VASCULAR DISEASE, VASCULITIS

WEGNERS GRANNULOMATOSIS MIMICKING HANSENS DISEASE

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Granulomatosis with polyangiitis (GPA), formerly known as Wegener granulomatosis, is a rare multisystem autoimmune disease of unknown etiology. Its hallmark features include necrotizing granulomatous inflammation and pauci-immune vasculitis in small- and medium-sized blood vessels. Other features include, chronic sinusitis being the most common initial complaint (67%). rhinitis and epistaxis.
The development of a saddle nose deformity is common. Serous otitis media and hearing loss are the presenting manifestations of GPA in some patients. Peripheral nervous system manifestations include mononeuritis multiplex, sensorimotor polyneuropathy, and cranial nerve palsies. Pulmonary disease may cause any of the following : Pulmonary infiltrate ,cough, hemoptysis etc
Leprosy can also cause saddle nose deformity, eye symptoms, tingling sensation and other systemic manifestations.
Here is a case of 45 year woman who presented to us with history of nasal bleed and nasal stuffiness with saddle nose deformity, redness and pain of both eyes No history of any hypopigmented patches but history of tingling sensation over the hands was present. She also gave history of difficulty in breathing and hard of hearing since 5 years. Her SSS was negative and nerve biopsy also did not show any changes of hansens.
On ophthalmic examination she was diagnosed with necrotizing scleritis with keratouveitis. On audiological examination she had sensory neural hearing loss. ANA profile: negative,
C-ANCA was positive,CRP was raised (130) and ESR was raised. CT chest showed collapse of superior segment of lower lobe. Hence a diagnosis of wegners granulomatosis was made and patient was started on steroids and cyclophosphamide.