Case Report

Microspherophakia and congenital superior divisional third nerve palsy: A rare presentation

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ABSTRACT

Unilateral microspherophakia without systemic associations is a rare condition and so is congenital palsy of the superior division of the third nerve. Here, we report both these rarities together in a 32-year-old male who had microspherophakia in his right eye and congenital palsy of the superior division of the third nerve in his left eye. The intraocular pressure (IOP) in the right eye was normal with no glaucomatous change in the fundus, but the patient had developed cataractous changes in the microspherophakia lens. The left eye of the patient had low vision due to congenital ptosis which led to stimulus deprivation amblyopia.

Key words: *Amblyopia, Cataract, Microspherophakia, Ptosis*

Microspherophakia is a rare congenital condition in which the crystalline lens is spherical with a decreased equatorial diameter and an increased anteroposterior diameter due to defective development of lens zonules during embryogenesis. The lens has no corticonuclear demarcation in such cases. The entire lens can be easily seen at the slit lamp on dilating the pupil. It is usually bilateral which may be isolated or familial associated with systemic anomalies such as Weill-Marchesani syndrome, Marfan's syndrome, hyperlysinemia, and congenital rubella. Rarely, it is seen unilaterally too [1,2].

Congenital palsy of the third nerve is also a rare condition. Affected individuals often have unilateral involvement and no other neurological abnormality [3]. Clinically, an isolated superior divisional third cranial nerve palsy is characterized by ptosis and limitation of supraduction, particularly with the eye in abducted position [4]. Amblyopia is frequently seen in such cases which require early and aggressive treatment. We report the case of a 32-year-old male who had microspherophakia in the right eye and congenital superior divisional third nerve palsy in the left eye. Fortunately, he had not developed glaucoma in his right eye, but unfortunately, he had developed amblyopia in his left eye.

CASE REPORT

A 32-year-old male presented to the eye Outpatient Department (OPD) at our Zonal Hospital with a history of decreasing vision in his apparently normal right eye for the past 6 months and a history of decreased vision in his left eye since birth. He came to our OPD for issuing of a visual disability certificate. He gave no history of trauma to his left eye or significant viral illness in childhood. He was non-diabetic and non-hypertensive.

The patient was a 32-year-old male adult of average built and height 168 cm. His skeletal development was normal with normal fingers of hands and toes. His IQ was within the normal range. He had no cardiovascular problem. The patient was vitally stable with a pulse of 76/min, blood pressure of 128/82 mmHg, and respiratory rate of 16 cycles/min. On ocular examination, the visual acuity in the right eye was 6/18 (with −2.50D Sph) and in the left eye was finger counting ½ m which showed no improvement with a pinhole. The visual axis showed a hypotropia in the left eye which was nearly 7°. The pupillary size and reaction were normal in both the eyes [Fig. 1]. There was a severe ptosis in the left eye with the upper lid covering the pupil in primary gaze. The ocular movements were normal in the right eye, and there was a restriction of elevation in the left eye more so in the abducted position, as shown in Fig. 2. Levator palpebrae superioris excursion was 11 mm in the right eye and 4 mm in the left eye. The corneal diameter was 11.2 mm vertically and 12.3 mm horizontally in both the eyes.

A slit-lamp examination revealed that the anterior chamber was shallow in the right eye (Van Herick Grade 2) and was normal in the left eye. The iris in the right eye seemed anteriorly pushed forward and was normal in the left eye. On dilating the pupils, the lens in the right eye was seen to be microspherophakia with early cataractous changes and was normal in the left eye [Fig. 3]. The lens power was 17.94D in the right eye and 20.0D in the left eye. The intraocular pressure was 13 and 14 mmHg in the right eye and left eye, respectively. The intraocular lens power (IOP) was 17.94D in the right eye and 17.56D in the left eye. The fundus examination was normal in both the eyes. Gonioscopy showed Grade II angles in the right eye and Grades III and IV in the left eye.

He was kept under close follow-up for his right eye to monitor the IOP and progression of cataract so that early intervention is

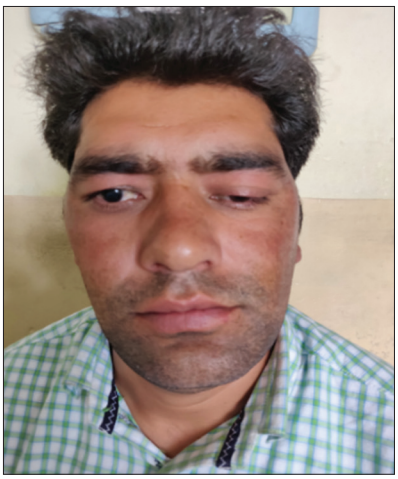

Figure 1: Ptosis and hypotropia in the left eye

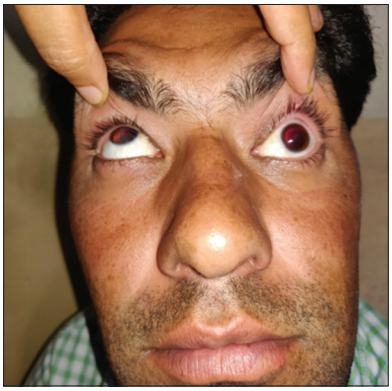

Figure 2: Limitation of supraduction in the left eye

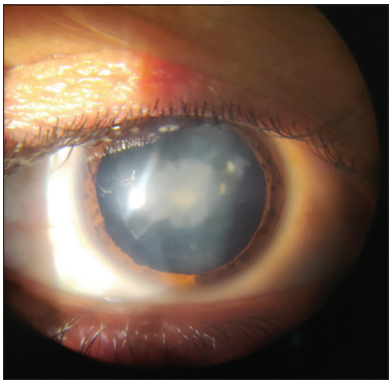

Figure 3: Microspherophakia in the right eye

planned as and when necessary. He was also told to report if felt any sudden pain in the right eye. All we could offer him in his left eye was a cosmetic correction of his ptosis and squint which he refused. The patient was asked to report after 2 months but lost to follow-up.

DISCUSSION

Unilateral microspherophakia without any systemic abnormality is a rare condition [1,5]. Faulty development of lens zonules during embryogenesis is believed to be the cause of microspherophakia [6]. Glaucoma in isolated spherophakia is less common [5]. In our case too, the microspherophakia was unilateral without any systemic association, and there were no glaucomatous changes, though early cataractous changes were present which had caused a gradual diminution of vision in his apparently normal right eye over a period of 6 months. Visual compromise in patients with microspherophakia is attributed to refractive error or secondary glaucoma [7]. This was not the reason for decreased vision in the right eye in our case.

Paralysis of the third cranial nerve can have a wide spectrum of presentation due to its wide innervation of the four extraocular muscles, the levator palpebral superioris, and the sphincter pupillae. The common causes of the isolated third nerve palsy in children are congenital (43%), trauma (20%), inflammation (13%), aneurysm (7%), and ophthalmoplegic migraine [8]. Congenital third nerve palsies are relatively rare and presumed to be due secondary to maldevelopment, intrauterine insult, or birth trauma [9]. They are typically unilateral and isolated. Some degree of ptosis and ophthalmoplegia is a rule and most of them are permanent [10].

In our case, only the superior division of the third nerve was involved and the cause of the palsy was most likely congenital as the patient gave a history of drooping left eyelid since birth and gave no history of trauma or significant viral illness or any other relevant history during childhood. As the patient presented to us at an age of 32 years, there was hardly any scope of restoration of binocular vision as the resultant amblyopia had already set in at an early age.

CONCLUSION

We can say that isolated microspherophakia presented coincidentally with superior divisional congenital third nerve paresis in the other eye. Both the entities are not related to each other but presented irrespective of each other.

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