Craniosynostosis

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

Soo Han Yoon, MD\textsuperscript{a}, Se-Hyuck Park, MD\textsuperscript{b,\*}

\textsuperscript{a}Department of Neurosurgery, Ajou University School of Medicine, Suwon 443-721, South Korea
\textsuperscript{b}Department of Neurosurgery, Kandong Sacred Heart Hospital, Hallym University, Seoul 134-701, South Korea

Received 28 March 2006; accepted 3 July 2006

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...
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).

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 mucocele 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-
ly aggravating neurologic symptoms, wide adhesion of mucocele with neural contents, and intracranial abscess.

Despite that mucoceles arise as a pathologic consequence of compromised sinus ventilation, it has been demonstrated that draining by marsupialization permits regeneration of the mucosa and normal epithelial movement by ventilation [5]. The marsupialization has been proved efficacious in a study by Har-El [4], who performed such a procedure with endoscopy in 108 patients and showed a very low 0.9% recurrence rate. Although the data necessitates further in-depth studies to ascertain more accurately the possible long-term complications of this procedure, the simple and easy endoscopic marsupialization procedure has been suggested to be the primary mode of treatment [4,9,14,15]. The patient in this study demonstrated a frontal sinus mucocele, which extended intracranially along the optic nerve, and was managed by removal of the mucosa and cranialization to restore optic nerve function as quickly as possible.

Furthermore, endoscopic ethmoidectomy was conducted to reduce possible recurrences.

The frontal sinus develops the latest among the sinuses, is absent at birth, and becomes aerated after 6 years of age [8]. There are, at present, 2 theories with regard to the development of the frontal sinus. One is that it develops because of progressive spread of the mucosa from the middle meatus or from ethmoid sinus, and the other is that it develops as a result of a certain stimulus in the frontal skull that also separates the inner and outer tables of the frontal bone [7,10]. However, in a study by Lykins et al [11], who performed unilateral frontal craniectomy including the frontal sinus and then filling with hydroxyapatite cement in 12-week-old cats, it was observed that the hydroxyapatite cement changed into bone, hence reforming the coronal suture, but there was no redevelopment of the frontal sinus. In a different study, Honig et al [7] reported development of the frontal sinus after orthotopical transplantation of the

Fig. 2. Brain computed tomogram reveals an enlarged and perforated frontal sinus and ethmoid sinus with a bony defect (black arrows) on the anterior skull base (A). Magnetic resonance tomogram shows intracranial extension of the cystic mass from the orbital fossa compressing the optic nerve. T1-weighted images of magnetic resonance tomogram shows slightly low signal intensity with moderate gadolinium enhancement of the cyst wall (white arrows) and low signal intensity of the cystic fluid that is slightly higher than the cerebrospinal fluid (B and C). T2-weighted images shows low signal intensity of the cystic wall and high signal intensity of cystic fluid with displacement of the left optic nerve (white arrows) (D).
frontal bone to the occipital area in 5-week-old infant Goettingen minipigs. He also suggested that the frontal skull, probably the metopic suture, induces frontal sinus development after observing that frontal sinus development was diminished or absent in some patients after craniosynostosis. The above 2 studies suggest that the frontal sinus develops from a certain stimulus of the frontal bone, rather than the hypothesis that postulates the frontal sinus develops because of progressive spread of the mucosa from the middle meatus. However, although it was not mentioned, the results from the above latter study by Honig et al seem to indicate that the orthotopically transplanted frontal bone does not induce the development of a drainage tract. Therefore, although the frontal sinus may originate from the frontal skull, at least a part of the sinus drainage tract may be induced from the middle meatus. Here, if frontal sinus development in humans does originate from the middle meatus, then it seems likely that the mucocele in the frontal sinus could not develop in a child after fronto-orbital advancement because a bony gap between frontal bone and the root of the nose produced because of surgery does not permit the development of the frontal sinus. On the other hand, if the human frontal sinus develops from the frontal bone, the development of the frontal sinus after fronto-orbital advancement is normally progressed, but without sinus tract development, resulting in development of a mucocele. However, although there have been numerous reports for the development of the frontal sinus after fronto-orbital advancement in infants and children [1,12,13], there are no reported cases of frontal sinus mucocele after fronto-orbital advancement, resulting in confusion for the developmental origin of frontal sinus, until the present case. From the present case, we can suggest more concretely that the frontal bone induces development of the frontal sinus after fronto-orbital advancement and bony gap produced by incomplete sinus tract development and compromised drainage and ventilation of sinus, resulting in a mucocele. This present case showed development of frontal sinus mucocele after fronto-orbital advancement, which may be additional evidence to support the frontal bone-inducing hypothesis in the development of the frontal sinus.

4. Conclusion

The authors suggest that this rare mucocele development after the fronto-orbital advancement surgery could support the theory that the frontal sinus originates from the frontal bone.

References


