PAEDIATRIC DERMATOLOGY

SNEDDONS SYNDROME-A CASE REPORT

Maheshwari Nallur Siddaraju [1] - Priyadarshini K [2] - Bhumika S [3]

Cosmetic Skin Care Clinic, Dermatology, Bangalore, India [1] - Kutiscience Clinic, Dermatology, Bangalore, India [2] - Specialist Hospital, Dermatology, Bangalore, India [3]

Background: Sneddon syndrome is a rare, non-inflammatory vasculopathy characterised by Livedo Racemosa and cerebrovascular-thrombotic attacks like dizziness, paresis and seizures. Livedo Racemosa is characterised by bluish/erythematous, closely arranged rings producing a netted pattern. Cutaneous features precede neurological symptoms in most cases. It is classified as either Primary, if no underlying etiology is found or Secondary, if associated with autoimmune disorders like Antiphospholipid Antibody Syndrome and Systemic Lupus Erythematosus, or Coagulation disorders.

Observation: An eight-year old girl was presented with diffuse erythematous net like lesions all over the body including palms and soles with relative sparing of central face, which was histopathologically confirmed as Livedo racemosa. An isolated Nevus Comedonicus, was noticed over the left knee as multiple grouped keratotic follicular plugs.
Neurological examination confirmed decreased intelligence and cognitive impairment but a normal speech, sensory, motor functions and tendon reflexes were noted at time of presentation. Ophthalmological examination showed a right retinal detachment leading to phthisis bulbi
A previous cerebral angiogram showed bilateral internal carotid artery narrowing with collaterals from the external carotid artery and vertebral artery, and bilateral segmental aneurismal features suggestive of vasculopathy. Magnetic resonance imaging of brain showed features of gliosis due to an old vascular insult in the left fronto-parietal and right frontal region.
Presence of Livedo Racemosa, neurological and ophthalmic features, histopathology and thrombotic signs in Magnetic resonance imaging helped us make a diagnosis of Sneddon syndrome. To the best of our knowledge, this is the first case report from India.

Key message: What is known- Sneddon syndrome (SS) patients can develop neurological and ophthalmological complications like seizures, paresis, aphasia, retinopathy, visual field defects and retinal neovascularization.
What's New- Presentation of multi-systemic involvement with classical findings in a very young girl which is extremely rarely reported. Nevus comedonicus was found as an isolated association with SS.