Unusual presentation of more common disease/injury

Unilateral tilted disc and ipsilateral keratoconus in the same eye

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Summary

The objective of this case was to report unilateral tilted disc in a boy with ipsilateral keratoconus. The tilted disc syndrome is a non-hereditary bilateral condition. This configuration is accompanied by situs inversus of the retinal vessels, congenital inferonasal conus, thinning of the inferonasal retinal pigment epithelium and choroid, and myopic astigmatism. Unilateral tilted disc syndrome is a rare condition. Keratoconus is a disorder characterised by progressive corneal steepening. The author present a case of unilateral tilted disc in a boy with ipsilateral keratoconus. Pterygium is a common disorder and tilted disc syndrome is a bilateral condition. But unilateral tilted disc in a boy with ipsilateral keratoconus is the first report in literature. Due to these clinical presentation, this report is an exception in literature and reported an unknown clinical coincident.

BACKGROUND

The tilted disc syndrome is a non-hereditary bilateral condition in which the superotemporal optic disc is elevated and the inferonasal disc is displaced posteriorly, which results in an optic disc of oval appearance with its long axis obliquely orientated.¹ This configuration is accompanied by situs inversus of the retinal vessels, congenital inferonasal conus, thinning of the inferonasal retinal pigment epithelium and choroid, and myopic astigmatism. These features presumably result from a generalised ectasia of the inferonasal fundus that involves the corresponding sector of the optic disc.

Keratoconus is a disorder characterised by progressive corneal steepening, most typically inferior to the centre of the cornea, with eventual corneal thinning, induced myopia and both regular and irregular astigmatism. The prevalence of keratoconus in the general population appears to be relatively high. The diagnosis of keratoconus can be made on the basis of observation of the presence of keratometry >47.20 D.² In the past, it was believed that more than 90% of cases were sporadic. But later on, the studies show keratoconus in some family members, which suggests an autosomal dominant pattern of inheritance. Ocular-associated disorders include Leber's congenital amaurosis, retinitis pigmentosa, Fuchs' dystrophy and posterior polymorphous dystrophy have been reported as well. Given the relatively high prevalence of keratoconus in the general population, some of these associations may be coincidental.³

I present a case of unilateral tilted disc in a boy with ipsilateral keratoconus. The tilted disc has an atypical form in which the temporal optic disc is elevated and the nasal disc is displaced posteriorly, which results in the optic of oval appearance with its long axis vertically orientated.

Figure 1 Topography of right eye is normal, topography of left eye show keratoconus.

Figure 2 Perimetry of right eye is in normal limit but perimetry of left eye is outside normal limit.

Figure 3 Fundus photography of the left eye: situs inversus of the retinal vessels showed by thin arrow, tilted disc showed by thick arrow and thinning of the nasal retinal pigment epithelium and choroid showed by arrowhead.

This configuration is accompanied by situs inversus of the retinal vessels and thinning of the nasal retinal pigment epithelium and choroid.

CASE PRESENTATION

A 13-year-old boy presented with diminished vision. There was no significant ocular or medical history. The birth history was normal.

His uncorrected distance visual acuity on Snellen's chart was 20/20 in the right eye and 5/100 in the left eye.

Keratometric measures were 43.25 D at axis of 110° and 43.75 D at axis of 20° at the right eye, 48.0 D at axis of 85° and 44.25 D at axis of 175° at the left eye.

SimK1 was 43.20 D at axis of 98 and SimK2 was 43.89 D at axis of 8 at the right eye, SimK1 was 47.80 D at axis of 73 and SimK2 was 43.55 D at axis of 163 at the left eye (figure 1).

Central 30-2 threshold test of the left eye was outside normal limits (figure 2).

Fundus examination revealed normal fundus appearance at the right eye and tilted disc accompanied by situs inversus of the retinal vessels and thinning of the nasal retinal pigment epithelium and choroid (figure 3).

DISCUSSION

The cornea and the optic disc form the anteroposterior opening of the sclera. Oblique entrance of optic disc to sclera accompanied by myopic astigmatism. But tilted disc is generally bilateral condition¹ and association with keratoconus has not been reported yet.

Diseases of posterior segment lesion reported in association with keratoconus are numerous including bilateral macular coloboma, gyrate atrophy, Leber's congenital amaurosis, retinitis pigmentosa, retinal dissection syndrome and retrolental fibroplasia.³

Here, I reported unilateral tilted disc and ipsilateral keratoconus in the same eye. Due to this clinical presentation, this report is an exception in literature and reports an unknown clinical coincident.

Learning points

- Tilted disc syndrome is a non-hereditary bilateral condition and unilateral tilted disc syndrome is a rare condition.
- The prevalence of keratoconus in the general population appears to be relatively high but in children it is a rare condition.
- The cornea and the optic disc form the anteroposterior opening of the sclera. Oblique entrance of optic disc to sclera accompanied by myopic astigmatism. But tilted disc is generally bilateral condition and association with keratoconus has not been reported yet.

Competing interests None.

Patient consent Obtained.

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