ผู้จัดทำ
วีชยุตม์ สกุลธรรมรักษ์
ศวัส ศรไชยธนะสุข
รุจ นานา
เภทรา วงศ์ภัทรกุล
สริตา ลิ่วคุณูปการ

Case Information

	54 years old Thai Female
Domicile	Bangkok
Occupation	Personal business (Car insurance)
Underlying disease	None
Coverage	Insurance

Chief Complaint Ptosis at left eye 2 months PTA

Present Illness

1 year PTA

The patient had a pulsatile headache at her left side of her head. Pain score 4/10. Each episode lasted for 7-10 days. No nausea/vomiting. No photophobia. No phonophobia. Her symptoms were relieved by pain control medications. Her headache recurred multiple times over the past year.

2 months PTA

The patient noticed a drooping eyelid in her left eye. At first, she could only partially open her left eye (approximately half) then it quickly progressed to complete ptosis. She had decreased vision and double vision at her left eye. Her left eye also became more sensitive to light. She felt numbness on the left upper half of her face. She still had similar episodes of headache, but they had gotten worse than previously- pain score of 7-8/10. She was often awakened by the pain at night. The patient had severe pain in her jaw when chewing food. She also had nausea and vomiting.

Her hearing was normal. She has no significant weight loss, weight gain, fever, or fatigue. She has no palpitations, lactation, deepening of voice, virilization, or nasal discharge.

Past History

Underlying disease	None
Current medication	None

Family history

Malignancy history	None
Surgical history	None
Trauma history	None
Alcohol drinking	None
Smoking	None

Physical Examination

Vital Signs	BP 123/71 mmHg	BT 36.5 C	PR 84/min	RR 20/min
Arthrometrics	55.5 kg	Height 155 cm		
GA	alert, good consciousness			
HEENT	no pale conjunctiva, anicteric sclera, no nasal discharge, no palpable cervical lymph nodes, no enlarged thyroid			
EYE	ophthalmoscope not seen papilledema			
CVS	full and regular pulse, normal S1 S2, no murmur, no JVP engorgement			
RS	clear and equal breath sounds both lungs			
Abd	soft, no distension, no tenderness			
Ext	no skin hyperpigmentation			
CNS				
Cortical Lobe	E4V5M6, oriented to time, place, person			
	no dysarthria			
	follow to two-step commands			
Motor power	grade V all extremities			
	reflex 2+ all extremities			
Sensation	equal pain, temperature sensations all extremities			
	equal pinprick sensation all extremities			
	proprioception normal			
Cranial Nerve	I	no anosmia		
	II	full FOV, RAPD negative,		
	VA	left eye 20/40, corrected 20/25	4 mm slow RTL	
		right eye 20/30, corrected 20/25	3 mm RTL	
	III/IV/VI	complete ptosis of left eye		
		left eye limited EOM all direction, pupil neutral position		
		right eye full EOM, no nystagmus		

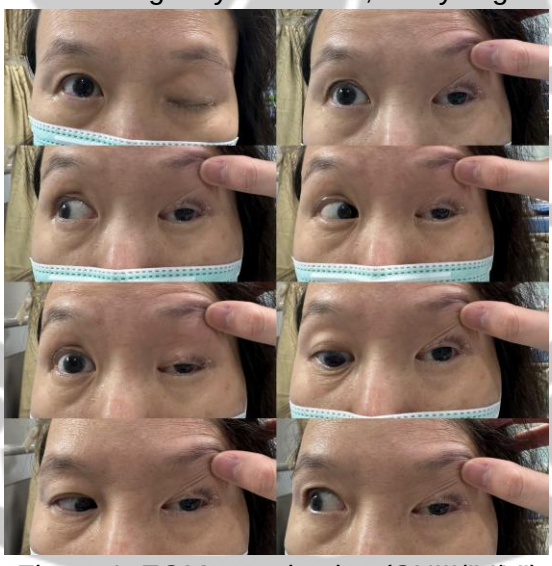


Figure 1: EOM examination (CNIII/IV/VI)

V		decreased pain and temperature sensation at left upper

	face, normal strength of masseter and temporalis muscle.
VII	no facial palsy,

Figure 2: Facial palsy examination (CN VII)

VIII	Rinne AC > BC both ears, Weber no lateralization
IX/X	no uvula deviation, normal gag reflex
XI	normal strength of trapezius and sternocleidomastoid muscle power
XII	no tongue deviation, no tongue fasciculation

Cerebellar Sign: no pronator drift, no truncal ataxia, normal gait, romberg sign negative, no tremor, finger to nose no swaying

Pertinent findings:

1. Left complete ptosis 2 month PTA
2. Limited EOM left eye 2 month PTA
3. Facial numbness 2 month PTA
4. Progressive unilateral headache with awakening pain 1 year PTA

Problem list:
Left complete ptosis with left facial numbness with ophthalmoplegia

Differential Diagnoses:

1. Cavernous Sinus Syndrome
2. Orbital Apex Lesion
3. Subarachnoid Space Lesion

Investigation:
Laboratory investigation

1. Complete blood count to access underlying infection:

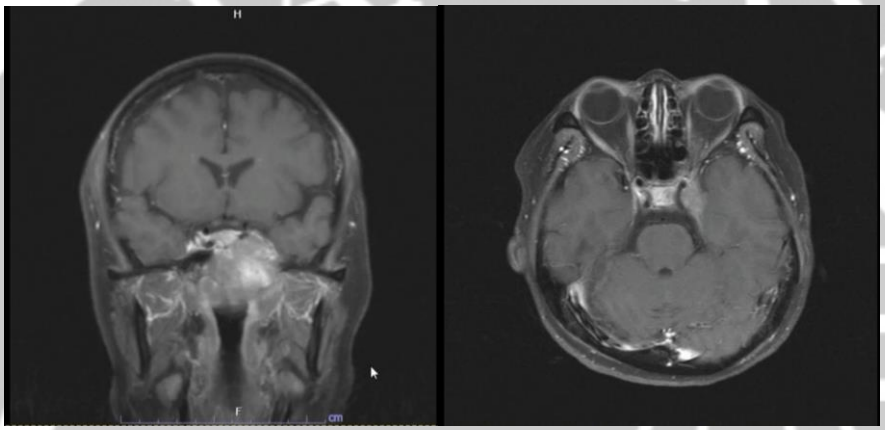
Results: no evidence of underlying infection

2. ESR, CRP, ANCA to evaluate underlying inflammation process:

Results: no evidence of underlying inflammation process

Imaging

1. MRI brain and orbits



Results: asymmetric enhancing lesion in the left cavernous sinus

Diagnosis:

- Cavernous Sinus Syndrome

Management:

- Medical Management: Pain control
- Surgical Management: Tumor removal (Endoscopic Transsphenoidal Approach)

Biopsy Results: involved by carcinoma

Discussion:

The patient presented with left complete ptosis with left facial numbness with ophthalmoplegia. The most possible cause of the patient's condition is neurogenic ptosis, since the patient has unilateral ptosis, ophthalmoplegia, and dilated pupils. The patient also exhibits signs of other cranial nerves abnormalities such as facial numbness and ophthalmoplegia which are the signs of cranial nerve V and IV, VI respectively. With these presenting symptoms, the most likely cause would be an existing lesion in the cavernous sinus affecting all these cranial nerves. Even though an orbital apex lesion could cause similar symptoms, it is less likely due to the absence of optic nerve dysfunction shown in this patient. Lastly, a subarachnoid space lesion could also cause some degree of abnormalities to the eye; however, the patient should also present with other symptoms such as motor weakness and sensory deficits, along with focal symptoms of seizure or alteration of consciousness. Overall, information gathered from history taking and physical examination lead to several possible differential diagnoses. Together with laboratory workups and imaging, diagnosis can be made, which in this case is the lesion at the cavernous sinus.

Knowledge

Cavernous Sinus Syndrome (CSS)

- **Epidemiology**

Most common cause of cavernous sinus syndrome is tumor in around 30% of all cases. Other causes may include trauma, inflammatory disease, vascular and infections.

- **Sign and symptoms**

The signs and symptoms of CSS are characterized by the compression and dysfunction of the structures within the cavernous sinus:

- Ophthalmoplegia

CN III palsy: partial or complete lack of ipsilateral eye elevation, depression, and adduction
CN IV palsy: Ipsilateral eye depression and partial or complete lack of abduction
CN VI palsy: partial or complete lack of ipsilateral eye abduction

- Face-sensing impairment

In the ophthalmic distribution, CN V1 loss causes a partial or complete lack of sensation.
Partial or complete lack of sensation in the maxillary distribution due to CN V2 loss

- Horner syndrome, which results from sympathetic plexus damage and a lack of sympathetic tone
- Proptosis and chemosis, which results from greater pressure in the cavernous sinus

Patients with CSS may not present with all the signs and symptoms listed above, it depends on the cause and the structures affected in the cavernous sinus.

- **Etiology**

Cause	Clinical Features
Tumor	Meningioma, chordoma, neuroma, pituitary adenoma, metastases, lymphoma, nasopharyngeal carcinoma, chondrosarcoma, hemangioma, neuroblastoma
Inflammatory Disease	Tolosa-Hunt syndrome, sarcoidosis
Trauma	Basal skull fracture, operative trauma to cavernous sinus after skull base surgery
Vascular	Intracavernous aneurysm, carotid-cavernous fistula, cavernous sinus thrombosis
Infection	Mucormycosis, aspergillosis, actinomycosis, nocardiosis, mycobacterium, herpes zoster

source: Bhatti. Cavernous Sinus Syndrome. EyeWiki. (2023)
https://eyewiki.aao.org/Cavernous_Sinus_Syndrome

Tumor Characteristics from MRI

Pathology	T1-weighted	T2-weighted	Contrast-enhanced	Other features
Meningioma	Iso	Iso	Homogeneous	Dural tail ICA narrowing when encased
Schwannoma	Iso-hypo	Hyper	Small – Homogeneous Large -Heterogeneous	Dumbbell shape Related to neurofibromatosis
ICA aneurysm	Flow void Iso-hyper if thrombosed	Flow void Iso-hypo if thrombosed	Luminal enhancement	MRA is useful
Metastatic lesion	Hypo	hyper	No specific	Known primary lesion

ICA internal carotid artery

- **Treatment**

The underlying etiology of CSS affects how it should be managed. Treatment is not similar as a result. Tumors are the most common cause of CSS, but various treatments are possible due to the variety of tumor pathophysiology. Potential therapies for the management of a tumor include surgery and/or radiotherapy. Traumatic cases may resolve on their own or may need orbital surgical decompression to treat cases with significant edema and serious damage.

Systemic glucocorticoid therapy is frequently effective for treating inflammatory disease. Vascular etiologies are frequently treatable with interventional radiology procedures like balloon or coil embolization. Infections should be controlled with antibiotics and drainage, if required.

References

1) Kuybu O, Dossani RH. Cavernous Sinus Syndromes. [Updated 2022 Jul 18]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK532976/
2) Fernández, Susana, et al. "Cavernous sinus syndrome: a series of 126 patients." Medicine 86.5 (2007): 278-281.
3) Keane, James R. "Cavernous sinus syndrome: analysis of 151 cases." Archives of Neurology 53.10 (1996): 967-971.
4) Das, Sunit, Bendock BR, et al. “Return of vision after transarterial coiling of a carotid cavernous sinus fistula: case report.” Surgical Neurology. 66 (2006): 82-85.
5) IDSA Practice Guidelines. Infectious Diseases Society of America. https://www.idsociety.org/practice-guideline/practice-guidelines/#/name_na_str/ASC/0/+/. Accessed March 30, 2020.
6) Bhatti. Cavernous Sinus Syndrome. EyeWiki. (2023) https://eyewiki.aao.org/Cavernous_Sinus_Syndrome