

A tale of ptosis, pharmacological tests, and Pancoast tumour

Muhammad Fadhli Ab Hamid^{1,2} Mimiwati Zahari^{1,2}, Norlina Mohd Ramli^{1,2}

¹Department of Ophthalmology, Faculty of Medicine, University of Malaya, Kuala Lumpur, Malaysia; ²University Malaya Eye Research, Faculty of Medicine, University Malaya,Kuala Lumpur Malaysia

Abstract

Horner syndrome (HS) classically presents with ipsilateral blepharoptosis, pupillary miosis, and facial anhidrosis and caused by a lesion along the oculosympathetic pathway from the hypothalamus to eye. The diagnosis of HS in a patient presenting with partial ptosis may be easily missed in the Asian patient. This is mainly due to the dark irides, making detection of anisocoria on direct visualization difficult. Index of suspicion must be high, especially in the absence of any extraocular motility or lid abnormalities. We present a case where a healthy asymptomatic patient presented with partial ptosis and diagnosis of Horner syndrome was eventually confirmed through pharmacological tests. Non-targeted imaging with a simple chest x-ray revealed an apical lung lesion which eventually turned out to be malignant. Although it is a typical textbook description, this case highlights the importance of careful history and examination in an otherwise healthy patient presenting with mild ptosis.

Keywords: Horner syndrome, Pancoast tumour

Correspondence: YDr. Muhammad Fadhli Ab Hamid, MBBS (UM), Department of Ophthalmology, Faculty of Medicine, University of Malaya, 50603 Kuala Lumpur, Malaysia. E-mail: messentry7@yahoo.com

Kisah ptosis, ujian farmakologi, dan tumor Pancoast

Abstrak

Sindrom Horner (HS) secara klasik menunjukkan adanya blefaroptosis ipsilateral, miosis pupil, dan anhidrosis muka dan ianya disebabkan oleh lesi di sepanjang jalan okulosimpatetik dari hipotalamus ke mata. Diagnosis HS pada pesakit yang mengalami ptosis separa mungkin mudah dilupakan pada pesakit Asia. Ini terutama disebabkan oleh warna gelap, menjadikan pengesanan anisocoria secara pemerhatian terus sukar di visualisasi. Indeks kecurigaan mestilah tinggi, terutama jika tidak ada pergerakan atau kelainan kelopak mata yang luar biasa. Kami menunjukkan kes di mana pesakit asimtomatik yang sihat dengan ptosis separa dan diagnosis sindrom Horner akhirnya disahkan melalui ujian farmakologi. Pengimejan tanpa sasaran dengan sinar-X dada yang sederhana menunjukkan lesi paru-paru apikal yang akhirnya disahkan malignan. Walaupun ini adalah keterangan buku teks yang biasa, kes ini menyoroti pentingnya sejarah dan pemeriksaan yang teliti pada pesakit yang sihat yang mengalami ptosis ringan.

Kata kunci: Sindrom Horner, tumor Pancoast

Case report

A 51-year-old man presented with a 3-day history of painless ptosis in the right eyelid. There was no associated diplopia or blurring of vision. He denied having a history of neck trauma. Systemic investigations were unremarkable. He admitted to smoking heavily but had quit smoking for the past two years. Examination revealed visual acuity of 6/6 in both eyes. His right eyelid had partial ptosis (vertical palpebral apertures were 5 mm and 7 mm for the right and left eye, respective-ly); both eyes had normal levator function (Fig. 1). There was slight anisocoria: 3 mm in the right pupil and 5 mm in the left pupil under room light. The anisocoria was more prominent in dim illumination, confirming the smaller pupil in the right eye as the pathological one. Cogan twitch sign, fatigability test, and ice pack compression were negative. Examinations of the anterior and posterior segments as well as the cranial nerves were unremarkable. Systemic examination did not reveal any other abnormalities.

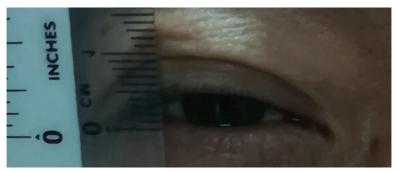


Fig. 1. Partial ptosis of the right eyelid.

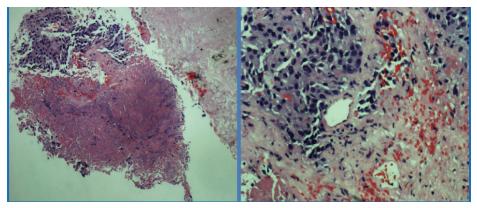


Fig. 2. (*Left*) Low-power view (H&E x 20) showing the clusters of malignant cells with surrounding wide areas of necrosis. (*Right*) High-power view (H&E x 100) showing malignant cells in clusters with pleomorphic hyperchromatic nuclei.

Based on the initial assessment, the provisional diagnosis was Horner syndrome (HS) in the right eye which is a clinical diagnosis of a disruption of oculosympathetic pathway from hypothalamus to the eye classically presents with ipsilateral blepharoptosis, pupillary miosis and facial anhidrosis.¹ This was confirmed a few days later by performing a 10% topical cocaine test where the right pupil failed to dilate. The test was done at a later date as due to difficulties in obtaining the cocaine solution on the same day.

A chest X-ray with apical views was ordered as an initial investigation to rule out an apical lung lesion. It showed lung consolidation at the right upper lobe. He was then referred to the respiratory team for a computerized tomography-guided lung biopsy. Biopsy results showed a poorly differentiated non-small cell carcinoma (Fig. 2). The patient unfortunately developed complications during surgery to remove the tumour and passed away.

Discussion

The presence of recent onset HS with anisocoria and ptosis in an otherwise healthy adult should always alert the clinician to an underlying systemic cause. In this case, the patient presented with ptosis as the main symptom. The role of imaging is thus very important in determining the aetiology. The yield of targeted imaging in cases where the aetiology of HS is unknown through history and examination is usually very low.¹ In a case series of 88 patients with HS, only one patient was found to have an asymptomatic malignancy as the aetiology.² In the current literature, only nasopharyngeal carcinoma and syringomyelia have been reported to present with isolated HS.^{2,3} Even in Pancoast tumours, 44% to 96% of patients have an initial presentation of shoulder pain and not HS.^{4,5} Early lesions of Pancoast tumour are known to present with constantly severe, constant pain in the shoulder radiating to the ulnar nerve distribution (pressure or damage of brachial plexus), ipsilateral weakness and atrophy of the small muscles of the hand (paraesthesia, dysesthesias), rib and vertebral body destruction, and HS (invasion of the sympathetic nerve).^{6,7} In our patient, the apical lung lesion was evident from a simple chest X-ray ordered on the same day. Hence there was no delay of this grave diagnosis.

This case highlights several important learning points in the assessment of a patient with HS. Firstly, the presence of anisocoria may be easily missed, especially in Asian eyes with dark irides. As this is usually not a presenting complaint, the clinician needs to be on the lookout for this clinical sign in patients presenting with ptosis. Confirmatory tests as described in textbooks (10% cocaine) are usually difficult to obtain on the spot in most clinical settings. Simple but potentially lifesaving investigations, such as chest X-rays, are absolutely essential in cases with isolated HS. These can usually be obtained faster and should not be delayed whilst wating for the confirmatory pharmacological tests.

In conclusion, non-targeted imaging in cases of isolated HS is essential and should not be delayed until confirmatory pharmacological tests are done. Although the yield is low, it may allow early diagnosis of potentially fatal lung lesions.

Acknowledgements

Due to the patient's untimely death during tumour removal, the authors were unable to seek consent for publication. However, all details contained in this case report have been sufficiently anonymized to protect the patient's identity.

References

- 1. Almog Y, Gepstein R, Kesler A. Diagnostic value of imaging in Horner syndrome in adults. J Neuroophthalmol. 2010;30(1):7-11.
- 2. Kerrison JB, Biousse V, Newman NJ. Isolated Horner's syndrome and syringomyelia. J Neurol Neurosurg Psychiatry. 2000;69(1):131-132.
- 3. Padungkiatsagul T, Poonyathalang A, Jindahra P, Cheecharoen P, Vanikieti K. Isolated Horner syndrome as a rare initial presentation of nasopharyngeal carcinoma: a case report. Int Med Case Rep J. 2018;11:271-276.
- 4. Van Houtte P, MacLennan I, Poulter C, Rubin P. External radiation in the management of superior sulcus tumor. Cancer. 1984;54(2):223-227.
- 5. Shahian DM, Neptune WB, Ellis FH, Jr. Pancoast tumors: improved survival with preoperative and postoperative radiotherapy. Ann Thorac Surg. 1987;43(1):32-38.
- 6. Foroulis CN, Zarogoulidis P, Darwiche K, et al. Superior sulcus (Pancoast) tumors: current evidence on diagnosis and radical treatment. J Thorac Dis. 2013;5 Suppl 4:S342-358.
- Arcasoy SM, Jett JR. Superior pulmonary sulcus tumors and Pancoast's syndrome. N Engl J Med. 1997;337(19):1370-1376.