# The significance of magnetic resonance imaging and computed tomography findings in sphenoid sinus agenesis

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## Abstract

*Agenesis of the sphenoid sinuses is a very rarely encountered anatomic variation. Findings on magnetic resonance imaging can mislead the radiologist and clinician. Therefore, the gold standard for diagnosis is computed tomography (CT) of the paranasal sinuses. We present the case of a 28-year-old man who was admitted to our hospital with a 3-month history of headache. CT of the paranasal sinuses revealed isolated bilateral sphenoid sinus agenesis.*


## Introduction

The development of the paranasal sinuses begins as evaginations of the nasal mucosa during the third and fourth fetal months. The sphenoid sinuses are undeveloped and nonaerated at birth. Thereafter, they undergo major expansion along with the development of the facial cranium and teeth. The aeration process begins at approximately 3 years of age in the anterior portion of the sphenoid bone in the area known as the *presphenoid,* and it progresses posteriorly into the basisphenoid area until it reaches the anterior wall of the clivus. The mature size of these sinuses is attained at approximately 12 to 14 years of age.[1,2]

Agenesis of the sphenoid sinuses is extremely rare. In this article, we describe a case of bilateral sphenoid sinus agenesis, and we discuss the features of previously reported cases that were diagnosed by computed tomography (CT).

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## Case report

A 28-year-old man was admitted to our hospital with a chief complaint of headache for the previous 3 months. He was concerned that his headaches were being caused by sinusitis or a tumor. The headaches were generalized in character, they occurred at all times of the day, and their duration varied.

Findings on the ENT and neurologic examinations were normal. Magnetic resonance imaging (MRI) of the brain detected no pathology. However, T1-weighted MRI revealed that sphenoid sinus aeration was absent; also, an area of high signal intensity was seen at the normal sites of the sphenoid sinuses (figure 1, A). In order to clarify the meaning of the high signal intensity, fat-saturated MRIs were obtained (figure 1, B). They showed what looked like fatty masses in the usual location of the sphenoid sinuses. To clearly define the bone structure of the sphenoid area, paranasal sinus CT was obtained, and these images indicated sphenoid sinus agenesis (figure 2). CT showed the presence of what appeared to be spongious bone where one would expect to find the sphenoid sinuses. The fatty mass seen on fat-saturated MRI actually represented yellow marrow in the corpus sphenoidale instead of air.

Further investigation revealed no craniofacial anomaly or skeletal disease and no family history of either. The patient was informed of his anatomic variation. For his headaches, treatment with analgesics was started.

## Discussion

Agenesis of the paranasal sinuses is an uncommon anatomic variation; it mainly appears in the frontal sinuses

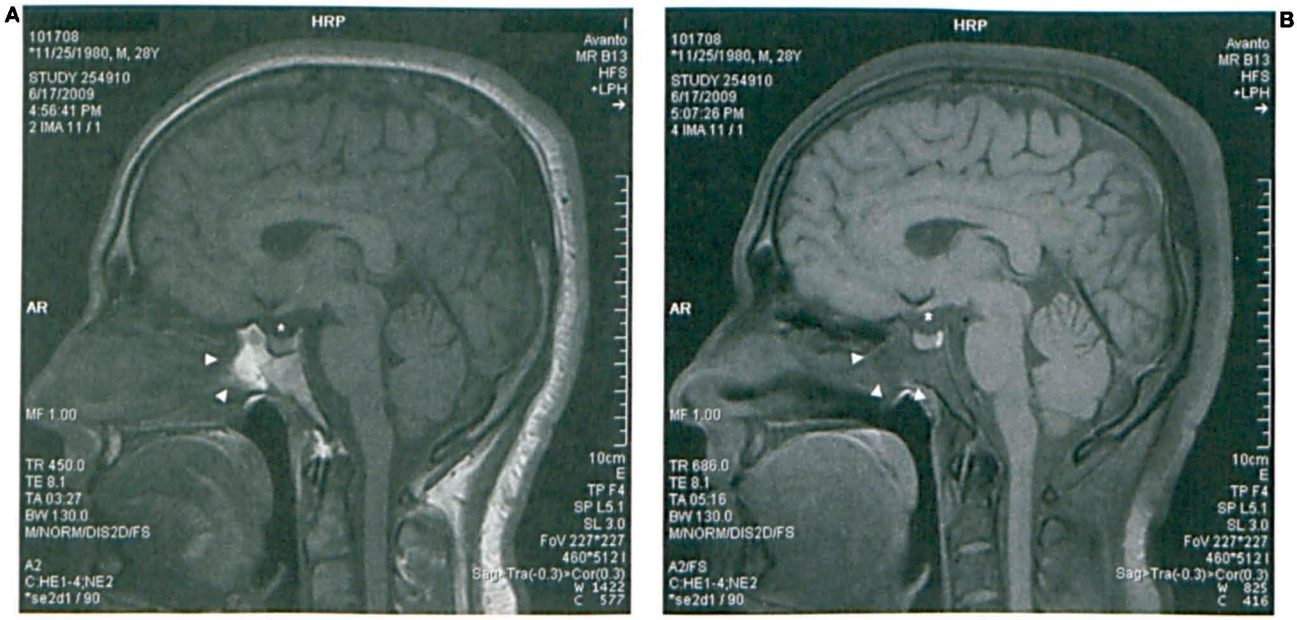


*Figure 1.* ***A:*** *The area of high signal intensity (arrowheads) is seen on T1-weighted MRI.* ***B:*** *Fat-saturated MRI shows the fatty-appearing area (arrowheads) that indicates the presence of yellow marrow instead of air in the corpus sphenoidale. The star symbol represents the pituitary gland.*

(12% of individuals) and next most often in the maxillary sinuses (5 to 6%).[1] These variations are seen more frequently in patients with syndromes of craniosynostosis, osteodysplasia, and Down syndrome (hypoplasia of the frontal sinus).[1,3] Hypoplasia of the sphenoid sinuses in patients with cystic fibrosis has been previously shown.[4]

Sphenoid sinuses are classified as three types based on the degree of pneumatization:

- type I: conchal-type or fetal-type sinuses;
- type II: presellar or juvenile sinuses; and
- type III: adult or postsphenoid sinuses.[1]

Pneumatization may extend into the greater and lesser wings of the sphenoid or pterygoid process and the anterior clinoid process; it can also extend into the occipital bone.[1,5]

Conchal-type pneumatization of the sphenoid sinuses may be misinterpreted as agenesis if CT is not used in the diagnosis.[3] Early anatomic studies indicated that the incidence of sphenoid sinus agenesis was 1 to 1.5 %,[6,7] but these studies had been conducted prior to the availability of CT. CT is now the gold standard for diagnosing sphenoid sinus agenesis.[8] Even so, there have been only 13 previously reported cases of bilateral sphenoid sinus

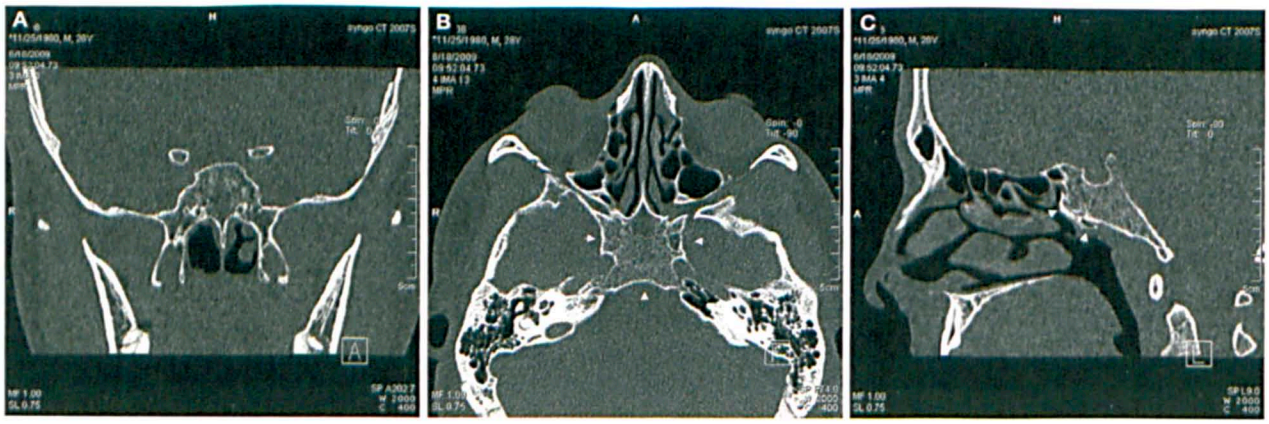


*Figure 2. Agenesis of the sphenoid sinuses is evident on coronal (A), axial (B), and sagittal (C) CTs. The other paranasal sinuses demonstrate normal aeration. Arrowheads indicate the expected location of normal sphenoid sinuses.*

**Table. Summary of reported cases of bilateral sphenoid sinus agenesis diagnosed by CT**

| Author | Sex/age | Agenesis of other sinuses | Associated disease |
|---|---|---|---|
| Antoniades et al,[1] 1996 | 32/M | No | Hand-Schüller-Christian disease |
| Keskin et al,[9] 2002 | 50/F | Posterior ethmoid sinus | Cushing disease secondary to pituitary adenoma |
| Degirmenci et al,[3] 2005 | 45/F | No | None |
| | 17/M | No | None |
| | 44/F | No | None |
| Anik et al,[10] 2005 | 22/F | No | Simple anisocoria |
| Haktanir et al,[11] 2005 | 25/M | Frontal sinus agenesis, maxillary and ethmoid sinus hypoplasia | None |
| Orhan et al,[12] 2010 | 50/M | No | None |
| Güven et al,[13] 2010 | 47/F | Frontal and maxillary sinus agenesis, ethmoid sinus hypoplasia | None |
| Sonbay et al,[14] 2010 | 3 cases* | See footnote* | None |
| Kandogan et al,[15] 2013 | 23/M | Frontal and maxillary sinus agenesis, ethmoid sinus hypoplasia | None |
| Uzun et al,† 2014 | 28/M | No | None |

** Sonbay et al reported 8 cases of sphenoid sinus agenesis—3 bilateral and 5 unilateral—but they did not specify sex and age other than to note that the 8 patients ranged in age from 13 to 66 years. One of the 3 patients also had bilateral frontal sinus agenesis. None had any associated disease.*

*† Present case.*

agenesis that were diagnosed by CT (table).[1,3,9-15] (There have been 8 reported cases of *unilateral* agenesis of the sphenoid sinuses.[14,16,17])

As the sphenoid bones prepare to pneumatize, red marrow changes to fatty yellow marrow.[2] On T1-weighted MRI, this change is reflected as a change from hypointensity to hyperintensity.[18] The focal area of high-intensity signal on T1 weighting can be misinterpreted as a hemorrhage, proteinaceous fluid, or a dermoid cyst.[2,5] In our patient, findings on the initial T1-weighted MRI resembled a fatty mass. Fat-saturated MRI supported this suspicion. It was not until CT was performed that the correct diagnosis was made, as possibly spongious bone was seen in the corpus sphenoidale instead of aeration. In our opinion, the area of high signal intensity was the result of the presence of yellow marrow in the sphenoid corpus. In the previous reports of sphenoid sinus agenesis, the fact that MRI features may be misleading was not emphasized.

We believe sphenoid sinus agenesis is of clinical significance for two reasons:

- On MRI alone, sphenoid sinus agenesis might be misdiagnosed as another condition.
- Aeration of the sphenoid sinuses may be of clinical importance for patients undergoing pituitary gland surgery. As a result, transnasal hypophysectomy may be difficult in patients with sphenoid sinus agenesis, and the risk of complications may be high.

Therefore, we recommend that if a patient's condition suggests the possibility of involvement of sphenoid sinus

pneumatization, CT should be obtained, even though sphenoid sinus agenesis is very rare.

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The treatment options for TOM include antituberculous chemotherapy and surgery. The current indications for surgical intervention are to obtain histologic material for diagnosis, drainage of a subperiosteal abscess, removal of sequestra and, according to some authors, decompression of the facial nerve.[8] However, the role of surgery in the treatment of TOM has obviously declined markedly since the introduction of antituberculous chemotherapy, the results of which are excellent.[3]

The two major problem areas in our case were the tuberculous meningitis and intracranial tuberculomas. The chemotherapeutic regimen for the treatment of tuberculous meningitis is divided into two phases: an intensive initial phase and a continuation phase.[9] During the intensive phase, which usually spans 2 months, the regimen includes a combination of four first-line drugs: rifampicin, isoniazid, pyrazinamide, and streptomycin. The continuation phase, which lasts for 7 to 10 months, includes only rifampicin and isoniazid.[9] Corticosteroids are strongly recommended for the treatment of tuberculous meningitis.[10] They significantly reduce mortality and neurologic deficits.

From a surgical perspective, cerebrospinal fluid diversion procedures are often used in patients who have tuberculous meningitis with hydrocephalus. Despite these efforts, the prognosis for patients with tuberculous meningitis is very poor, as antituberculous treatment prevents death or disability in less than 50% of patients.[9] Early diagnosis and treatment are important for a better prognosis, and the routine use of corticosteroids is strongly recommended.

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