Parry Romberg Syndrome

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Parry Romberg Syndrome, also described as Progressive Facial Hemiatrophy is a rare acquired, neurocutaneous disorder characterized by slowly progressive atrophy of one side of the face, primarily involving skin, subcutaneous tissues, fat, muscle and bone in some cases without motor weakness. Its etiology is unknown and some consider it to be autoimmune in origin due to overlap with linear scleroderma and its associations with other autoimmune diseases, with ophthalmic and thyroid involvement.

The disease varies in severity from those with barely perceptible asymmetry to severe disfigurement and has a diverse presentation. Patients have been reported to have coexistent epilepsy, migraine, trigeminal neuralgia, visual symptoms, unusually cold hands, jaw symptoms, dental abnormalities, anxiety and depression.

The disease tends to progress intermittently and so the effects of the medications tried is difficult to attribute to them. Medications like Steroids, Methotrexate, Azathioprine, Penicillamines, and Intravenous Immunoglobulin have been tried in a few cases with variable success. No single drug or a combination of drugs has proved to have sustained effect on this disease. Most of the drugs tried have potentially hazardous side effects so the therapy must be individualized. Surgical treatment with lipoinjection, flap or pedicle procedure, bone implant have been tried with variable success when the manifested deformity of disease is static for a period of two years. Patient is advised to be on regular follow up with medical care.

References