermittent periods of remission and reoccurrence. While often referred to as sporadic in etiology, documented cases of familial PRS suggest that there is a multifactorial genetic trait, with those multifactors including any combination of the following: genetic predisposition to autoimmunity, hormones, stress, infection from virus/bacteria, and/or even very mild trauma.

Initial diagnosis is based upon clinical observation; and frequently, serological testing confirms that autoimmunity plays a role in the disease. In cases where there are symptoms but not a significant notable atrophy, it is interesting to note that the practitioner can make a more informed diagnosis by standing behind a suspected PRS patient while that patient looks into the mirror. Reverse image or mirror image reflection enhances the evidence of asymmetry in a PRS patient, making the disease easier to diagnose.

PRS patients seek treatment from the medical specialty that ministers to their individual symptoms, and those complaints are treated symptomatically or surgically, in the case of facial reconstruction.

More information regarding PRS can be accessed through The Romberg Connection at http://www.geocities.com/HotSprings/1018/. ■

—This information was provided by AARDA member Sally VanRaemdonck who has had personal experience with Parry Romberg Syndrome.