ORBITAL APEX SYNDROME AND RUBIOSIS IRIDIS: RARE COMPLICATION OF HZO

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Introduction:

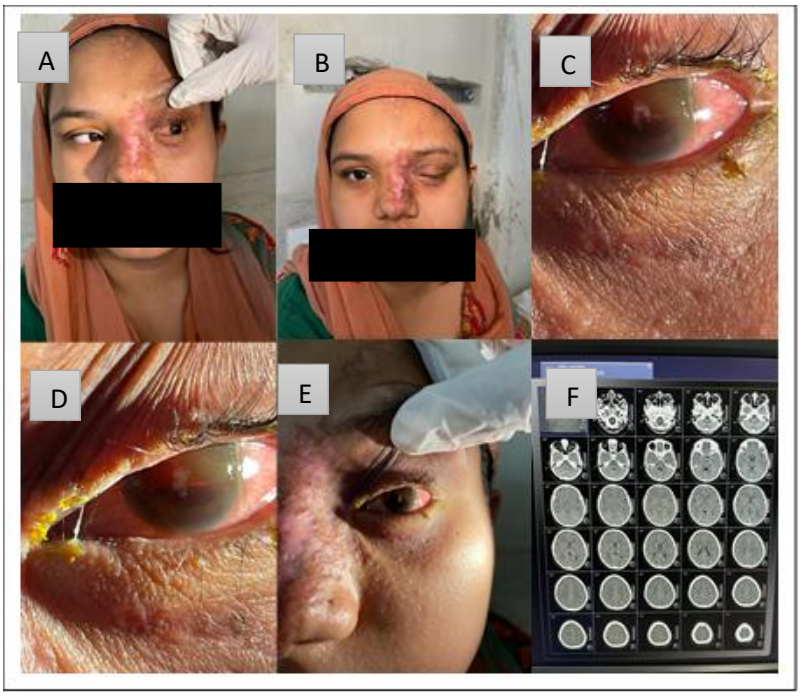
In early childhood, infection with Varicella Zoster virus (VZV) results in a disease entity known as chickenpox. This virus remains dormant in the sensory ganglion of the cranial nerve for decades and its reactivation causes Herpes Zoster in the affected dermatome. Herpes Zoster ophthalmicus is a term used when the Varicella-Zoster virus is reactivated in the dermatome supplied by the ophthalmic division of the Fifth cranial nerve. Most patients present with a periorbital vesicular rash distributed according to the affected dermatome. Ocular involvement occurs in 20–70% of patients with HZO and may include Blepharitis, Keratoconjunctivitis, Iritis, Scleritis, and Acute Retinal necrosis. Our article will be the first to report a case of a child who developed OAS and Rubiosis iridis as a complication of HZO.

Case history

A 16-year-old girl from Ghaziabad presented in the Ophthalmology department of Santosh medical college and hospital with complaints of a sudden painful loss of vision in LE with the inability to open her eyes for the last 3 days. There was a history of fever with the eruption of vesicle lesions over the forehead and nose associated with edema and erythema around the left eye 3 weeks back and the patient was diagnosed as Herpes Zoster ophthalmicus. She took treatment for the same elsewhere. On examination it was found that the patient's vision in affected eye was PL +ve and PR inaccurate in nasal and inferior quadrant.

There was complete ptosis(Fig 1 :B), conjunctival and ciliary congestion was seen(Fig 1:C,D). The cornea was clear and Anterior Chamber was totally filled with organized Hyphema(Fig 1:C,D). Iris and Lenticular details were not visible. Pupillary reaction direct and consensual were not appreciated due to hyphema but in the normal eye, consensual response was absent. In Fundus no glow was seen. The ocular movement was restricted in lateral gaze(Fig 1:A). Digital tension was high.

Fig. 1: Picture showing Clinical Presentation of the case



There was the presence of scarring over the tip and lateral wall of the nose and forehead on the affected side(Fig 1:A,B). All the investigations pertaining to identifying the cause were carried out. As this condition is uncommon in a child all blood investigations to rule out immunocompromised status were carried out including HIV, Diabetes. Chest X-Ray PA and Montuax test to rule out tuberculosis. CT Scan brain and orbit and B Scan were done. All the reports were normal. MRI Brain and orbit was also advised but due to financial issues, patient attendants were unable to get it done. Paediatric reference was also done so as to rule out any kind of systemic diseases.

The clinical diagnosis of orbital apex syndrome with hyphema post-Herpes Zoster Ophthalmicus infection was made.

The patient was suspected to have Orbital Apex Syndrome as there was the involvement of three different nerves. After paediatric consultation full doses of drugs were given as the weight of the child was 68 kgs. The patient was started on systemic Acyclovir, 800 mg (5 times a day) along with systemic Steroids – Tab Omnacortil, 60 mg (once a day), Tab Diamox – 250 mg BD along with Vitamin supplements in form of Vitamin C, 500 mg once daily and Vitamin A, 25000 IU – 8 capsules stat. Local treatment in the form of antibiotic, anti-viral, and steroid drops was started for the patient. The patient started showing remarkable improvement after 3 days and complete resolution of ocular movement after 1 week of steroid treatment. The Ptosis was partially resolved in 2 weeks. But there was no improvement in Hyphema in spite of giving local and systemic steroids. The patient was taken for an Anterior chamber (AC) wash. After the AC wash, it was observed that the Iris was having new vessels over it and there was complete obscuration of the pupil. In spite of the Ac wash, the patient did not show any improvement in vision.

Discussion

Herpes zoster ophthalmicus is a disease entity that occurs after the reactivation of the varicella-zoster virus in the 5th cranial nerve (2). The mean time from onset of vesicles to ocular involvement is 1.82 weeks (range 1-4 weeks) (1) Hutchinson sign states that when the tip and side of the nose are involved by the vesicles there is a strong possibility of the involvement of ocular structures too. Nasociliary nerve involvement was associated with subsequent ocular disease. (1) The incidence and severity of disease increase after 60 years of age (4, 5) . Ocular involvement occurs in 20–70% of patients with herpes zoster ophthalmicus (6). Ocular involvement with the virus commonly causes blepharitis, conjunctivitis,keratitis, and uveitis. Neurological complications are rare and may cause ophthalmoplegia, ptosis, optic neuritis, and rarely OAS(7). The direct viral invasion leads to conjunctivitis and epithelial keratitis. Secondary inflammation and occlusive vasculitis may result in more severe diseases like Scleritis, Optic neuritis, and cranial nerve palsies. In 11 to 29% of cases of HZO, ophthalmoplegia has been reported (8). The most frequent nerve involved is the oculomotor nerve followed by Abducent (9). OAS is characterized by paralysis of cranial nerves II, III, IV, and VI and the ophthalmic branch of the cranial nerve V, caused by the inflammatory, infectious, neoplastic, traumatic, vascular, and sometimes iatrogenic causes along the ophthalmic canal (12). Mucormycosis and Aspergillosis are the most common infectious causes of OAS . Therefore this condition should be considered in immuno-compromised, diabetic and patients with malignancy. Paranasal sinuses are primarily involved, after which the disease progresses to the orbital apex (14). Sometimes a patient of HZO may develop OAS a very rare complication where there is the involvement of the third nerve, 4th nerve, 5th nerve, and 6th nerve associated with involvement of 2nd nerve (10,11) First such case of it was reported by Ramsell in 1967(13) Exact mechanism for the development of extraocular muscle paresis and optic neuropathy is not completely understood in the patients of HZO and OAS but different suggested mechanisms of the involvement of ocular tissue include extensive inflammation around the posterior ciliary nerve and ocular vessels, direct compression of 2nd, 3rd, 4th, 6th cranial nerve by soft tissue edema. The disease manifestation will vary according to the mechanism involved. In our patient as three nerves (2nd, LPS branch of 3rd nerve, 6th nerve) were involved, we diagnosed it as an early case of orbital apex syndrome through exclusion and curbed its progress through early and rigorous treatment. The duration of development of OAS is unclear but in our patient, it developed after 3 weeks of vesicular eruption. A similar case was observed by kocaoglu et al. where the patient had developed OAS in the second week of treatment (22). There are not many reported cases of such a condition. We found only 10 such cases while looking for this condition out of which four patients were in their sixties (15-18), two patients were in their seventies (19,20) and two were in their eighties (21,22). There was only one reported case in the younger age group, a 29-year female who was immuno-compromised (23). In our case, the age of the child was only 16 years who otherwise was a healthy female. Only a few articles were found in which HZO occurred in children. According to a study done by

Denise De Freitas etall the mean age of presentation was 8.7 years, two patients (20%) had decreased visual acuity and nine (90%) had some degree of abnormal corneal sensitivity and corneal opacity despite good final visual acuity (24). Our patient not only developed OAS and rubiosis iridis but also had no major improvement in her vision.

The virus causes arterial occlusion by vascular remodeling as well as the accumulation of immune complexes formed due to infection on the vessel wall. These processes result in occlusive and thrombotic granulomatous arteritis. If the central retinal artery gets involved then it may lead to CRAO.There is only one reported case of Neovascular glaucoma post HZO (25). In our case it it difficult to say what caused rubiosis iridis. But there is high possibility that the patient developed CRAO which lead to the development of Rubiosis iridis as in the case reported by Ahmad SS et all. where the patient developed Neovascular glaucoma after 3 weeks of vesicular eruption (25) In our case also, the patient developed hyphaema after 3 weeks of vesicular eruptions.

While the therapeutic modality for OAS secondary to HZO is unclear, the current treatment modality involves combined administration of sytemic acyclovir and systemic steroids (22,26). In all the reported cases, patients have shown improvement with the above mentioned treatment. In our case as well, we have noticed improvement in the patient's ptosis as well as ocular movements, but the vision was not fully restored.

Complete or near complete resolution of ophthalmoplegia due to HZO has been reported in about 76.5% of cases and may take between 2 weeks to 1.5 years (27). In our patient, complete resolution of ophthalmoplegia occurred in about 2 weeks but residual ptosis remained and visual acuity did not improve due to extensive rubiosis iridis, obscuration of pupil by membrane andcataractous changes in lens furthermore, prevented fundus evaluation for any related retinal vascular disease like CRVO or CRAO which might have led to neovascularization.

While searching for this diseased condition, we came across few more rare complications of HZO, which are as follows-

1) Isolated Internal Ophthalmoplegia was reported by karti et al., where the patient had developed Anisocoria and in the affected eye pupillary reaction was not there. The rest of the external ocular structures and fundus were normal (28)
2) Neovascular glaucoma due to HZO, was reported by Ahmad SS etall (25)
3) Optic neuritis, a rare complication was observed by de Mello Vitor B etall. (29)

Conclusion-

Herpes zoster opthalmicus in a child is a very rare condition. All the treating doctors must take into account that this disease can lead to severe complications in the child and the elderly and it should be promptly treated. While going through so many articles we found that a case of herpes zoster opthalmicus should be followed for at least 3 weeks after vesicular eruption as all the major complication occurs at around 3 weeks. and not only the cornea but the fundus and angle structures should also be monitored vigilantly for early detection of any sequelae of the disease .our patient

developed complications only after 3 weeks of vesicular eruption and vesicles were fully dried which shows that the scarring after drying up of vesicles does not mean that the disease is not active and treatment can be stopped. We should first examine all the ocular structures before stopping treatment.

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