

Neurosciences Quiz

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A 45-year old man with headache, double vision, and facial numbness

Clinical Presentation

A 45-year-old man presented to the emergency room with a 6-month history of headache and a one-week history of right-sided facial numbness. Three days before presentation, he developed double vision, which was worse on looking to his right. He was a smoker, but had no significant history. Examination revealed normal pupils, but there was a limitation of right eye abduction, and a right facial hypoesthesia affecting the forehead and cheek. Motor and sensory functions were normal in his limbs, as was the systemic examination. Laboratory investigations were all normal. Brain MRI performed before, and after IV gadolinium is shown in **Figure 1**.

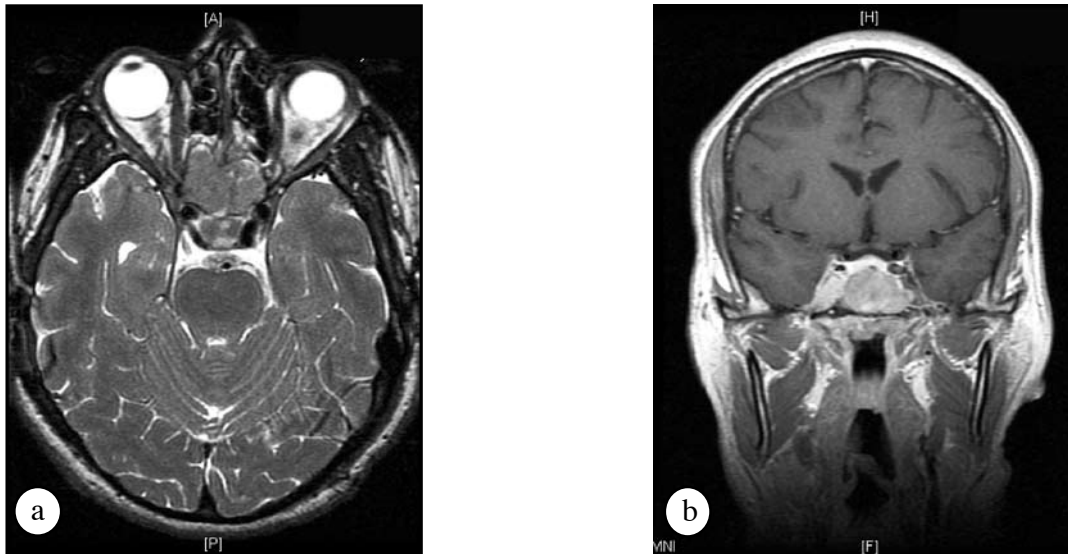


Figure 1 - Brain MRI scans in a) axial T2-sequence, and b) coronal T1-sequence following IV gadolinium of a 45-year-old man with headache, facial numbness, and double vision.

Questions:

1. What is the clinical diagnosis?
2. List the 3 most likely pathological diagnoses.

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Answers

1. Right cavernous sinus syndrome causing sixth and fifth nerve palsies.
2. Mucosal carcinoma, salivary gland tumor, and ectopic pituitary adenoma.

Discussion

The brain MRI shows a contrast-enhancing soft-tissue mass filling the sphenoid sinuses and extending into the right Meckel's cave and cavernous sinus, findings that are consistent with the differential diagnoses described above. The patient underwent a surgical biopsy, which revealed an infiltrating and poorly-differentiated sino-nasal carcinoma of the squamous type grade 3 (**Figure 2**). Chest and abdominal CT scans, and radioisotope bone scan showed no evidence of metastasis. He was then started on chemotherapy and radiotherapy. The cavernous sinus was so named in 1732, however, it was William Gowers in 1888 who observed that “*all the nerves to one eye may be compressed in the wall of the cavernous sinus by an aneurism of the internal carotid or by a growth in this region...*”¹ The cavernous sinus syndrome manifests as ophthalmoplegia due to involvement of the third, fourth, and sixth cranial nerves, and facial pain or numbness due to involvement of the ophthalmic and maxillary divisions of the trigeminal nerve.² The syndrome may be partial or complete, and is bilateral in up to 4% of cases. Common causes include trauma, nasopharyngeal carcinoma, pituitary adenoma, and meningioma, while carotid aneurysms, infections, lymphoma, and the Tolosa-Hunt syndrome are less common.^{2,3} A potentially life-threatening form of this syndrome is cavernous sinus thrombosis, commonly resulting from orbital cellulitis or rhinocerebral mucormycosis. Emergency antimicrobial and anticoagulant therapy are indicated in cavernous sinus thrombosis, while other forms of the cavernous sinus syndrome are treated according to underlying etiology.

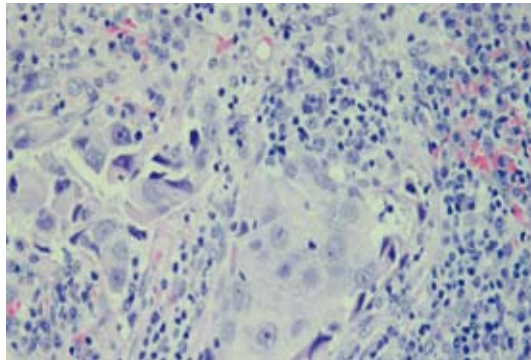


Figure 2 - A histological specimen from the sphenoid sinus of a 45-year-old man with cavernous sinus syndrome showing an infiltrating and poorly-differentiated sino-nasal carcinoma of the squamous type grade 3.

References

1. Gowers WR. A manual of diseases of the nervous system. Philadelphia (PA): P Blakiston Son & Co; 1888. p. 616-617.
2. Keane JR. Cavernous sinus syndrome. Analysis of 151 cases. *Arch Neurol* 1996; 53: 967-971.
3. Goldenberg-Cohen N, Curry C, Miller NR, Tamargo RJ, Murphy KPJ. Long term visual and neurological prognosis in patients with treated and untreated cavernous sinus aneurysms. *J Neurol Neurosurg Psychiatry* 2004; 75: 863-867.