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Endoscopic optic nerve fenestration amongst pediatric idiopathic intracranial hypertension: A new surgical option

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ABSTRACT

Objective: To assess the effectiveness of transnasal trans-sphenoid endoscopic optic nerve fenestration for the reversal of vision loss in pediatric idiopathic intracranial hypertension (IIH).
Material and methods: This is a single center observational retrospective case series. Fifteen diagnosed pediatric patients of IIH satisfying the modified Dandy criteria and reported to the out-patient services of otolaryngology, Post Graduate Institute of Medical Education and Research (PGIMER), Chandigarh, India were included in this study. All children underwent thorough clinical examination, complete neuro-ophthalmological work up including visual acuity (V/A), visual field charting(V/F), fundus venogram and radiological work up with MRI for special optic nerve sections in sagittal reconstruction. cerebro-spinal fluid pressure (CSF) measured pre operatively for all children. Standard endoscopic optic nerve sheath fenestration was performed on all children. Visual improvement was assessed by comparing pre-operative ophthalmological findings.
Results: Improvement in vision was taken as a positive outcome. Vision improved in all except two children, who had pre-existing optic nerve atrophy.
Conclusion: Endoscopic optic nerve fenestration is an effective minimally invasive procedure to revert visual loss in pediatric Idiopathic Intracranial Hypertension.

Introduction

Idiopathic intracranial hypertension (IIH, also known as pseudotumor cerebri) is a disorder associated with increased intracranial pressure without clinical or radiological evidence of a space-occupying lesion and with normal cerebrospinal fluid (CSF) constituents [1–5]. The diagnosis is established according to the modified Dandy criteria [6]. The incidence of IIH in the general population is 0.9:100,000 [5]. It is well described in the adult population, and is usually associated with female gender, obesity, and child-bearing age. The natural history of IIH is unknown. In some cases, it is self-limited, while in others, the intracranial pressure remains elevated for many years, even if systemic and visual symptoms resolve. The effects of self limited IIH on the visual system may be catastrophic; approximately 25% of patients may have significant visual impairment and about 10% cases with bilateral visual loss [7,8]. The condition is relatively rare among young children, and only few studies have described it in the pediatric population. Some studies have suggested that the clinical profile of children with IIH may be different than that of their adult counterparts and that the precipitating factors may be considerably different as well [7,9,10]. Treatment modalities vary from nonsurgical to surgical options. Nonsurgical options include use of acetazolamide, furosemide, oral corticosteroids, oral glycerol, and repeated lumbar punctures [11]. Surgical modalities vary from minimally invasive endoscopic optic nerve sheath decompression (ONSD) to shunting procedures to medial or lateral orbitotomy and trans-conjunctival ONSD to sub-temporal craniotomy [12–17]. The author A.K.G. was the first to report endoscopic endonasal ONSD in the world literature [12]. The present study aimed at showing effectiveness of Optic Nerve sheath fenestration (ONSF) to reverse vision lost and to prevent further deterioration in all those cases who failed medical treatment which is less invasive and less morbid.

Material and methods

This is a retrospective single center observational study conducted in the department of Otolaryngology, Post Graduate

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Institute of Medical Education and Research (PGIMER), Chandigarh, India between 2002 and 2011 including 15 cases of proven pediatric idiopathic intracranial hypertension (IIH) fulfilling the modified Dandy's criteria (Table 2).

Inclusion criteria

After getting approval from the Institutional Review Board (IRB), children younger than 15 years, normal brain MR imaging with or without signs of elevated intracranial pressure, intracranial pressure greater than 250 mm H_2O with normal cerebrospinal fluid content, edema of the optic disc, and a non-focal neurologic examination were considered in the inclusion criteria.

Exclusion criteria

Children with concurrent illnesses such as dural sinus thrombosis, otitis media, traumatic intra-parenchymal or sub-arachnoid hemorrhage, systemic lupus erythematosis, Lyme disease and renal failure were excluded.

Apart from routine blood investigations all the children were subjected to an initial pre-operative complete neuro-ophthalmological work up including CSF pressure measurement, visual acuity, slit lamp examination, applanation tonometry, fundoscopy, perimetry visual field charting to know the pattern of visual field defect, and fundus venogram. Patients were subjected to computed tomography (CT) scan and magnetic resonance imaging (MRI) of the brain to rule out any neurologic factors of raised intracranial pressure (ICP). MRI imaging with special sagittal sections for optic nerve to see the characteristic findings of flattening of posterior orbit dilated tortuous optic nerve. MR venography was performed in all cases to look for any venous sinus abnormality, and CT scan of the paranasal sinuses was performed to look for the DeLano type of optic nerve and the degree of sphenoid pneumatization.

All children were assessed for postoperative improvement in vision and perimetry. Postoperative assessment of cerebrospinal fluid (CSF) pressure was performed in all the cases. The cases were followed up for a period ranging from 3 months to 45 months to look for further improvement or deterioration in vision or perimetry.

Children with visual deterioration or intractable headache despite maximum medical treatment were referred for surgery from the department of pediatric neurology.

Surgical technique

Standard trans-nasal trans-ethmoidal endoscopic optic nerve fenestration was performed using 0° rigid nasal endoscope under hypotensive general Anesthesia. Skeletonization of ethmoidal gallery after medialization of middle turbinate was done to expose the lamina paparacae (LP) followed by removal of posterior 1/3rd of LP to get optic tubercle into the view (Fig. 1(a)). Further Optic tubercle was drilled to expose the optic canal and internal carotid artery along with Carotico-Optic recess in the lateral wall of sphenoid (Fig. 1(b)). Thin bone of Optic canal was removed using periosteum elevator in its entire course till it enters into skull base (Fig. 1(c)). Two separate windows were made one at the level of annulus of Zinn and the second one in the course of optic nerve to incise the sheath without damaging the nerve fibers (Fig. 1(d)). Out flow of CSF was seen.

Criteria for improvement

Reversal of vision loss, relief from headache and improvement in visual acuity of two or more scales on the Snellen's chart was taken as improvement.

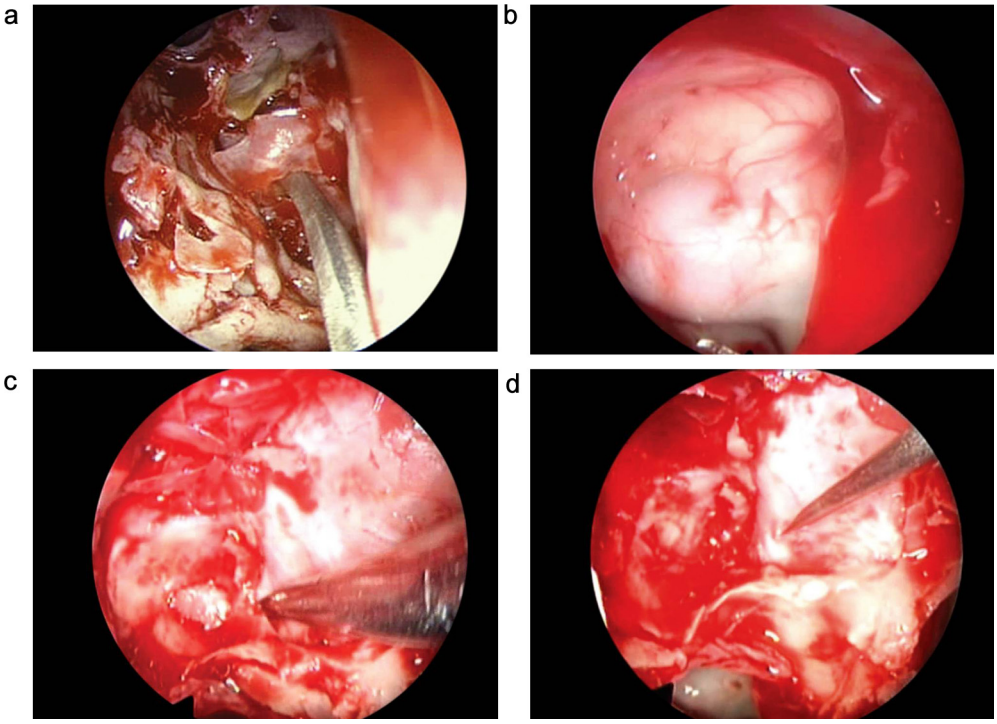

Fig. 1. (a) 0° Rigid endoscopic view of skeletonization of the ethmoids to expose the lamina papyracea. (b) 0° Rigid endoscopic view showing widened anterior face of sphenoid to make posterior ethmoids and roof of sphenoid continuous. (c) 0° Nasal endoscopic view showing a blunt Freer elevator being used to remove the posterior 1/3rd of Lamina papyracea, ~1 to 1.5 cm anterior to the junction of the posterior ethmoids and sphenoid. (d) 0° Nasal endoscopic view of Two separate windows using a sharp sickle knife are made ~3 × 2 mm, one just behind, ~1 mm behind the level of annulus of Zinn and the second one in the course of optic nerve ~3–4 mm before the nerve enters the skull base, by incising the sheath without damaging the nerve fibers.