A case of optic nerve hydrops with tolosa hunt syndrome: A case report

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ABSTRACT
A 36-year old female patient presented with complaints of discomfort in left eye associated with mild morning pain since 3 days duration. Patient had history of facial nerve paresis in the right side, two years back that recovered spontaneously. On examination, the visual acuity was 6/6 in the right eye and 6/6P in the left eye. Anterior segment of both eyes was normal in both eyes. Fundus examination of the right eye was normal while of the left eye showed mild disc oedema. Patient was kept under observation and was reviewed in two days. She complained of persistent discomfort in the left eye and double vision. On examination the visual acuity, color vision was normal. She developed abduction weakness and elevation deficit in the left eye. The disc oedema had increased in the left eye. She underwent MRI of Brain and orbit, which showed optic nerve hydrops in the left side. Neurological examination was normal except for the findings noted above.

With consultation with neurologist and internist she was started on tablet acetazolamide 250 mg thrice a day and intravenous methylprednisolone 1gm daily for 3 days. The initial MRI features were suggestive for Optic nerve hydrops due to Benign Intracranial Hypertension when a follow up MRA / MRV showed features of optic nerve hydrops with Tolosa Hunt Syndrome. The optic nerve hydrops presenting with Tolosa Hunt Syndrome is unusual in our case.

Keywords: optic nerve hydrops, tolosa hunt syndrome

CASE REPORT
A 36-year old female patient presented with complaints of discomfort in left eye associated with mild morning pain since 3 days duration. Patient had history of facial nerve paresis in the right side, two years back that recovered spontaneously. On examination, the visual acuity was 6/6 in the right eye and 6/6P in the left eye. Anterior segment of both eyes was normal in both eyes. Fundus examination of the right eye was normal while of the left eye showed mild disc oedema. Patient was advised to undergo MRI Brain and Orbit for evaluation of the optic disc edema. While the patient was waiting for the CT scan she developed drooping of the left lid and double vision. On examination, she had the best-corrected visual acuity of 1.0 in both the eyes. There was a moderate left upper lid ptosis, limitation of abduction and elevation. The pupils were normal size and reactive in both eyes. Fundus examination revealed slightly increased optic disc edema in the Left Eye. She was advised to undergo MRI Brain and Orbit which revealed left optic nerve sheath hydrops with no intracranial mass. She was referred to internal medicine physician who advised MRA / MRV for this patient. The patient was started on intravenous Methyl Prednisolone 1gm a day for 3 days along with proton pump inhibitors.

The Visual acuity, color vision were monitored during the course of treatment which remained normal. The patient had marked improvement in her symptoms, her Diplopia disappeared and the Ptosis improved. The ocular movements also showed improvement in abduction and elevation. The MRA / MRV study showed dilated cavernous sinus and a diagnosis of Tolosa Hunt Syndrome with Optic nerve
Hydrops was made.

After 3 days of Intravenous methylprednisolone she was put on Oral prednisolone 40 mg per day in tapering doses with proton pump inhibitors and Vitamin B12 supplementation. She continued to show improvement in symptoms. Her visual Acuity and color vision remained stable throughout the treatment. At the last follow-up she had no Diplopia, no Ptosis with full ocular movements. She has been advised a follow up MRA / MRV scan.

DISCUSSION

Optic nerve dural ectasia is saccular dilatation of the optic nerve sheath. It is characterized by expansion of the cerebrospinal fluid (CSF) around the optic nerve without orbital or cerebral neoplasm and inflammation. It was first described in the year 1918. It is commonly associated with Idiopathic Intracranial Hypertension. However in our case the optic nerve hydrops was associated with Tolosa Hunt Syndrome which is very rare and only few cases have been reported. Tolosa-Hunt syndrome is a painful ophthalmoplegia caused by nonspecific inflammation of the cavernous sinus. It presents with retroorbital or periorbital pain, diplopia, visual loss, paresthesias along the forehead due to involvement of first division of the trigeminal nerve. The cause of Tolosa Hunt syndrome is idiopathic.

Our case was treated as a case of Optic Nerve Hydops with Tolosa Hunt syndrome with systemic steroids and oral acetazolamide. This led to a quick improvement in the patient symptoms of diplopia and pain. The association of the optic nerve hydrops and Tolosa hunt syndrome is not clear and has not been reported in the literature. It is postulated that some factor triggered the inflammation leading to development of Tolosa Hunt Syndrome and Optic Nerve Hydrops.

A follow up MRA / MRV was advised for follow up.

REFERENCES