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Craniosynostosis

Delayed development of frontal mucocele after fronto-orbital advancement in a child with craniosynostosis

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Abstract

Background: Sinus mucoceles rarely develop as a consequence of inadequate sinus ventilation that arises due to inflammation, allergy, polyps, tumors, surgery, and trauma. The development of frontal sinus is delayed until older than 6 years. Therefore, the development of the mucocele in the frontal sinus after fronto-orbital advancement surgery in young children with craniosynostosis may provide essential information for the development of the frontal sinus.

Case Description: We report a rare case of a 22-year-old man presenting with a frontal mucocele manifested by dull headache, proptosis, and diplopia, and which developed 16 years after fronto-orbital advancement surgery for craniosynostosis. Magnetic resonance imaging demonstrated that a multiple cystic mass extended from the frontal sinus to the retro-orbital space along the optic nerve. During surgery, we found that the cyst consisted of mostly thin, yellow mucosa, which developed from an anomalously overdeveloped frontal sinus containing yellow pus-like intracystic fluid. There was no gross local invasion by the cyst. We easily dissected and removed the mucosal cyst from the large frontal sinus completely with frontal sinus obliteration. We cranialized the anomalously large frontal sinus by removal of the posterior wall of the frontal sinus and then widening the ethmoidal drainage with endoscopic ethmoidectomy.

Conclusion: We report the first case of a frontal sinus mucocele that developed after fronto-orbital advancement surgery in the literature and suggest that the mucocele development after fronto-orbital advancement supports the hypothesis of frontal bone-inducing role in frontal sinus development.

Keywords: Craniosynostosis; Development; Frontal sinus; Fronto-orbital advancement; Mucocele

1. Introduction

Frontal sinus mucoceles develop as a consequence of inadequate sinus ventilation that arises due to inflammation, allergy, polyps, tumors, surgery, and trauma [3,9,14,16,18], and it is known that improved ventilation will lead to normally functioning sinus epithelium [5]. Treatment of mucoceles consists of removal of the mucocele by an obliteration procedure and a marsupialization that attempts to enhance ventilation. It has been recommended that in patients who do not have progressively aggravating neurologic symptoms, marsupialization may be the primary mode of treatment, followed by an additional obliteration procedure, if necessary [4,9,14,15].

The frontal sinus differs from other sinuses in that it is absent at birth and has delayed development until older than 6 years [2,8]. From the point of delayed development of the frontal sinus, the development of the mucocele in the frontal sinus after fronto-orbital advancement surgery in children with craniosynostosis may provide essential information for the development of the frontal sinus. Although there have been numerous reports that describe the development of the frontal sinus after fronto-orbital advancement in infants and children [1,12,13], no literature has been reported to date regarding the formation of a frontal sinus mucocele after

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fronto-orbital advancement in infants and children, which may support the hypothesis of the frontal bone origin. The authors of this study report a case of a child with delayed development of a mucocele after fronto-orbital advancement surgery for the treatment of craniosynostosis and suggest that this case supports the theory that the frontal sinus development originates in the frontal bone.

2. Case report

A 22-year-old male patient presented with supraorbital pain of 6 months that gradually increased in severity and accompanied by left proptosis and diplopia (Fig. 1A). The patient had a history of mental retardation and frontal bar advancement including expansion cranioplasty for treatment of multiple craniosynostosis 16 years ago (at 6 years of age). At that time, medical records revealed that the frontal sinus had not developed. At the time of presentation, there was no evidence of neurologic abnormality, and mental retardation had improved slightly to an intelligence quotient of 80. The measured visual acuity was 1.0/0.6, and the right-side visual field was normal, whereas on the left, it was restricted on the nasal side. Simple skull x-rays showed numerous wire fixations and a markedly developed frontal sinus (Fig. 1B). Computed tomogram of the brain demonstrated enlargement of the frontal, sphenoidal, and ethmoidal sinuses and also a defect of the anterior skull base (Fig. 2A). T1-weighted magnetic resonance images revealed a cystic mass, consisting of a slightly low-intensity wall and intracystic fluid of a slightly higher intensity than cerebrospinal fluid, which extended intracranially from the orbital fossa compressing the optic nerve (Fig. 2B, C). The T2-weighted images showed that the cyst wall had a high signal intensity, whereas the internal fluid had an intensity lower than the cerebrospinal fluid and with moderate gadolinium enhancement (Fig. 2D).

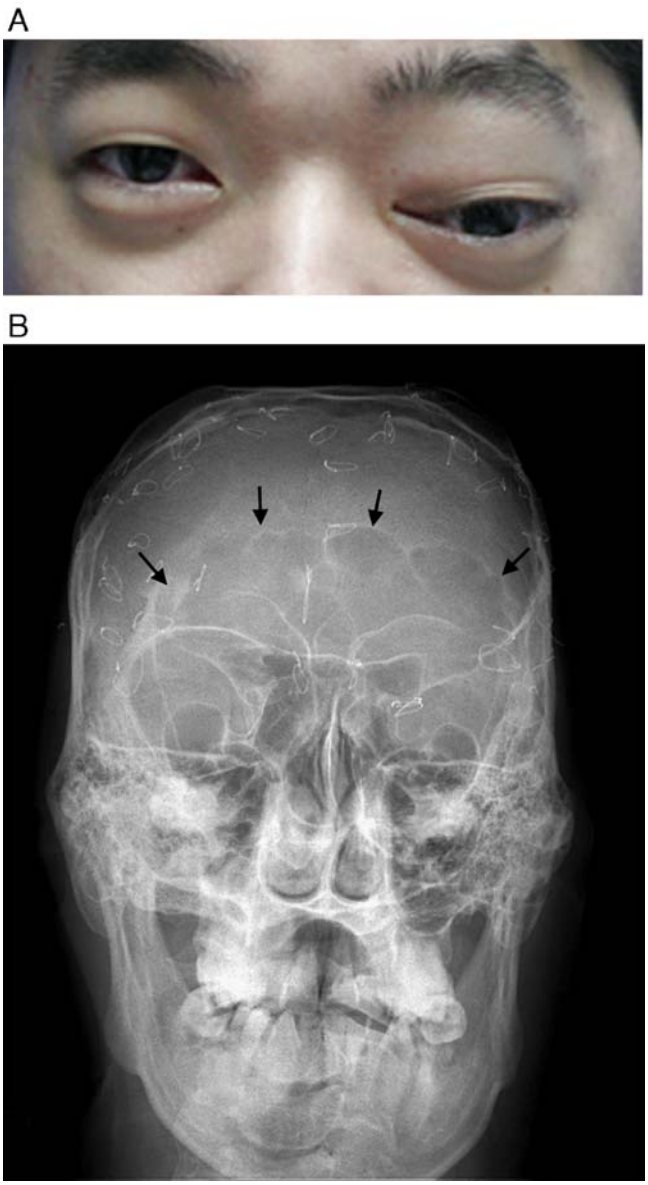

Fig. 1. A: Photograph shows proptosis and protrusion of the supraorbital area in a 22-year-old male patient presenting with dull headache. B: Anteroposterior simple skull x-ray shows a large frontal sinus and multiple wires for bone fixations after fronto-orbital advancement.

A frontal craniectomy was performed and revealed a frontal sinus mucocele with yellowish mucosa and yellow pus-like internal fluid that partially communicated with the ethmoid and sphenoid sinuses (Fig. 3A). Because of the intracranial extension that compressed the optic nerve, the frontal sinus mucosa was completely excised, and the frontal sinus was cranialized with a drainage ostium that was formed by partial resection of the temporal muscle and whole removal of the posterior wall (Fig. 3B). The anterior cranial base was reinforced with a pericranial flap, and endoscopic ethmoidectomy and widening of sphenoid ostium were also performed (Fig. 3C). After surgery, the patient showed improved proptosis, visual acuity, and field, and the pain had disappeared (Fig. 3D).

Pathologic examination confirmed that the lesion was frontal sinus mucocele. At 1 year of follow-up after surgery, there were no abnormal manifestations evident, and the magnetic resonance images also showed no recurrence of disease.

3. Discussion

Mucocele is known to develop from the frontal sinus in 51% to 98% of cases after sinus surgery and less than 1% after frontal craniotomy of routine neurosurgeries [3,14,16-18]. Because 67% to 100% of mucoceles occur several years or decades after surgery, some authors have recommended that patients who have received frontal sinus trauma or surgery should be followed up for at least 5 to 10 years or for their remaining lifetime [2,6,7,19]. Classical treatment of mucoceles consists of transcranial removal of the mucocele by an obliteration procedure that is still recommended especially for patients who have progressive-