



## Alternating orbital symptoms in cavernous sinus syndrome due to isolated sphenoid sinusitis

### İzole sfenoid sinüzite bağlı kavernöz sinüs sendromundaki değişken orbital semptomlar

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A 40-year-old male with left-sided headaches, ptosis, proptosis and extra-ocular muscle paralysis developed right-sided headaches, proptosis, chemosis, diplopia, extra-ocular muscle paralysis and trigeminal sensory loss. An enhancing left orbital, intrasellar and parasellar cavernous nodule on magnetic resonance imaging progressed into a right cavernous sinus and orbital apex soft tissue mass. Cavernous sinus syndrome from sphenoid sinusitis resolved after endoscopic transnasal sphenoidotomy. This diagnostic complexity and its treatment options are discussed according to current literature.

**Key Words:** Cavernous sinus syndrome; diagnostic complexity; endoscopic transnasal sphenoidotomy; isolated sphenoid sinusitis.

Kırk yaşında erkek hasta, uzun süredir var olan sol tarafta baş ağrısı, ptozis, propitozis ve ekstraoküler kas paralizisi ve sonradan gelişen sağ tarafta baş ağrısı, propitozis, kemozis, diplopi, ekstraoküler kas paralizisi ve trigeminal duyu kaybı nedeniyle başvurdu. Manyetik rezonans görüntüleme sol orbital, intrasellar ve parasellar alana doğru genişleyen kavernöz nodül ve sağ kavernöz sinüs ve orbital apekse doğru ilerleyen yumuşak doku kitlesi saptandı. Sfenoid sinüzite bağlı kavernöz sinüs sendromu endoskopik transnazal sfenoidotomi sonrası iyileşti. Bu tanısal karmaşa ve tedavi seçenekleri güncel literatür eşliğinde sunuldu.

**Anahtar Sözcükler:** Kavernöz sinüs sendromu; tanısal karmaşa; endoskopik transnazal sfenoidotomi; izole sfenoid sinüzit.

Cavernous sinus syndrome (CSS) is a rare and potentially life-threatening condition caused by various conditions, including neoplasms, vascular pathologies, bacterial or fungal infections and noninfectious inflammations. Because the cavernous sinus includes a venous plexus, the carotid artery, cranial nerves and sympathetic fibers, cavernous sinus syndrome is characterized by multiple cranial neuropathies. The clinical presentation includes impairment of ocular motor nerves, Horner's syndrome, and sensory loss of the first or

second divisions of the trigeminal nerve in various combinations.<sup>[1]</sup>

We report a patient with cavernous sinus syndrome presenting with alternating orbital symptoms, diagnosed to have isolated sphenoid sinusitis.

#### CASE REPORT

A 40-year-old male consulted for severe headache and proptosis of the right eye.

Six months prior, he began having intense, squeezing, left fronto-temporal headaches, temporarily relieved by analgesics. Five months before admission, these were accompanied by undocumented fevers, then left ptosis, proptosis and gaze diplopia. An ophthalmologist noted left extraocular muscle palsy and an magnetic resonance imaging (MRI) revealed a left orbital, intrasellar and parasellar small enhancing cavernous nodule. No intrinsic abnormality on the orbital globe, optic nerves, extraocular muscles, optic chiasm and the rest of the brain parenchyma was noted (Figure 1). Some relief was afforded by steroids prescribed by a neurosurgeon, and the patient continued medications without follow-up.

He was asymptomatic until three months prior when intermittent intense squeezing right fronto-temporal headaches radiating to the right face, unrelieved by analgesics and interfering with sleep, prompted consult and admission in another hospital. Right ptosis, proptosis and extraocular muscle paralysis were noted by a second ophthalmologist. Visual acuity then was 6/12+1 OD and 6/15+1 OS. Intraocular pressure was elevated in both eyes

(OD=45 mmHg and OS=60 mmHg). He improved with several ophthalmic ointments, diuretics and intravenous antibiotics. Magnetic resonance imaging revealed interval development of a soft tissue mass in the right cavernous sinus and orbital apex with a similar contrast-enhancing soft tissue mass in the left cavernous sinus compared with the previous scan (Figure 2).

One month prior to admission, the senior author noted the persistent intense headaches, right proptosis, chemosis and ophthalmoplegia with Horner's syndrome and decreased sensation over V 1-3 on the right. Suspecting a sinus etiology for the cavernous sinus syndrome, paranasal sinus computed tomography (CT) scans were requested, revealing isolated right sphenoid sinusitis with cavernous sinus involvement (Figure 3). The patient was started on flucloxacillin and methylprednisolone and admitted for endoscopic direct transnasal sphenoidotomy and biopsy. Mucopurulent and serosanguinous discharge were drained, and granulation tissue and debris cleaned from the sinus. Histopathologic findings were consistent with thrombosis (Figure 4).

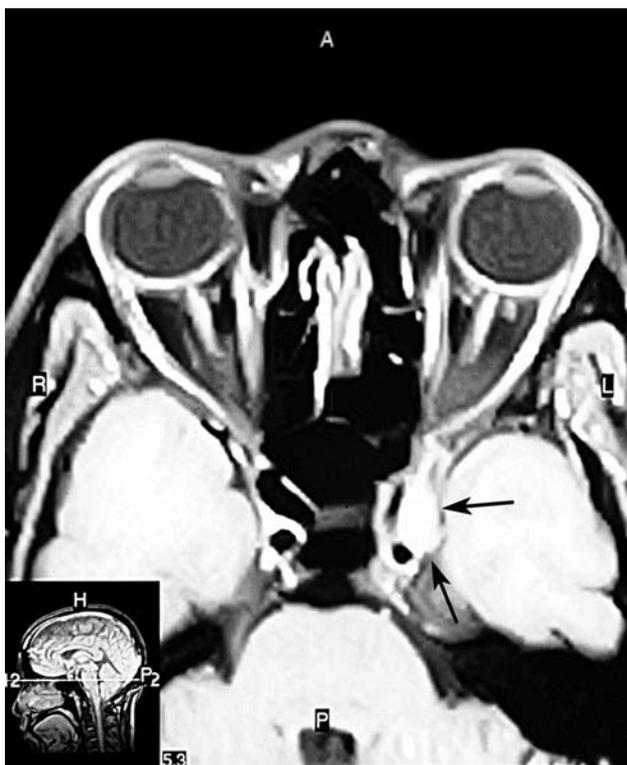


Figure 1. Magnetic resonance imaging with a well-defined, intensely-enhancing ovoid small nodule in the left orbital apex extending into the left parasellar cavernous region (black arrows).



Figure 2. Magnetic resonance imaging showing development of soft tissue mass in the right cavernous sinus and orbital apex and enhancing soft tissue lesion in the left cavernous sinus extending into the superior orbital fissure (black arrows).

The nasal discharge cleared on postoperative follow-up, proptosis and chemosis resolved, followed by resolution of diplopia and headaches as he completed a 28 day course of co-amoxiclav, ranitidine and saline douches. He was clinically well after three, and at six months.

## DISCUSSION

Described by Winslow in 1732, "the cavernous or lateral sinuses of the Os Sphenoides are reservoirs of a very particular kind, containing not only blood, but considerable vessels and nerves; and likewise a spongy or cavernous substance full of blood, much like the spleen or corpus cavernosum of the urethra".<sup>[2]</sup>

The cavernous sinus syndrome clinically presents with paralysis of the oculomotor nerves together with the ophthalmic branch of the trigeminus, and is caused by disease in the area of the cavernous sinus in the anterior middle fossa. The main causes of CSS are tumors (30%) and trauma (24%); self-limited inflammation (23%); carotid aneurysms and fistulas, infection, and other causes (12%). Tumors were the most frequent cause of CSS and of multiple cranial nerve palsies in general. Pituitary adenoma (29%) predominate followed by meningioma (25%), nasopharyngeal cancer (22%), lymphoma (18%) and metastasis (18%) while the aneurysms of the internal carotid artery (16%) and carotid-cavernous fistulas (9%) were the most frequent vascular abnormalities.<sup>[3-5]</sup> Miscellaneous cavernous sinus lesions include Tolosa-Hunt syndrome, herpes zoster and sarcoidosis.<sup>[5]</sup> The presentation is later with aneurysms (average age, 52

years) and tumors (average age, 47 years) than with self-limited inflammation (average age, 35 years) and trauma (average age, 29 years).<sup>[4]</sup>

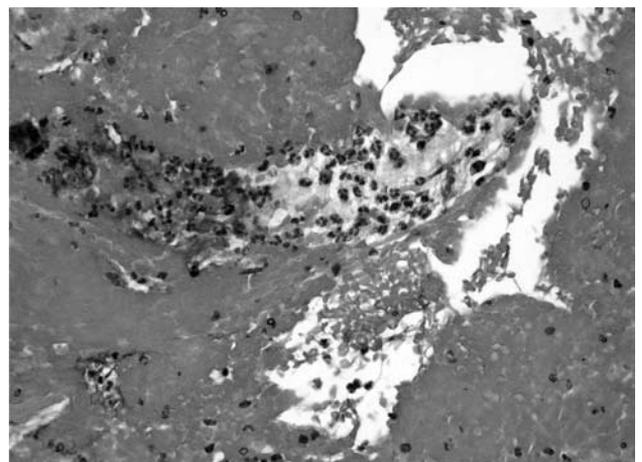
Isolated sphenoid sinusitis accounts for only about 1-2% of all sinus infections. Predisposing factors include entry of infected water into the sinus during swimming, cocaine abuse, steroid ingestion, radiotherapy, fractures of the orbital floor and ethmoid bones, diabetes mellitus and obstruction of the sphenoid ostium by polyp or tumor.<sup>[6]</sup>

Headache is the most common initial symptom in isolated sphenoid sinusitis. The pain ranges from "dull and constant" to "intense and sharp," interfering with sleep and not relieved by pain medications as in the case of our patient. The two most common patterns, also seen in our patient, are unilateral headaches involving the frontal, temporal and occipital region as well as pain or paresthesia of the nervus ophthalmicus to nervus mandibularis areas. Nasal symptoms may be scarce.<sup>[6]</sup>

Computed tomography scan and MRI are currently the most sensitive tools for diagnosis, but may not be specific enough to provide an etiologic diagnosis. Biopsy is technically difficult, and is usually a last resort in patients with cavernous lesions in whom the etiology cannot be determined by less invasive techniques. Isolating the organism responsible is hindered by the difficulties of obtaining bacteriological samples. Many different organisms have been incriminated including *streptococci*, *staphylococci*, *haemophilus* and anaerobic organisms.<sup>[6]</sup>



**Figure 3.** Paranasal sinus computed tomography scan revealed isolated right sphenoid sinusitis (black arrow).



**Figure 4.** Histologic section of tissue from the sphenoid sinus showing blood clots and fibrin enmeshed with leukocytes. Scanty fragments of endothelium-lined vascular channels are noted at the periphery (H-E x 40).

The complications of sphenoid sinusitis are accounted for by the proximity of the sinus to the cavernous sinus laterally, and the superior orbital fissure and optic nerve antero-laterally. The most catastrophic and feared complication of sinus infection and orbital involvement is cavernous sinus thrombosis. Because the dural sinuses and veins draining into them have no valves, the direction of blood flow and spread of infection is subject to pressure gradients.<sup>[7]</sup> The freely-anastomosing valveless venous system of the paranasal sinuses allows retrograde spread of infection to the cavernous sinus via superior and inferior ophthalmic veins. There may be bilateral orbital involvement or rapid progression of inflammation from one side to the other with severe headache, symptoms of meningitis, eye pain and rapidly diminishing vision, and high, swinging temperature patterns. Bilateral proptosis, total ophthalmoplegia and blindness compromise the usual scenario.

Prompt intravenous antibiotic therapy is recommended, and a variety of combinations involving gentamicin, chloramphenicol, nafcillin, penicillin and metronidazole have been used.<sup>[8]</sup> Surgery is indicated if symptoms worsen or continue for 24 to 48 hours on antibiotic therapy, or if there are signs of complications.<sup>[9]</sup> Urgent surgical exploration and drainage is indicated for isolated sphenoid sinus disease accompanied by neurological or ocular symptoms or when a major complication arises.<sup>[10]</sup>

The endoscopic surgical approaches include the transthemoid, transseptal and transnasal, each of which may be combined with an extracranial approach. A transthemoid approach provides better exposure in a narrow nose or when other sinuses are involved, and facilitates postoperative ventilation and debridement. A transseptal approach provides wider exposure of the entire anterior face of the sphenoid and allows the use of binocular vision. Transnasal endoscopic sphenoidotomy has become increasingly popular in recent years.<sup>[11,12]</sup> Using the superior turbinate as a key landmark, it is most useful in patients with spacious noses; otherwise, partial middle turbinate resection may be required.

The diagnosis of CSS remains difficult despite available imaging techniques. Sphenoid sinusitis is an uncommon etiology and easily missed on initial evaluation. The sinus is not accessible on clinical examination and often not clearly seen on routine radiographs. Surgery should be considered in the presence of complications or when the diagnosis cannot otherwise be confirmed. In conjunction with intravenous antibiotic therapy, endoscopic transnasal sphenoidotomy is a viable surgical option.

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