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Dural Ectasia of Optic Nerve – An Important Clinical Finding in Marfan's Syndrome

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Abstract

Marfan's Syndrome (MS) is a hereditary, systemic, connective tissue disorder having multisystem involvement. Ghent Nosology proposed new criteria for the diagnosis of MS of which dural ectasia in the lumbosacral region is one of the diagnostic criteria. We report the findings of optic nerve sheath dural ectasia in three brothers of the same family with MS. We propose that dural ectasia of the optic nerve sheath (ONSDE) be considered as one of the markers of the disease, like dural ectasia of the lumbosacral region.

Keywords

Marfan's syndrome; dural ectasia; criteria; optic nerve sheath

Abbreviations

MS: Marfan's Syndrome; ONSDE: dural ectasia of the optic nerve sheath; BCDVA: best corrected distant visual acuity; MRI: Magnetic resonance imaging; USG: Ultrasonography; CT: Computed tomogram; Sph.: spherical; Cyl.: Cylinder

Introduction

Marfan's syndrome (MS) is a genetic connective tissue disorder with variable systemic manifestations involving the eyes, dural sac surrounding the spinal cord, the skeleton, the lungs, and the hard palate. Optic nerve sheath dural ectasia is a rare structural anomaly occurring either as a primary entity or associated with other local or systemic conditions [1]. In 2010, the Ghent nosology proposed new criteria to lead to a diagnosis of MS. Dural ectasia in the lumbosacral region is one of the major diagnostic criteria of MS [2]. However, dural ectasia of the optic nerve sheath (ONSDE) in patients with MS has been described only once before [3]. We report the findings of optic nerve sheath dural ectasia in a family of three brothers with MS and propose that ONSDE be considered as one of the markers of the disease, like dural ectasia of the lumbosacral region.

Case Presentation

Patient 1: An 11-year-old boy presented to us with blurred vision in both eyes (BE) and a past history of headache since many years. He had a past medical history of bronchial asthma and inguinal hernia surgery 2 years back, and left eye (LE) trabeculectomy surgery 1 year back, elsewhere. The medical records of the patient revealed that the patient had undergone a trabeculectomy for a secondary

glaucoma in the LE. Systemic review highlights were elongated facies, high arched palate, marfanoid habitus, and an arm span of 139 cm versus a height of 130 cm. There was no evidence of heart disease, vertebral abnormalities, wrist sign or thumb sign. He had two brothers (one younger and one older to him) and a sister and the detailed history revealed that the two brothers were also suffering similar kind of visual problems. His best corrected distance visual acuity (BCDVA) was 6/18 with -10 D Sph. / -3.50 D Cyl. X 40 in the right eye (RE), and counting fingers at 1 meter in the LE. He had a small failing bleb with slight corneal haze and scarring along with cataract in the LE. The anterior chamber was extremely shallow in the RE with a lens that looked microspherophakic and a nearly flat anterior chamber in the LE. BE had a patent peripheral iridectomy. A visible posterior pushing mechanism by the lens was evident in BE. A dilated exam could not be done at that time, as the pupils were non-dilating. Ultrasonography (USG) of the LE revealed a normal posterior segment, but a cystic mass was noted in each eye in the optic nerve region immediately behind the globe (Figure 1.d). Magnetic resonance imaging (MRI) of the brain and orbits revealed tube like enlargement of the anterior portion of the orbital optic nerve of BE (Figure 1.a, b), isointense with cerebrospinal fluid. A neuroradiologist opinion labeled them as dural ectasia of the anterior optic nerve sheath, immediately behind the globe. He underwent lensectomy with anterior vitrectomy in the RE for the subluxated lens. Dilated fundus exam of the RE post operatively was within normal limits, with no evidence of optic nerve head edema.

Patient 2: His 15- year-old elder brother also presented to us with the similar complaint of diminution of vision which was there for many years with a similar history of being operated for inguinal hernia at the age of 2 years. Systemic examination revealed elongated facies, high arched palate, positive wrist sign and thumb sign, and an arm span of 175 cm versus a height of 165 cm. There was no evidence of heart disease or vertebral abnormalities. His BCDVA was 6/18 with - 25 D Sph. / - 6 D Cyl. X 110 in the RE and 6/9 with - 17 D Sph. / - 3 D Cyl. X 80 in the LE. The edge of the subluxated lens in his RE could be seen through the undilated pupil (Figure 2.a). Posterior segment examination was within normal limits. He also underwent lensectomy with anterior vitrectomy under general anesthesia in the RE, and achieved a 6/9 vision in that eye with a contact lens. After the dural ecstasias were noted in his younger brother, USG and MRI of the brain and orbit were ordered that revealed dural ectasia of the anterior optic nerve sheath in BE (Figure 2.b,c). The dural ectasia was larger in the RE compared to the LE.

Patient 3: The younger brother aged 7 years who was also having visual problems was also brought for an examination by the father after having his two sons operated with us. He was also found to have ocular and systemic features of MS and a previous Computed tomogram (CT) scan done showed bilateral dural nerve ectasia (Figure 3.a, b). An orbital MRI was done which confirmed the findings of the CT scan.

Lumbar dural ectasia being major Ghent criteria for diagnosis of MS, an MRI of the lumbar area was done for all the three brothers and only the eldest brother was found to have a dural ectasia of the lumbar region where as the other brothers had no signs of lumbar dural ectasia (Figure 2.d & 3.c).

Discussion

ONSDE has been known to occur either primarily, or secondarily in association with orbital lesions such as meningioma, glioma, vascular hamartoma, neurofibromatosis, hemangioendothelioma, Von-Hippel-Lindau disease, Hajdu-Cheney syndrome, Arnold-Chiari malformations and Idiopathic intracranial hypertension [4]. To the best of our knowledge, only one other case has been reported in

association with MS [3].

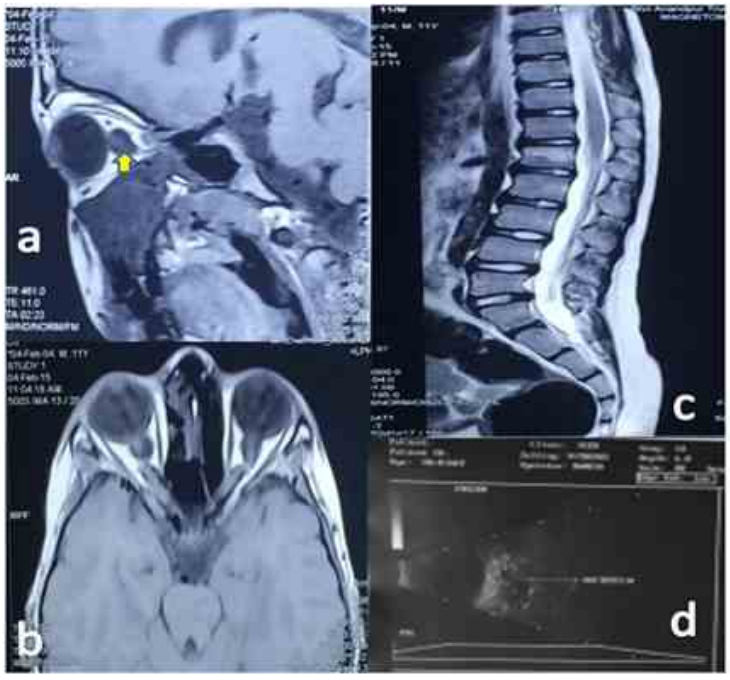
Structurally, it is a saccular dilatation of the meninges surrounding the orbital portion of the optic nerve, filled with cerebrospinal fluid. Histopathologically, other than the cyst, the meninges appear to be normal [5-7]. It can present with blurred vision, pain, proptosis, an enlarged blind spot, visual field changes, optic nerve head edema, choroidal folds, optic disc shunt vessels [1,5,8]. None of these symptoms or signs was seen in the cases reported by us, the dural ectasia being incidental findings.

MRI with fat suppression and high spatial resolution clearly shows tube like enlargement of the optic nerve sheath isointense with cerebrospinal fluid [6]. The MRI also allows its differentiation from other optic nerve masses. In our cases, the dural ectasia were first noted on USG, and later confirmed by MRI. USG may thus be used as a quick and less expensive procedure to look for dural ectasia of the anterior optic nerve in patients with MS.

Treatment is aimed at relieving optic nerve compression and its features if present. Corticosteroids and systemic carbonic anhydrase inhibitors have been used previously, with variable success. Surgical decompression can be considered for cases with persistent compression [8]. Since there were no symptoms or signs of optic nerve compression in our cases, no intervention was planned, and the brothers have been kept under observation.

Dural ectasia of the lumbosacral region is considered one of the major criteria in the diagnosis of MS using the Ghent nosology, and is prevalent in 63-92% of patients [2]. In patients with MS, defective fibrillin may lead to weaker connective tissue support and in the lower spine where, due to the effects of gravity, the dura may be more predisposed to developing ectasia. However, since the effect of gravity does not play any role in the region of the optic nerves, dural ectasia of this area cannot be explained by the same mechanism. Perhaps a normal level of cerebrospinal fluid pressure may lead to ectasia in these individuals with weak tissue support.

Figures



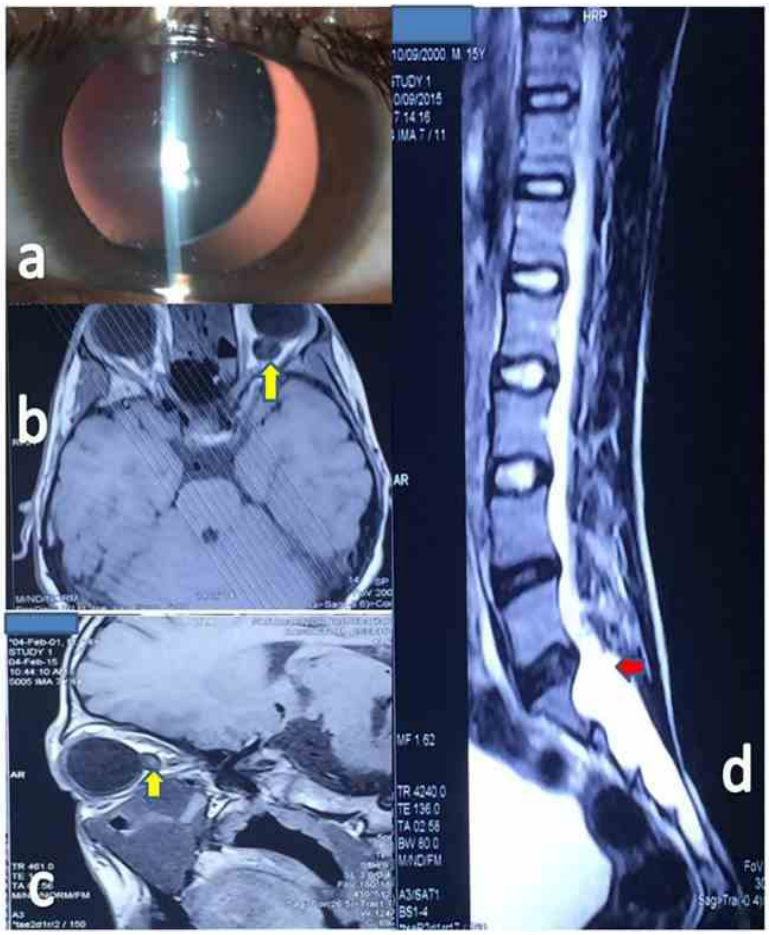
(a) Sagittal section showing ONSDE (yellow block arrow).

(b) Axial scans showing dural ectasia of both optic nerve sheath.

(c) MRI of the lumbosacral region not showing the dural ectasia of the lumbosacral region.

(d) USG of the RE orbit showing the cystic lesion suggestive of the dural ectasia of the optic nerve.

Figure 1: Imaging in Patient 1



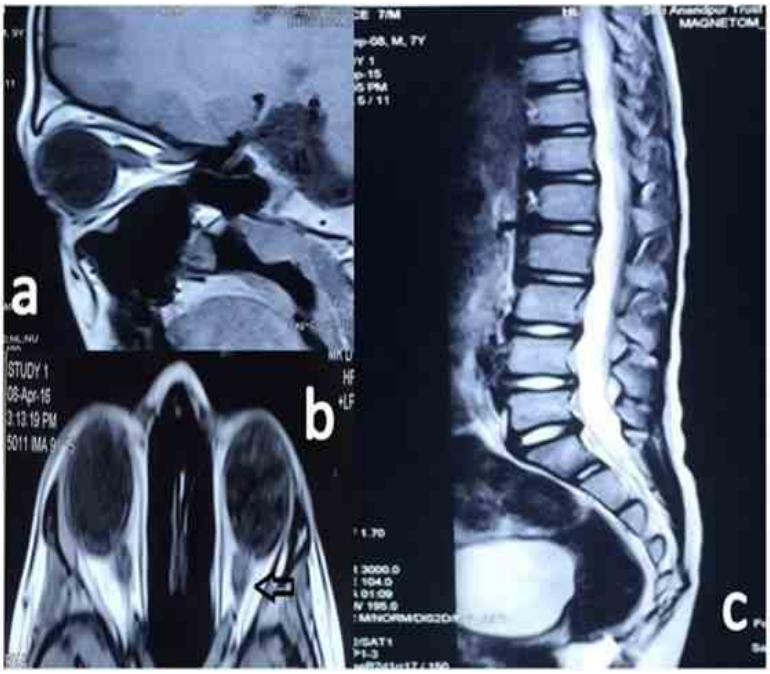
(a) Slit-lamp photography of the RE showing ectopia lentis and temporal subluxation of the lens.

(b) Axial scans showing ONSDE(yellow block arrow).

(c) Sagittal section showing ONSDE(yellow block arrow).

(d) MRI of the lumbosacral region showing the dural ectasia of the lumbosacral region (red block arrow)..

Figure 2: Imaging in Patient 2



(a,b) Axial and saggital scans showing ONSDE (black arrow).

(c)MRI of the lumbosacral region not showing dural ectasia of the lumbosacral region.

Figure 3: Imaging in Patient 3

Conclusion

In conclusion, ONSDE is a less often looked for anomaly in cases of suspected MS. USG of the orbit may easily rule out any dilatation of the anterior orbital part of the optic nerve in such cases. The optic nerve sheath may be considered as one of the regions to look for dural ectasia, as part of the work-up of these patients.

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