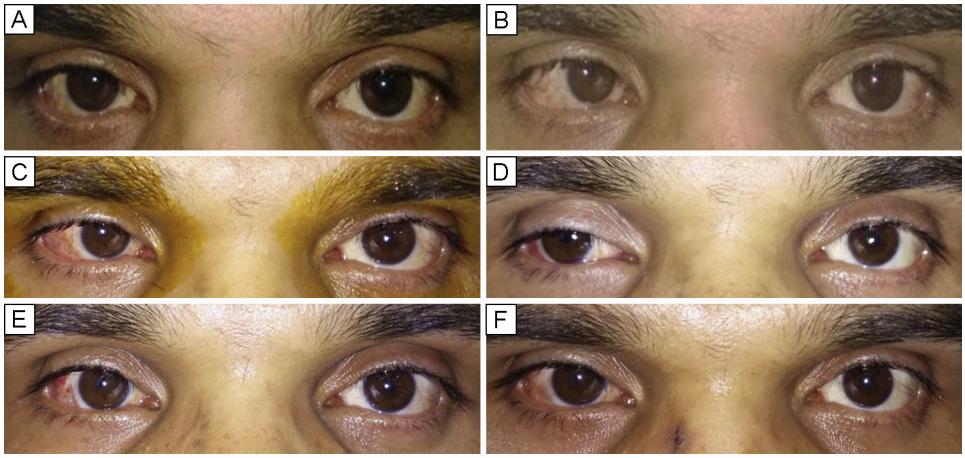

FIG 1. A-B, Cyclic esotropia at presentation. C-D, Deviation present on esotropic days (C) and exotropic days (D). E-F, Orthotropia following the second strabismus surgery seen on former esotropic and exotropic days.

due to the overcorrection of the initial esotropia. The cyclic nature of the strabismus persisted after the first strabismus surgery. The deviation of 45^{Δ} of esotropia alternating with orthotropia changed to esotropia of 20^{Δ} alternating with exotropia of 20^{Δ}. Our surgical plan in the second surgery involved lateral rectus recession of 8.5 mm and medial rectus posterior fixation 13.5 mm from the muscle insertion. The recession over the resected muscle probably acted as a combined recession-resection surgery or faden operation on the lateral rectus muscle, controlling the exodeviation on the exotropic days, with the medial rectus posterior fixation managing the esodeviations on the esotropic days.

Literature Search

PubMed was searched without date restriction on May 15, 2016, using the following terms: *cyclic esotropia*, *cyclic strabismus*, *adult-onset cyclic esotropia*, and *cyclic heterotropia*.

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OCT-documented optic atrophy in nonsyndromic craniosynostosis and lacunar skull

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We report the case of 6-year-old boy who presented with mild redness in the left eye. On fundus examination, disk pallor was noted in both eyes. He did not complain of headache, vomiting, or blurred vision. Three-dimensional computed tomography (CT) imaging was suggestive of craniosynostosis and lacunar skull (lückenschädel). Magnetic resonance imaging findings were suggestive of intracranial hypertension. HD-OCT imaging revealed optic neuropathy in both eyes. The patient underwent sutural release and expansion cranioplasty surgery.

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Submitted May 11, 2016.
Revision accepted August 24, 2016.
Published online January 10, 2017.
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J AAPOS 2017;21:78-81.

1091-8531/$36.00
http://dx.doi.org/10.1016/j.jaapos.2016.08.021

Craniosynostosis refers to premature fusion of a cranial suture, potentially causing characteristic deformations of the skull. It often presents in infancy or childhood due to anomalous skull shape, associated facial anomalies, developmental delay, or signs and symptoms of neurologic impairment or elevated intracranial pressure (ICP), more commonly in syndromic craniosynostosis. Lacunar skull (lückenschädel) is reported in approximately 10% of patients with craniosynostosis.

Case Report

A 6-year-boy of Indian origin presented at Aravind Eye Hospital, Tirunelveli, for evaluation of mild redness in his left eye of 2 days duration. There was no associated headache, blurring of vision, or vomiting. There were no neurological deficits, and growth and development were normal for his age. He had a high forehead, and both eyes had mild proptosis and shallow orbit resulting in scleral show (Figure 1A). On examination, his visual acuity was 20/20 in each eye. Slit-lamp examination was normal for the right eye; the left eye had minimal conjuctival congestion. Dilated fundus examination revealed optic disk pallor with normal vessels in both eyes (Figure 1B). Intraocular pressure by noncontact tonometry was 14 mm Hg in the right eye and 15 mm Hg in the left eye. Color vision (36 plate Ishihara) and automated perimetry visual field testing (Vision Monitor; Metrovision, Perenchies, France) were normal in each eye. Flash visual evoked response showed normal P100 latencies and amplitudes.

All biochemical results, including blood glucose, serum electrolyte (calcium, phosphorus, inorganic phosphorus), renal function, and liver function test were within normal limits. His skull X-ray showed multiple focal areas of radiolucency, bounded by dense bony ridges (Figure 2). Magnetic resonance imaging (MRI) revealed complete fusion of all cranial sutures, with resultant oxycephaly, bilateral prominent perioptic subarachnoid spaces with posterior scleral flattening, vertical buckling of optic nerve, and partial empty sella suggestive of intracranial hypertension (Figure 3). His magnetic resonance venogram showed mild hypolastic left transverse sinus. His MRI revealed normal optic canals, with no optic nerve compression.

Peripapillary retinal nerve fiber layer (RNFL) thickness measured by spectral domain optical coherence tomography (Cirrus HD-OCT; Carl Zeiss Meditec Inc, Dublin, CA) confirmed fundus findings of optic neuropathy. His average RNFL thickness was 37 μm in the right eye and 36 μm in the left eye (eSupplement 1, available at jaapos.org). He was referred to a neurosurgeon, who advised computed tomography (CT) with 3D reconstruction of the skull, the result of which suggested craniosynostosis with universal fusion of all skull vault sutures resulting in oxycephaly and prominent gyral impression on the inner table of the skull vault consistent with lacunar skull (lückenschädel; Figure 4, eSupplement 2, available at jaapos.org). The MRI findings were attributed to intracranial hypertension.

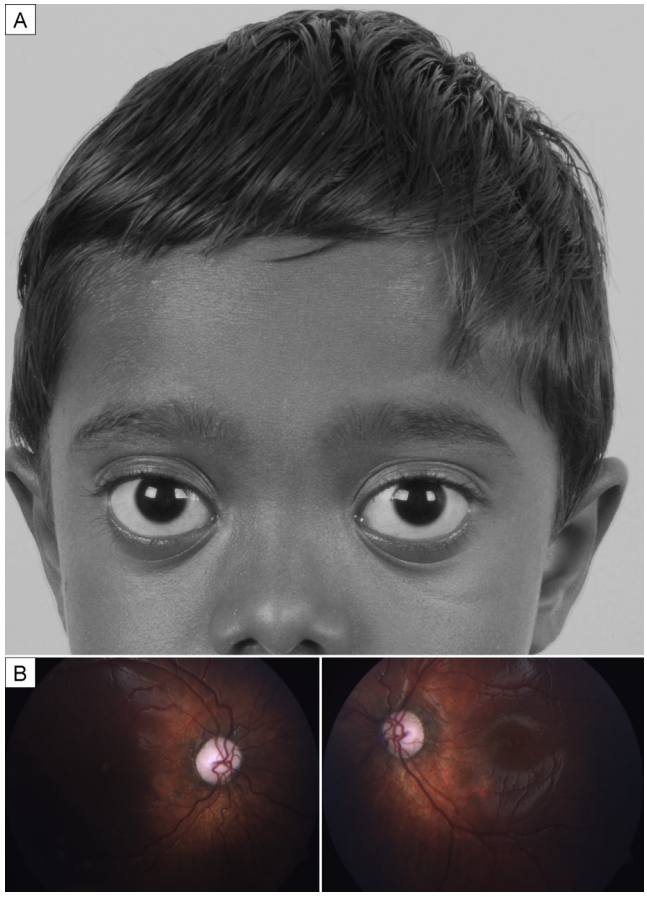

FIG 1. A, Clinical photograph of a 6-year-old boy with mild form of craniosynostosis with high forehead and scleral show in both eyes. B, Fundus images of the right and left eyes showing optic disk pallor, with normal vessels.

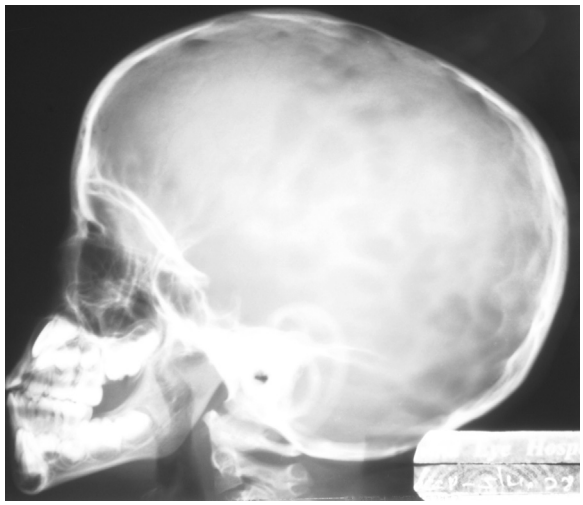

FIG 2. A, Skull X-ray showing multiple focal areas of radiolucency in the skull bounded by dense bony ridges.

The left eye redness disappeared 1 week after decongestant drops were started. He underwent uneventful sutural release and expansion cranioplasty under general

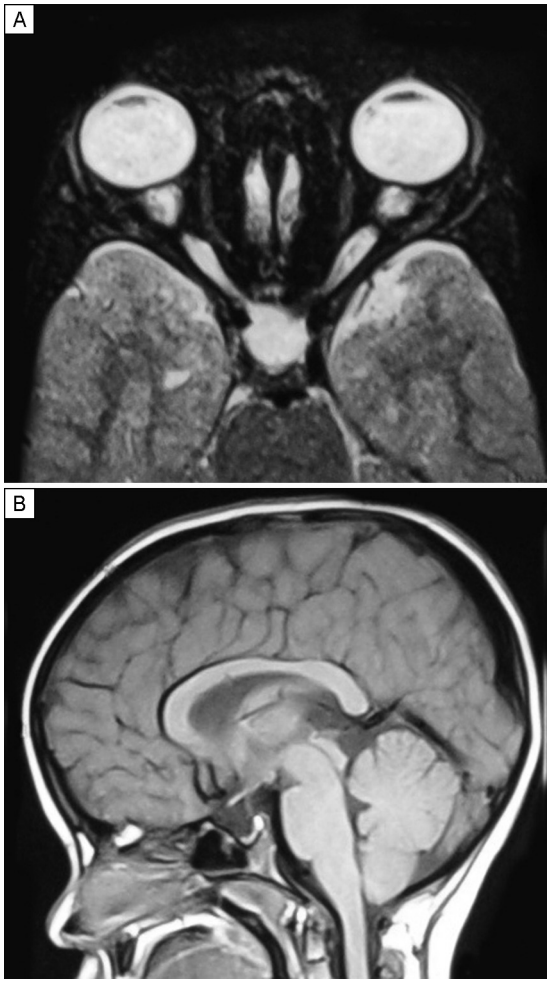

FIG 3. Magnetic resonance imaging showing the vertical buckling of the optic nerve (A), partially empty sella (B), suggestive of raised intracranial pressure.

anesthesia at a tertiary neurosurgery center and recovered well without postsurgical complications.

On follow-up ophthalmologic evaluation 8 weeks after surgery, visual acuity was 20/20 in each eye, with no change in optic neuropathy detected preoperatively.

Discussion

The incidence of nonsyndromic craniosynostosis is approximately 0.4 to 1 in 10,000 live births.[1] The majority of patients with craniosynostosis have various skull alterations, which are severe in syndromic type and are mainly considered to have arisen from compensatory growth of the skull after stenosis of some sutures and high ICP. Lacunar skull results from dysplasia of the membranous skull vault and is typically characterized by multiple, round or oval, radiolucent defects, separated by dense strips of bone (honeycomb-like configuration), which tend to cluster in the cranial vault on plain skull film. It can be a consequence of elevated ICP, or it can be a self-limited phenomenon seen in children with myelomeningocele. It

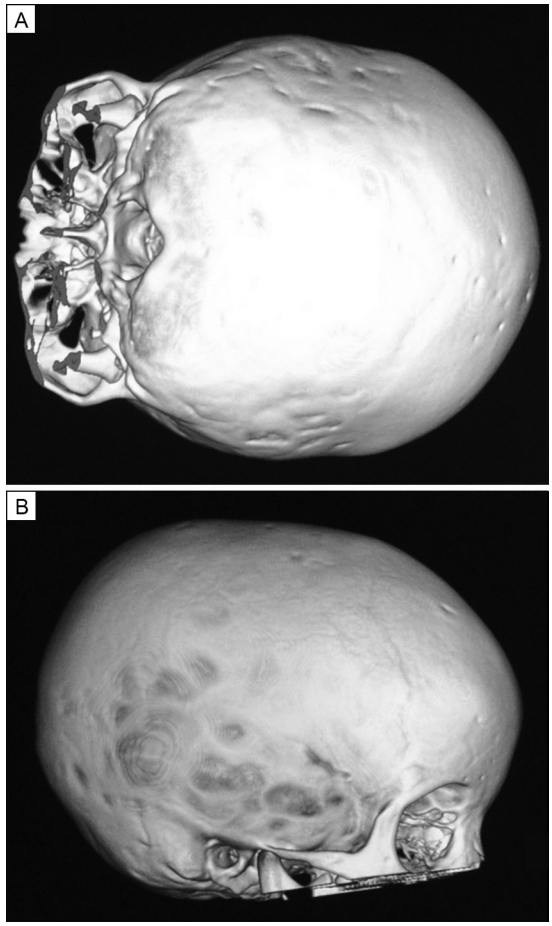

FIG 4. Superior (A) and lateral (B) views of the calvarium on 3D computed tomography scan demonstrating suture fusion and lacunar skull.

has been estimated to occur in 10% in cases of craniosynostosis. Lacunar skull is associated with Chiari malformations (seen in up to 80% of such cases).[1-3] Syndromic craniosynostosis are commonly associated with papilledema and optic nerve atrophy.[1] Diagnosis of a mild form of nonsyndromic craniosynostosis is detected later in childhood or adolescence when symptoms of increased ICP arise, such as headaches and vision changes.[4-8] The literature is uninformative on the percentage of patients with any form of craniosynostosis experience symptoms, especially in late childhood.[7]

Dagi and colleauges[9] reported peripapillary RNFL thickness measured by SD-OCT provides adjunctive evidence for identifying optic neuropathy in patients with craniosynostosis and appears more sensitive in detecting optic atrophy than papilledema.[9] Our patient had decreased peripapillary RNFL thickness in both eyes compared to normative data for Indian children, suggestive of optic atrophy.[10] The optic disk appearance showed pallor in both eyes, but no papilledema. MRI features and OCT findings suggested that intervention was necessary to prevent further loss. This case demonstrates that nonsyndromic craniosynostosis may be occult and insidious,

with subtle facial changes. The patient was asymptomatic, with elevated ICP. His condition was discovered incidentally. The facial appearance of a high forehead with ridging, which was initially not recognized, proved to be important, along with scleral show due to mild proptosis, and shallow orbits. The patient's facial appearance together with optic disk pallor warranted X-ray and neuroimaging.

Visual acuity of 20/20, with preserved visual fields, may be seen in a patient with optic atrophy. Various studies have found children with normal vision and normal visual fields despite a significantly decreased RNFL.[6] No longitudinal studies in pediatric patients have proven the floor values of RNFL with different versions of OCT, which correspond to vision loss and optic disk neuropathy.

In our patient cranial expansion surgery and sutural release was performed to prevent further functional loss of vision. All patients with proven synostosis should be followed closely for asymptomatic elevations of ICP. Case management should involve a multidisciplinary team representing ophthalmology, neurology, and interventional radiology. Careful review of facial features, radiologic images for signs of premature suture fusion, and clinical suspicion must be undertaken to identify craniosynostosis and optic neuropathy; OCT should be performed to investigate possible optic neuropathy. Prompt treatment in cases of elevated ICP in craniosynostosis and lacunar skull is recommended to prevent vision loss in later life.

Acknowledgments

The authors gratefully acknowledge the assistance of Dr. M. Nair, Head of Neurology, Shree Chitra Institute of Medical Sciences, Trivandrum, and Dr. Dilip Panikar, MS, Mch Neurosugeon, Aster Medcity, Cochin, India.

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Nontraumatic orbital roof encephalocele

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Intraorbital meningoencephaloceles occur most commonly as a complication of traumatic orbital roof fractures. Nontraumatic congenital orbital meningoncephaloceles are very rare, with most secondary to destructive processes affecting the orbit and primary skull defects. Treatment for intraorbital meningoencephaloceles is surgical repair, involving the excision of herniated brain parenchyma and meninges and reconstruction of the osseous defect. Most congenital lesions present in infancy with obvious globe and orbital deformities; we report an orbital meningoencephalocele in a 3-year-old girl who presented with ptosis.

Case Report

A 3-year-old white girl presented to the pediatric ophthalmology clinic at Nemours Children's Hospital for progressive left upper lid swelling and drooping. Her medical history was remarkable for molluscum contagiosum involving the left eye 1 year prior; there was no history of significant head trauma or major accidents.

On ophthalmological examination, visual acuity was 20/30 in the right eye and 20/50 in the left eye; refraction was +1.00 sphere in the right eye and plano + 1.00 ×100 by streak retinoscopy. External examination revealed a 9 × 5 × 3 mm area of fullness in the left upper eyelid, with no palpable mass or changes with posture and two remaining molluscum lesions on the temple and cheek from the previous episode (Figure 1A). There was moderate ptosis of the left upper eyelid and a left hypoglobus but no

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Submitted May 26, 2016.
Revision accepted August 27, 2016.
Published online December 16, 2016.

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J AAPOS 2017;21:81-83.

1091-8531/$36.00
http://dx.doi.org/10.1016/j.jaapos.2016.08.020