Seeing Is Believing: Diffuse Large B-cell Lymphoma Presenting as Horner Syndrome and Syncope

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Introduction

- Horner syndrome (HS) consists of the classic triad of symptoms: **miosis, ptosis, and anhidrosis**.
- The oculosympathetic pathway is made up of a three-neuron system – central, preganglionic, and postganglionic – and HS can be caused by a lesion anywhere along this pathway **(Figure 2)**.
- Diffuse large B-cell lymphoma (DLBCL) commonly presents in the form of a mass due to its aggressive nature and rapid growth.
- Here we present a unusual case of DLBCL with initial manifestation due to Horner syndrome and syncope.

Case Presentation

- A 66-year-old previously healthy woman presented to her PCP with right-sided ear pain and was treated supportively for eustachian tube dysfunction.
- Her symptoms persisted and one month later she presented to the ED following a syncopal event.
- On arrival, her vitals were notable for a mild tachycardia.
- Physical exam was notable for right-sided ptosis and miosis but was otherwise non-focal.
- CT head was normal, but CT neck revealed a right neck mass with narrowing of the nasopharyngeal airway.

Neuron	Pathology
Central	Stroke (hypothalamus, LMS), tumor, spinal cord demyelination
Preganglionic	Apical lung tumors, brachial plexus injury, subclavian artery aneurysm, trauma
Postganglionic	ICA (dissection, aneurysm, trauma, tumor), cluster headache, cavernous sinus lesion

Table 1: Differential diagnosis of Horner sydrome based on order of neuron. ICA = internal carotid artery; LMS = lateral medullary syndrome

Imaging

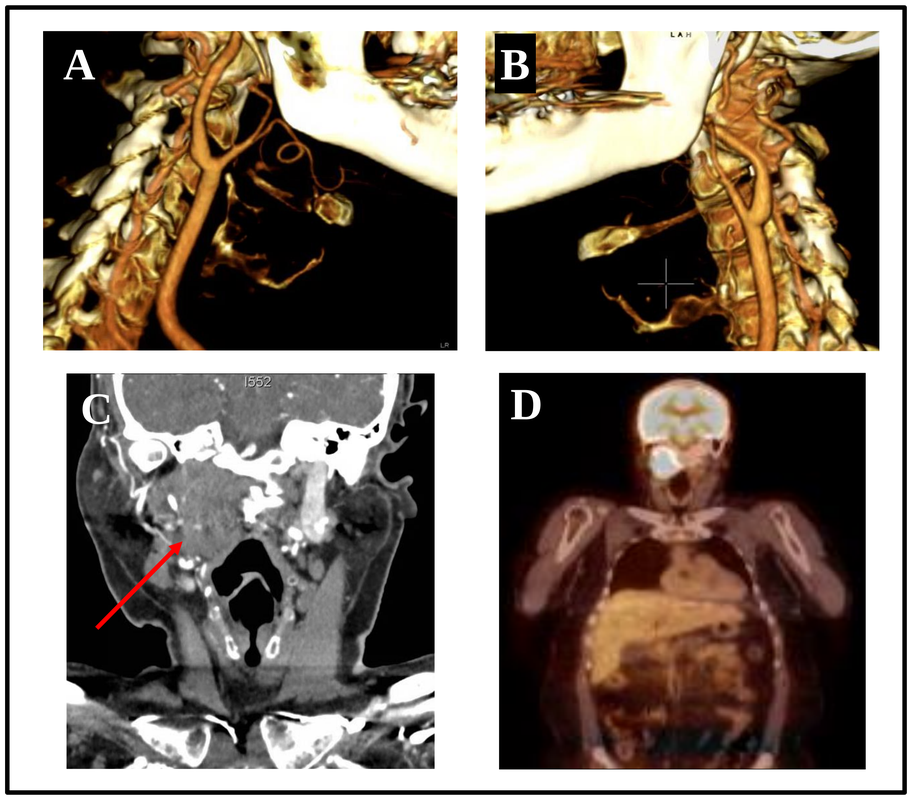

Figure 1: 3D reconstruction of carotid vasculature showing splaying of the right internal and external carotid arteries (A) as compared to the left (B). CT-A neck showing large soft tissue mass involving the right styloid parapharyngeal space (C). PET scan showing intense abnormal uptake corresponding to the right parapharyngeal mass (D).

Clinical Course

- CT-A neck was obtained that demonstrated encasement and displacement of the right cervical internal carotid artery by mass **(Figure 1c)**.
- Fine needle aspiration of the mass was performed and the pathology was suggestive of lymphoma.
- The patient subsequently had an incisional biopsy that revealed the final diagnosis of DLBCL.
- She was initiated on treatment with R-EPOCH (rituximab, etoposide, prednisone, vincristine, cyclophosphamide, and doxorubicin).
- Lumbar puncture was performed and CSF studies were negative for CNS involvement.
- Follow up CT neck two months later showed significant treatment response and reduction in mass size.
- With initiation of chemotherapy, the patient's syncopal episodes resolved.

Discussion

- Causes of HS range from benign to life-threatening, and neck imaging is essential to rule out mass or carotid dissection **(Table 1)**.
- The majority of HS cases are caused by a second- or third-order neuron lesion, as was the case for our patient.
- Her syncope was explained by mass effect on the right carotid sinus; her HS was likely due to disruption at the superior cervical ganglion.
- Carotid artery invasion is commonly seen in squamous cell carcinoma, while only a few case reports exist describing involvement with DLBCL.

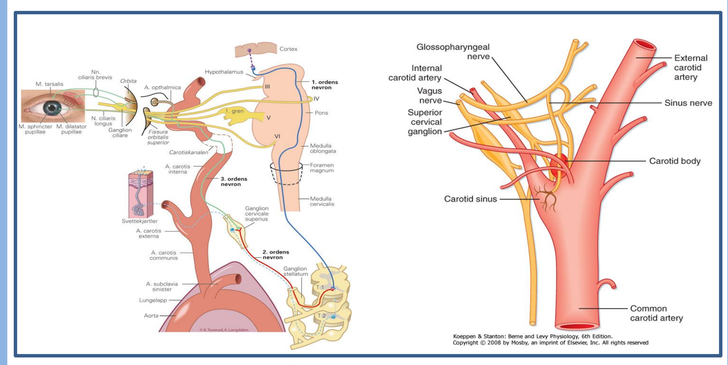

Figure 2: Illustrations of the oculosympathetic pathway and anatomical relationship between the carotid artery and carotid sinus.

Key Points

- It is important to keep malignancy at the top of the differential diagnosis in adults presenting with neck mass.
- HS with neck pain should be treated as carotid artery dissection until proven otherwise.
- Swift recognition of HS is essential to not only diagnose the underlying cause but to prevent neurologic morbidity.

References

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