

Case report

Combined sphenoid and frontal sinus aplasia accompanied by bilateral maxillary and ethmoid sinus hypoplasia

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Abstract. We describe CT scans of a case with bilateral aplasia of frontal and sphenoid sinuses with symmetrical hypoplasia of the ethmoid cellules and maxillary sinuses. This case appears to be first in the English-language literature with these combined findings.

Paranasal sinuses are prone to a great diversity of anomalies. It is important for surgeons to be aware of variations that may predispose patients to increased risk of intraoperative complications and help avoid possible complications and improve success of management strategies. CT is an excellent cross-sectional imaging technique that is used to map sinus anatomy prior to surgical intervention to limit morbidity.

In this report, we present CT findings of a unique association of combined paranasal sinus hypoplasias and aplasias in an adult man that, to the best of our knowledge, has not been reported previously.

Case report

A 25-year-old man was referred to our department with a pre-diagnosis of sinusitis. Coronal and axial CT scans were performed with 5 mm slice thickness and 1 mm increment. Acquired images have delineated that the bodies of the sphenoid and frontal bones were normal and consisted of symmetrical bony structures with a total lack of pneumatization (Figures 1 and 2). These appearances were indicative of aplasias of frontal and sphenoid sinuses. Axial and coronal CT scans also revealed hypoplasias of the ethmoid cellules and maxillary sinuses with a slight nasal septum deviation to the left side (Figure 1 and 2).

There were also mucosal thickenings in maxillary sinuses and ethmoid cells with bilateral air-fluid levels in maxillary antra (Figures 1 and 2). No skeletal, systemic or haematological abnormality was detected on clinical and laboratory examinations. Since paranasal sinus agenesis is frequently seen in cystic fibrosis [1] we tested our patient for that disease. In clinical and laboratory examinations cystic fibrosis was not revealed. After a 10 day medical therapy for acute sinusitis, the patient was relieved from complaints.

Discussion

The development of paranasal sinuses begins as an evagination of the mucosa from the nasal cavities during the third and fourth fetal months. They undergo major expansion after birth, along with the development of the facial cranium and teeth [2]. The underdevelopment or aplasia of the paranasal sinuses is a rare phenomenon that refers mainly to the frontal (12%) and secondarily to the maxillary sinuses (5–6%) [3]. This occurs more frequently in syndromes of craniosynostosis, osteodysplasia (Melnick-Needles), as well as in cases of Down's syndrome (hypoplasia of the frontal sinus) [4]. Agenesis of the sphenoid sinuses is an extremely rare phenomenon. Based on their anatomical studies, Wertheim and Grunwald reported in the early 1900s that agenesis of the sphenoid sinuses can indeed occur in 1–1.5% of cases [5, 6]. Using the same techniques, Peele, in the early 1950s, also referred to the agenesis of the sphenoid sinuses [7]. However, to our knowledge, there is no case reported in the English-language literature with associated aplasia of sphenoid and frontal sinuses which is also accompanied by hypoplasia of maxillary and ethmoid sinuses.

Major studies in the world literature on the prevalence of anatomical features of paranasal sinuses show that the term "anatomical variations" is preferable to "bony abnormalities" [8]. Previous anatomical studies have led to the identification of sphenoid sinus aplasia in an estimated 1–1.5% of cases [5–7]. However, these studies, most of which date back to the first half of the 20th century, lack the support of CT. We have not encountered any case in the literature regarding aplasias of the multiple paranasal sinuses identified by CT.

Eggesbo et al [1] reported the developmental anomalies of paranasal sinuses in cystic fibrosis patients by comparing them with non-cystic fibrosis controls with inflammatory sinusitis. They reported five control subjects with frontal sinus aplasia (4%) and 6 bilateral maxillary sinus hypoplasias (5%) in their 126 controls at 12 years old and over. No sphenoid sinus aplasia were reported in patients or controls from all age groups in their study.

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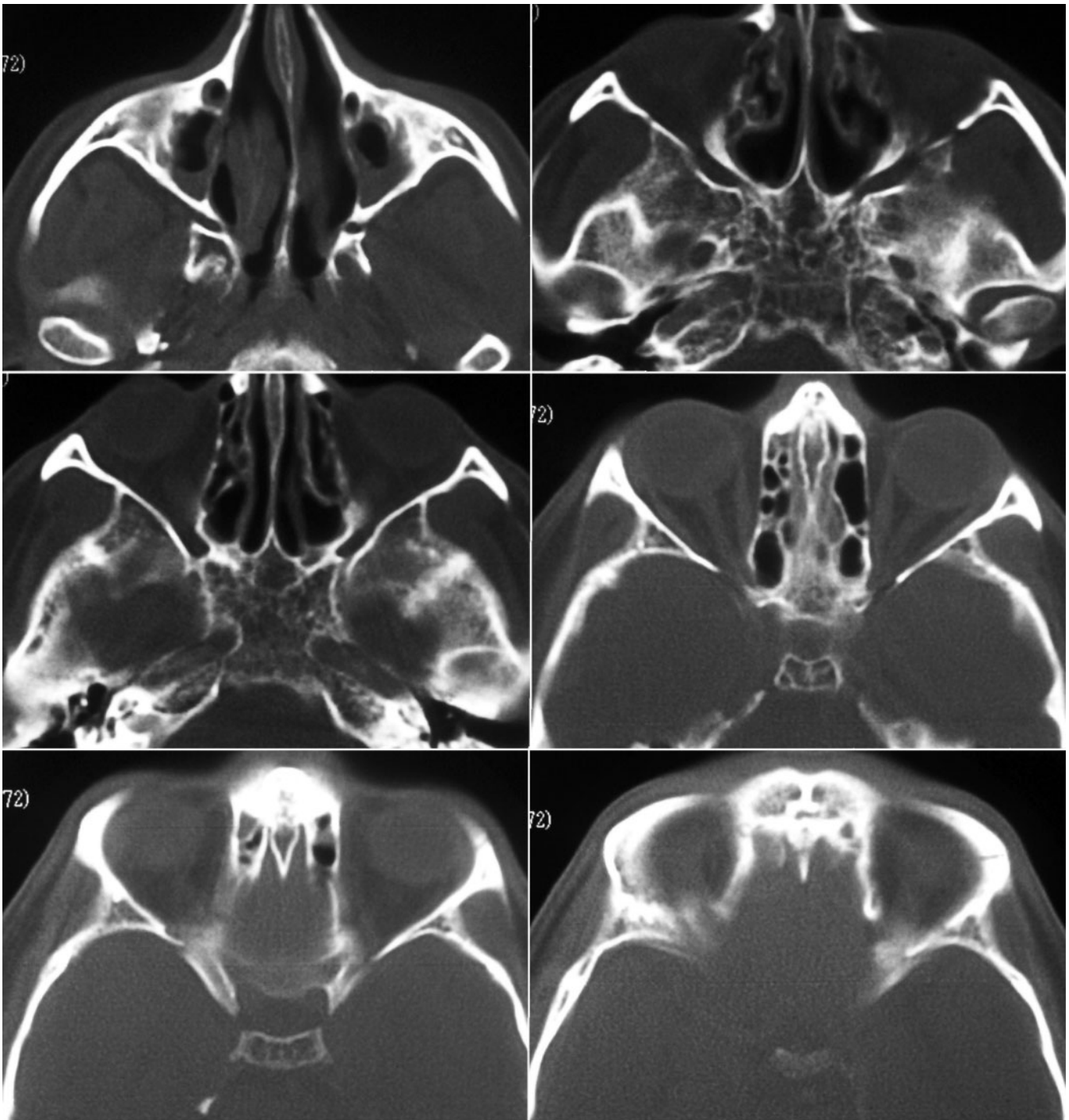


Figure 1. Consecutive axial CT slices of paranasal sinuses demonstrates the lack of pneumatization in expected locations of sphenoid and frontal sinuses and bilateral hypoplastic maxillary and ethmoid sinuses. A slight deviation of nasal septum towards the left side and bilateral soft tissue densities in maxillary sinuses are noticed (top left).

The diagnosis of sphenoid sinus hypoplasia is potentially important in patients in whom trans-sphenoidal hypophysectomy is contemplated. Kesgin et al [9] have identified sphenoid sinus aplasia in an adult woman using CT. No additional sinus abnormality was reported in their case. Another case with CT scans, which has the diagnosis Hand-Schuller-Christian disease, has been reported as well [10]. Our patient did not have any other skeletal or craniofacial anomaly.

Maxillary sinus hypoplasia is an uncommon condition that may be misdiagnosed as chronic sinusitis

[11]. Bolger et al found the prevalence of unilateral hypoplastic maxillary sinus to be 10.4% on coronal CT scans [12]. Maxillary sinus hypoplasia predisposes to orbital penetration during endoscopic sinus surgery; therefore this bony abnormality must be recognized as well as associated anatomic variations, especially prior to sinus surgery.

In conclusion, our case seems to be the first having combined aplasias of the sphenoid and frontal sinuses with hypoplastic ethmoid and maxillary sinuses without any systemic or skeletal disease.

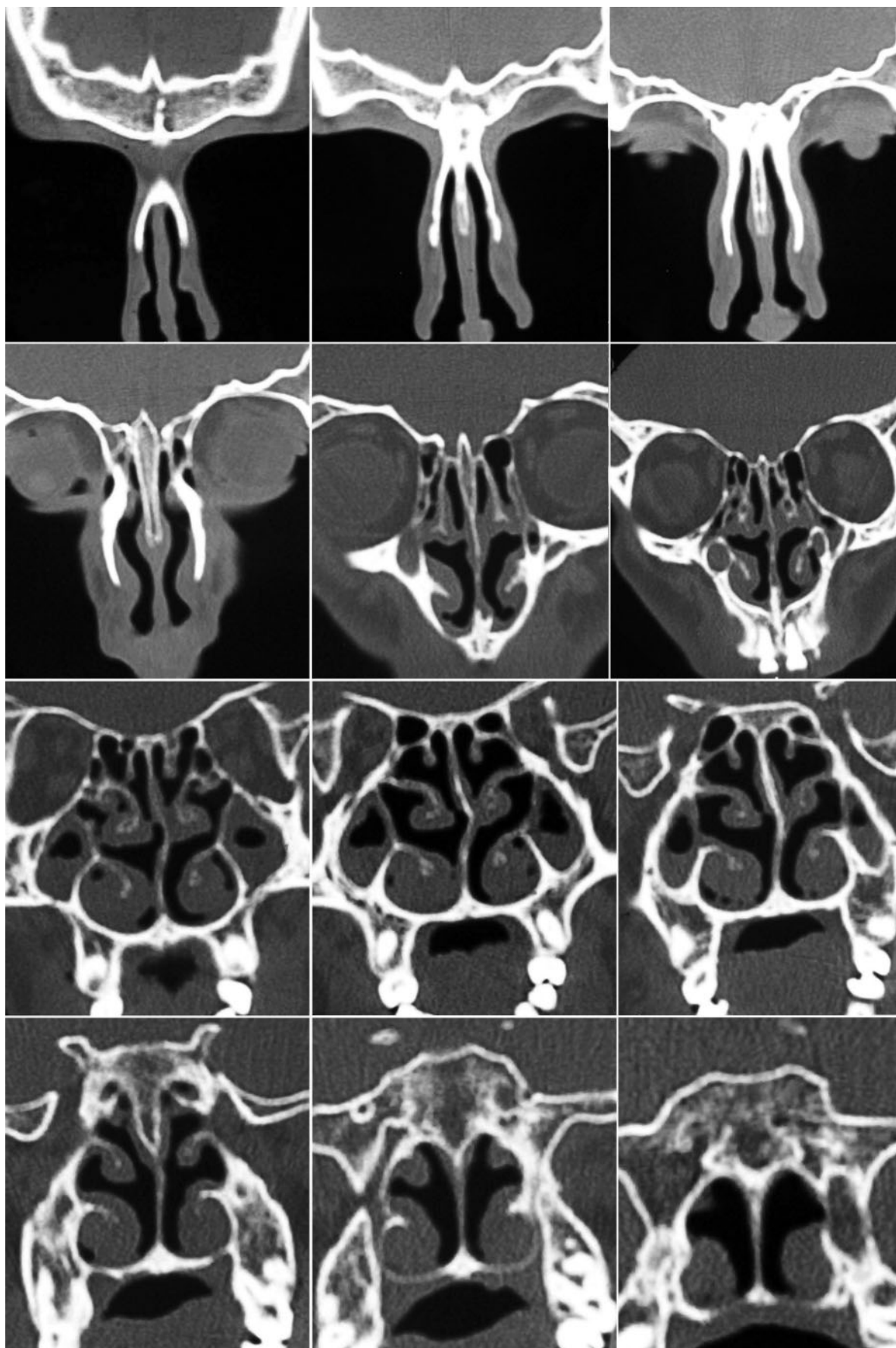


Figure 2. Sequential CT slices of paranasal sinuses in coronal plane shows the same findings in Figure 1: bilateral hypoplastic maxillary and ethmoid sinuses and bilateral frontal and sphenoid sinus aplasias. A slight deviation of nasal septum towards the left side and bilateral soft tissue densities in maxillary sinuses are also seen.

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