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Arnold-Chiari Malformation and It's Curse to the Innocent Eye- a Case Report

Dr. Shivani Bansal[1], Dr. Preeti Rawat[2], Dr. Vijay Bhaisare[2]
[1]*Resident, Department of Ophthalmology, MGM Medical College and M.Y. Hospital, Indore, Madhya Pradesh*
[2]*Professor, Department of Ophthalmology, MGM Medical College and M.Y. Hospital, Indore, Madhya Pradesh*
Corresponding Author: Dr. Shivani Bansal

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ABSTRACT: This paper aims to report a rare presentation of Arnold Chiari Malformation with unilateral Proptosis and endophthalmitis.

KEYWORDS: Arnold Chiari, Proptosis, Enucleation, Endophthalmitis

I. INTRODUCTION

Arnold-Chiari or Chiari malformations consist of a group of deformities of the posterior fossa and hindbrain, consisting of the cerebellum, pons, and medulla oblongata. Other associated intracranial or extracranial defects such as hydrocephalus, encephalocele, syrinx, or spinal dysraphism may be present. Chiari malformation has herniation of posterior cerebellar vermis for more than 3 mm below the foramen magnum. There are four subtypes, and all of them feature herniation of posterior cerebellar vermis.

Chiari malformations are classified according to their morphology and severity of anatomic defects, via imaging. Chiari I is least severe and is characterized by one or both pointed cerebellar tonsils that project 5 mm below the foramen magnum. Chiari II consists of brainstem herniation with a towering cerebellum and an open distal spinal dysraphism/myelomeningocele. Chiari III has herniation of the hindbrain into a low occipital or high cervical meningoencephalocele. Chiari IV is now considered obsolete.

Visual symptoms like blurred vision, photophobia, diplopia, or retro-orbital pain are found in up to 80% of patients with Chiari malformations.[2] Other ocular manifestations are nystagmus (most often down-beat), cranial nerve palsies (most common abducens palsy), transient visual disturbances, acquired comitant esotropia, or alternative hypertropia.[2] They occur secondary to either cerebellar compression, elevated ICP or brainstem compression. Papilledema can occur in association with idiopathic intracranial hypertension (IIH).

II. CLINICAL HISTORY

A 2 year female presented to the OPD with complaint of gradually progressive protrusion of Left eye ball since birth. The birth and prenatal history were unremarkable. Right eye anterior segment examination was normal, and fundus showed disc edema. On examination of the left eye severe proptosis was present. Further examination revealed vision as perception of light, ill sustained pupillary reaction, conjunctiva congested, cornea showed signs of exposure keratopathy. On dilated fundus examination Disc edema was found. Facial features showed antimongoloid slant, hypertelorism, severe midface deficiencies, and enlarged head circumference. Magnetic Resonance Imaging (MRI) was advised which lead to the diagnosis of Chiari Malformation with Syrinx, Craniosynostosis and Hydrocephalus. The patient was then referred to pediatric-surgery department, where she underwent Ventriculo-peritoneal (VP) shunt surgery for hydrocephalus. After 15 days her left eye vision was No Perception of light, with red tender swollen lids, mucopurulent discharge and severe Exposure Keratopathy with worsening proptosis. USG Bscan was done which showed Endophthalmitis with preseptal cellulitis. Broad spectrum systemic antibiotics were started. Routine Blood investigations along with culture and sensitivity were sent. There were no signs of systemic sepsis and the VP shunt site showed no signs of infection. The blood reports were normal, culture and sensitivity reports were negative. There was no response to the antibiotics and the proptosis was worsening. Enucleation of the Left eye was performed as the last resort. The condition improved post enucleation. VP shunt showed good results.

III. CONCLUSION

This was an unusual presentation of Chiari Malformation with severe proptosis and endophthalmitis. The cause of endophthalmitis was unclear. Enucleation was a salvage surgery to

prevent the systemic spread of infection which could have lead to VP Shunt failure. Timely diagnosis and intervention can prevent mortality in such cases.

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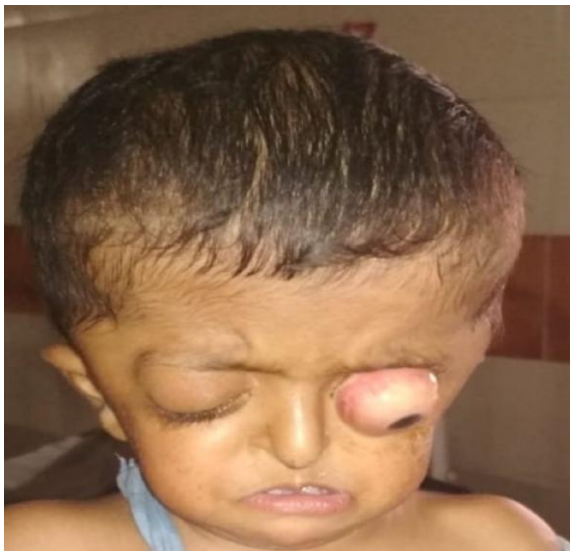

Arnold Chiari Malformation with Proptosis

Post Enucleation Socket