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Case Report

Pituitary Macroadenoma Versus Orbital Apex Syndrome: How well do you know your Neuroanatomy?

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Abstract

Orbital Apex Syndrome (OAS) is a rare cause of cranial nerve deficits. We report a patient presenting with headache known to have a pituitary adenoma who developed unilateral blindness and proptosis and who was diagnosed with orbital apex syndrome and incidental pituitary macroadenoma.

Case Report

A 70-year-old male presented to the emergency department with two weeks of headache and loss of vision in the left eye. He stated that he was recently evaluated by a neurologist who diagnosed him with a "brain tumor" that would require surgery. It was recommended he transfer to our facility for further treatment. In addition to the headache and loss of vision, the patient reported left sided eyelid and facial droop. The patient's medical history included uncontrolled type 2 diabetes and hypertension. His medications included Losartan and a sulfonylurea. His family history was non-contributory and social history was negative for alcohol, illicit drugs and tobacco use.

Vitals signs in the emergency department included a heart rate of 86 beats per minute, the blood pressure was 205/94mmHg, and the temperature of was 36.4°C. The patient was GCS 15, alert and oriented with normal speech. He had decreased visual acuity in the left eye with no light perception. He had ptosis as well as panophthalmoplegia in the left eye with no pupillary response to light on the left side. Mild decreased light touch sensation in the forehead on the left side was also noted. The remainder of the general physical and neurological examination was unremarkable.

Computed tomography (CT) demonstrated a 1.6 X 1.4 X 1.9cm sellar/suprasellar mass interpreted as a possible pituitary adenoma versus aneurysm (Figure 1). Magnetic

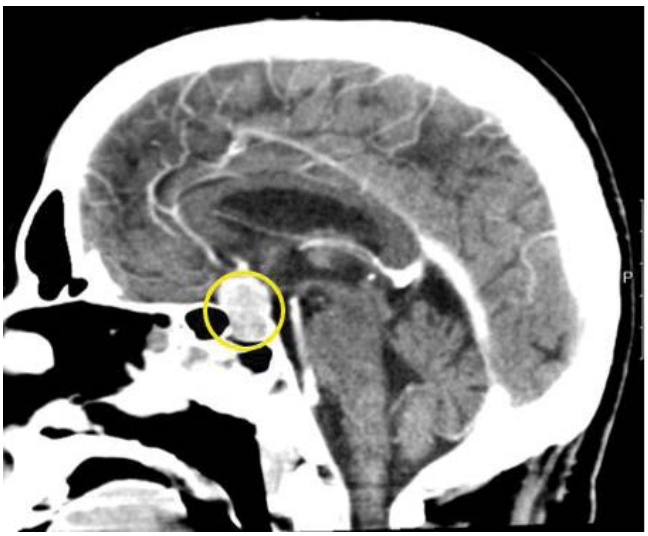

Figure 1: CT Head with contrast: yellow circle depicts a sellar/suprasellar enhancing mass measuring 1.6X1.4X1.9cm consistent with pituitary macroadenoma versus anterior communicating artery aneurysm.

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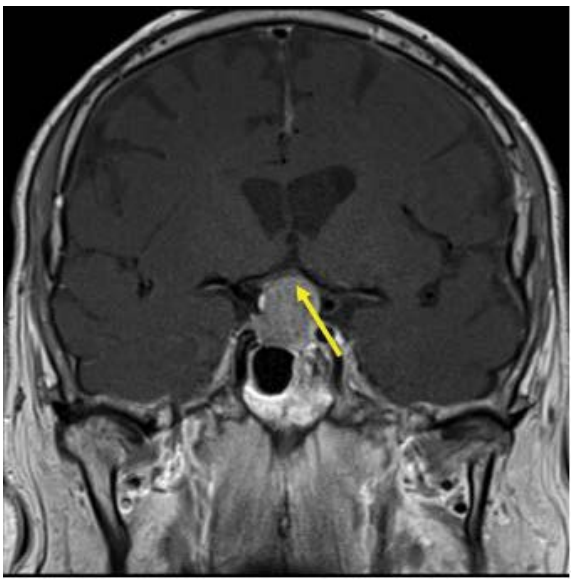

Figure 2: MRI Brain sagittal image: Yellow arrow represents where the 1.5X1.3X2.4 cm sellar/suprasellar mass expanding the sella with suprasellar extension leads to elevation and thinning of the optic chiasm consistent with pituitary adenoma.

resonance angiogram (MRA) of the brain was not consistent with aneurysm. Magnetic resonance imaging (MRI) of the brain confirmed a mass with extension into the optic chiasm consistent with pituitary adenoma. Chronic left sphenoid sinus disease was noted with otherwise normal sinuses (Figure 2). Laboratory work was significant for a white blood cell count of 11.12 X103/UL and a glucose of 257 mg/dL. Basic serum chemistries and blood counts were otherwise normal. Thyroid stimulating hormone was low at 0.30 mlU/L, Free T3 was low at 1.80 pg/mL, and Free T4 ng/dL was normal at 1.25. A morning cortisol was measured at 27 µg/dL. Follicle stimulating hormone, luteinizing hormone, prolactin, adrenocorticotropic hormone, and insulin-like growth factor-1 were all within normal limits.

Initially, the patient was treated supportively while awaiting consultations from endocrinology and neurosurgery. His blood pressure as well as his blood sugars improved with treatment. Because our work-up was consistent with a non-secreting macroadenoma which did not require urgent surgical intervention, it was recommended on hospital day three that the patient follow-up on an outpatient basis with neurosurgery. On that same day, his daily neurological examination revealed a new finding of left eye proptosis in addition to the previous exam findings (Figure 3). While severe headache and blurry vision are very common features of pituitary macroadenoma, proptosis is never a finding associated with this diagnosis. A stat MRI of the orbits was ordered and was consistent with acute and chronic left spheno-ethmoid sinusitis with secondary extension into the left orbit pterygopalantine fossa and left infrazygomatic masticator space (Figure 4). The diagnosis was changed from non-secreting macroadenoma to orbital apex syndrome with incidental pituitary macroadenoma.

Ampicillin-sulbactam and liposomal amphotericin B were immediately initiated for broad spectrum antimicrobial coverage in this patient with uncontrolled diabetes and concern for

Figure 3: Proptosis.

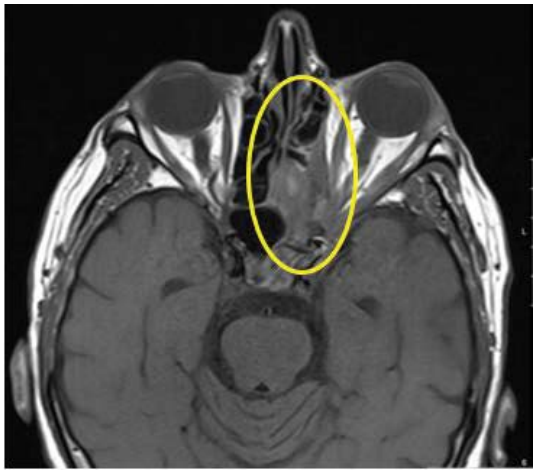

Figure 4: MRI Face and Orbits axial image: Yellow oval demonstrating acute and chronic left sphenoethmoid sinusitis with secondary extension into the left orbit pterygopalantine fossa and left infrazygomatic masticator space.

bacterial and fungal sinusitis. A single dose of dexamethasone was given due to concern for possible mass effect from surrounding edema. The case was promptly managed as a surgical emergency. Unfortunately, there was not an Otolaryngologist available at our facility and the patient was transferred to the nearest facility with these capabilities. The patient underwent orbital apex decompression. Intra-operative cultures subsequently grew Aspergillus. The patient was discharged in stable condition on voriconazole. On follow up, the patient had limited light perception in the left eye. The patient was instructed to follow-up as an outpatient with neurosurgery regarding his non-secreting pituitary macroadenoma.

Discussion

Orbital Apex Syndrome is characterized by impairment of CN II, III, IV, VI and the ophthalmic branch of CNV which results in loss of sensation to the ipsilteral forehead, opthalmoplegia, a fixed and dilated pupil, and ptosis often associated with proptosis [1]. Our patient presented with each of these classic findings.

Cranial Nerve Involvement	Orbital Apex Syndrome	Pituitary Macroadenoma
I	No	No
II	Yes	Yes
III	Yes	Yes
IV	Yes	Yes
V	Yes, *Ophthalmic Branch*	No
VI	Yes	Yes
VII	No	No

Table 1: Cranial Nerve Deficits in Orbital Apex Syndrome vs. Pituitary Macroadenoma.

The classic neurologic presentation for pituitary adenoma on the other hand includes headache and bitemporal hemianopsia [2]. While CN III, IV, V, VI deficits are rarely associated with pituitary macroadenomas, proptosis with monocular blindness are extremely rare in pituitary macroadenoma and should alert the clinician to an alternative diagnosis [3] (Table 1).

Orbital apex syndrome is diagnosed by a thorough history and physical exam followed by a high resolution MRI of the orbit which is the preferred imaging modality to diagnose OAS in the absence of contraindications [4]. Once diagnosed, surgical evaluation is required as this is often a surgical emergency and prompt surgical decompression is indicated to increase the likelihood of the patient regaining vision as well as to obtain cultures for tailoring of antimicrobial therapy. Causes of OAS include infection, neoplasm, trauma, inflammatory disorders such as Churg-Strauss syndrome, and vascular disorders such as carotid-cavernous fistulas, cavernous carotid aneurysms, and sinus thrombosis [1].

Treatment is based on the underlying etiology. Neoplasm or inflammatory conditions are treated based on the underlying disease process itself [1]. In patients with suspected orbital cellulitis as a cause of OAS, broad-spectrum intravenous antibiotics should cover common respiratory pathogens, most commonly streptococci and staphylococci [5]. Coverage for anaerobic organisms should also be strongly considered. Levofloxacin plus clindamycin or ampicillin-sulbactam are excellent choices in these patients. Many institutions are also choosing to provide coverage for methicillin resistant staphylococcus aureus with vancomycin as well [5]. In patients with immune compromising disorders such as in our patient with uncontrolled diabetes, additional coverage with amphotericin B should be added with concern for Aspergillus and Mucor infections [6].

In addition to IV antibiotics, surgical decompression is indicated in all patients with OAS. Once culture results are completed, antibiotics should be tailored. If options for emergency surgery are unavailable, an attempt to lower intraorbital pressure medically using mannitol or acetazolamide systemically or dorzolamide, timolol or brimonidine topically may be attempted [1]. However, these are only to be used in order to attempt to decrease pressure temporarily and are not a substitute for surgical intervention. Use of corticosteroids in infectious OAS has not been extensively studied, but has shown benefit in at least one study [1]. There have been studies for the use of corticosteroids for traumatic OAS that have shown mixed results and their use should be based on a risk benefit analysis on a case by case basis [1]. Cavernous sinus thrombosis is managed with anticoagulation and antibiotics if associated with cellulitis [1].

OAS is a rare syndrome, which can easily be missed especially when patients have co-existing neurological diagnoses complicating their examination. Because these patients are at an extremely high risk for blindness, serial careful and detailed neurological examinations must be done to rule out this syndrome in all patients presenting with cranial nerve deficits. In this case, a change in the patient's physical examination to include the new finding of proptosis prompted an MRI specifically of the orbits. The finding of opthalmoplegia, monocular visual loss and proptosis should alert the examiner to an orbital process rather than a lesion outside of the orbit such as pituitary mass, even prior to the development of proptosis.

References

1. Warburton RE, Brookes CC, Golden BA, Turvey TA. Orbital apex disorders: a case series. *Int J Oral Maxillofac Surg.* 2016;45(4):497-506.
2. Levy A Pituitary disease: presentation, diagnosis, and management. *Journal of Neurology, Neurosurgery & Psychiatry.* 2004;75:iii47-iii52.
3. Chong, K, Lee, S. L. Pituitary macroadenoma with invasion into cavernous sinus, cranial nerve palsies. 2010.
4. Yeh S, Foroozan R. Orbital apex syndrome. *Current Opinion in Ophthalmology*. 2004;5(6):490-498.
5. Cashin L, Cashin B, Joseph M. Orbital Cellulitis Complicated by Subperiosteal Abscess Due to Streptococcus pyogenes. *Infectious Diseases in Clinical Practice*. 2010;18(5):344-345.
6. Molitch ME. Diagnosis and Treatment of Pituitary AdenomasA Review. *JAMA*. 2017;317(5):516-524.