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Case Report

ACQUIRED ADULT HORNER'S SYNDROME: CLINICAL PERSPECTIVES

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Background: Horner's syndrome is a rare entity diagnosed clinically. The underlying disease process lays hidden as it may be the only presentation of a mortal disorder. Imaging modalities such as MRI, CT are important to diagnose the causative factors of Horner's syndrome.
Case reports: Two consecutive patients received in a tertiary care hospital with Horner's syndrome resulting from varied aetiology are being presented
Discussion: Clinical perspectives of central, preganglionic and postganglionic Horner's syndrome have been discussed and an attempt has been made in devising a protocol involving combined clinical and safe radiological approach of a patient for early diagnosis.

INTRODUCTION

Horner syndrome results from dysfunction of the ipsilateral oculosympathetic pathway and is also called oculosympathetic paresis (Moon *et al*. 2015). Classic triad of signs of Horner syndrome include ipsilateral mild ptosis involving upper lids due to paralysis of the Müller's muscles innervated by the sympathetic pathway, unilateral miosis, dilation lag in the dark (slow dilation of the affected pupil, with or without anhydrosis of face on affected side. (Kong *et al* 2007). Pseudo-enophthalmos is observed because of the reduced palpebral fissure. The diagnosis of Horner syndrome is most often just based on the findings of anisocoria, with the small pupil not dilating well in the dark and decreased ipsilateral palpebral fissure.

The long anatomical oculosympathetic pathway makes it vulnerable to many pathological processes. (Davagananam *et al*. 2013) Neurologic symptoms and signs such as anhidrosis of ipsilateral face may occur with preganglionic lesions (first or second order). Cranial nerve dysfunction with or without brainstem & spinal cord signs and symptoms suggest a first-order Horner syndrome. Associated arm pain, hand weakness, history of neck surgery, or neck trauma suggest a second-order Horner syndrome. Third-order Horner's syndrome points to lesions of the internal carotid artery such as dissection or cavernous sinus aneurysms presenting as unilateral head and neck pain and incomplete Horner's syndrome (Sivshakthi and Miller 2015).

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The diagnosis can be established by a detailed clinical history and examination and conjunct use of diagnostic imaging. It is not common for patients to present with classical signs and symptoms, but asymptomatic Horner's syndrome pose a great diagnostic challenge.

The present study will discuss two cases with a variable clinical presentation and aetiology received at a tertiary care centre. Identifying its cause is a diagnostic challenge in spite of its apparent presence. The importance of a proper clinical examination is vital. The use of clear combined clinical and recent diagnostic imaging techniques for diagnosis of Horner's syndrome for clinicians is being stressed upon. Familiarity with the pathway, knowledge of related structures and affecting diseases help in early evaluation. Its importance lies as Horner's syndrome may be the only manifestation of a mortal disorder.

Clinical Report

Case 1*:* A 50 year old man was hospitalised due to fever for last ten days and repeated vomiting for the past six days. Fever was of moderate intensity, sudden in onset, intermittent, associated with chills unresponsive to simple analgesics. The patient also complained of dizziness and giddiness since four days. 7-8 episodes of projectile vomiting per day whitish colour which were not blood stained containing food particles ingested were reported by the patient. Patient was non smoker and non alcoholic with no chronic illness in family. There were no remarkable features in the history. No complaints of chest pain, abdominal pain, cough, joint pain, eye or ear discharge, burning micturition or any complaint of diarrhoea was stated. On inspection, patient appeared well oriented, vitals normal. Chest and cardiovascular system appeared

normal. Liver function tests, renal function tests, serum electrolytes and haematological reports were within normal limits. Ultrasound of abdomen reported a simple parenchymal cyst in the upper pole of right kidney measuring 18X4 mm. Diagnosis of Horner's syndrome was made on the basis of clinical examination. Patient had mild ptosis of left eye with miosis and loss of sweat secretion on the left forehead. (Fig 1, 2) The patient was advised MRI brain and cerebral angiography to further investigate the underlying cause. Patient was put on nil per oral and advised intravenous fluids and antibiotics. Sugar charting and input output charts and blood pressure monitoring was done. Vertigo, dizziness and vomiting did not subside much on treatment.

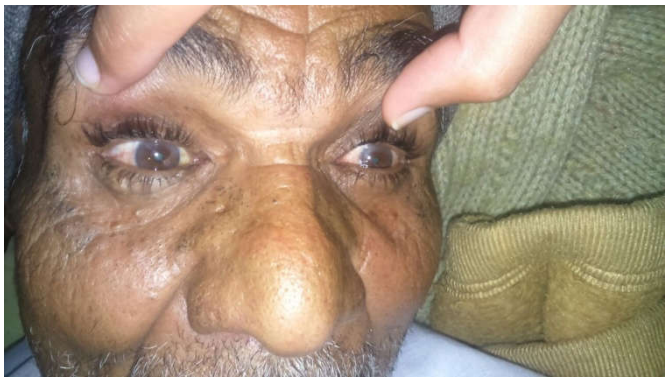

Fig 1 Patient has miosis of left eye

Fig 2 Ptosis of left eye in the patient of Horner's syndrome

MRI brain observed focal altered signal intensity area showing hyperintensity on T2 and FLAIR and hypointensity on T1W sequence with diffusion restriction observed in temporo-parietal occipital lobe, left medulla oblongata and left cerebellar hemisphere suggestive of acute infarct. Elsewhere cerebral parenchyma appeared normal. Morphology of corpus callosum also appeared normal. No midline shift was observed and brainstem as well as cerebellum showed no pathological signs. However signs of diffuse atrophy were indicated by presence of prominent superficial sulci and ventricular systems (Fig 3).

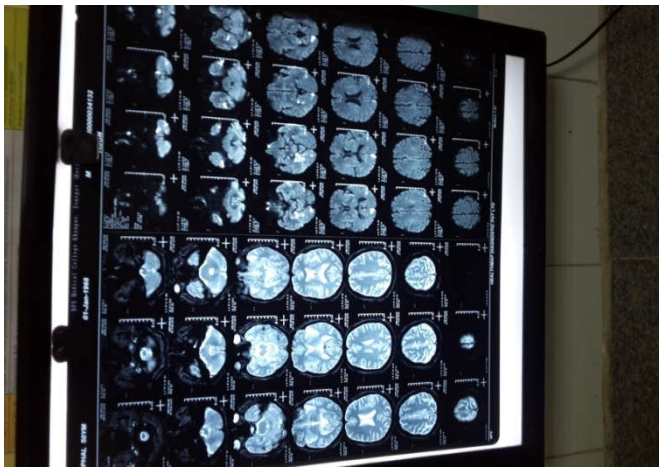

Fig 3 MRI of brain showing hyperintensity on T2 and FLAIR and hypointensity on T1W sequence with diffusion restriction observed in temporo-parietal occipital lobe, left medulla oblongata and left cerebellar hemisphere suggestive of acute infarct.

Vertebral basilar insufficiency was suspected and cerebral angiography was done to confirm the cause. CT angiography was done using MDCT from base of skull to vertex after administration of contrast media. Findings revealed subtle hypodensity in left half of medulla, left cerebellum and right inferior temporal lobe infarcts.

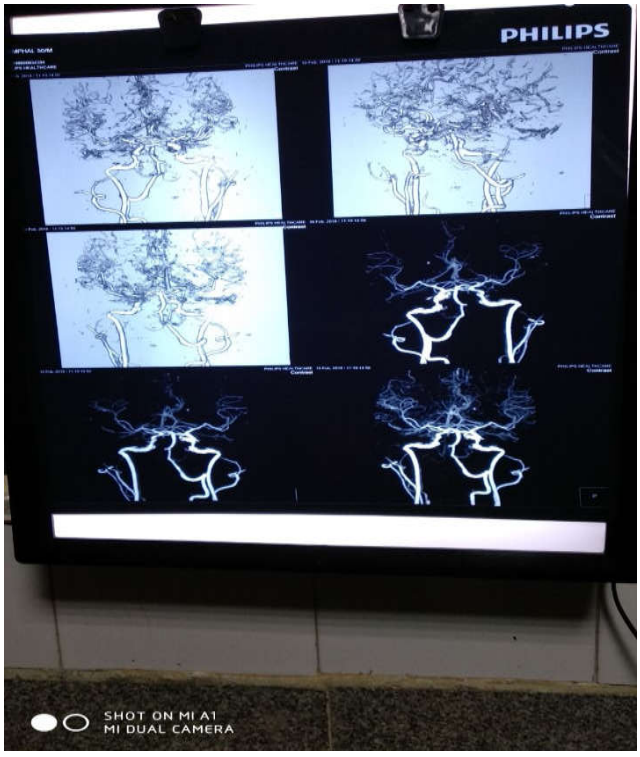

Fig 4 CT angiography showing hypoplastic left vertebral artery with stenotic or occluded V3 &V4 segment were seen on film

Both cerebral hemispheres and thalamo-ganglionic regions appeared normal. Ventricular system, basal cisterns and cortical sulci appears prominent suggestive of mild cerebral atrophy. Angiography calcific plaque were noted in intra cerebral portions of bilateral internal carotid arteries. Left vertebral artery was narrow in lumen as compared to right side and showed faint opacification upto C1 vertebral foramina only after which no contrast opacification was seen. The possibility of hypoplastic left vertebral artery with stenotic or occluded V3 &V4 segment were seen on films (Fig 4). Vertebral artery stenosis as a cause of Horner's syndrome was confirmed and patient referred to higher centre for vertebral artery revascularisation.

Case 2*:* A 35 year male presented in outpatient department with loss of appetite for the last one month. Patient reported loss of weight since 6-7 days. No other major complaint was expressed by the patient. On abdominal examination patient had hepatosplenomegaly. CNS examination revealed ptosis, miosis and anhydrosis. CVS examination revealed bradycardia. Patient was investigated for causes of Horner's syndrome. Apart from hematogram, serum viral markers, renal function tests, liver function tests were conducted. The total leukocyte count of the patient was raised. The viral markers Hepatitis B surface antigen (HBS Ag) and antibodies such as anti HBC were negative. However total protein was decreased. CT chest revealed lymph nodes compressing cervical sympathetic chain. Ultrasound guided lymph node biopsy was done. Histopathological examination revealed Hodgkin's lymphoma. The patient was referred to the oncology department for chemotherapy.

DISCUSSION

The oculosympathetic pathway comprises of three neuronal chains extending from hypothalamus to the eye (Patel and Ilsen 2003). The first order neurons arise from posterolateral hypothalamus descend through the brain stem and terminate at ciliospinal centre of "Budge and Waller" in the intermediolateral column of C8-T1 segments of spinal cord. The preganglionic second order neuronal fibres exit via the ventral roots of T1 spinal nerves and traverse the inferior and middle cervical ganglia or stellate ganglia of paravertebral sympathetic chain and terminate in the superior cervical ganglia. The T1 root travels close to the apex of lung. The vertebral artery lies anterior to the inferior cervical ganglia and the fenestrated posterior cord of middle cervical ganglia surrounds the vertebral artery. The third order neurons (postganglionic) exit from the superior cervical ganglion to form a plexus surrounding the internal carotid artery. The plexus ascends through cavernous sinus along the 6th cranial nerve and finally follows the ophthalmic division of trigeminal nerve to supply Muller's muscle and dilator of iris in orbit. Fibres to sweat glands and blood vessels follow the external carotid artery and supply ipsilateral half of the face. (Kong *et al* 2007).

Horner's syndrome is quiet uncommon. Table 1 illustrates differential diagnosis of Horner's syndrome. All characteristic features of Horner's syndrome; ptosis, miosis and anhydrosis may not appear in every case depending upon the lesion. Evaluation of 450 cases of Horner's syndrome (Maloney *et al*. 1980) claimed that classic traid of symptoms occurred less frequently while miosis was observed in 98% of cases, ptosis occurred in 88% and only 4% had anhydrosis. The distribution of anhydrosis varies from the entire half face to a small patch.

Table 1 Differential diagnosis of Horner's syndrome

First- order neuron lesions (Central)	Second-order neuron lesions (pre-ganglionic)	Third -order neuron lesions (post ganglionic)
Arnold -Chiari syndrome	Injury to lower brachial plexus(Pancoast syndrome)	Internal carotid dissection
Syphilitic meningitis	Cervical rib	Cluster headache
Lateral medullary syndrome	Aneurysm or dissection of aorta, common carotid artery	Carotid artery thrombosis
Neck trauma	Trauma or surgical injury (thyroidectomy, carotid angiography, chest tube insertion	Carotid cavernous fistula
Intrapontine hemorrage	Lymphadenopathy eg. Hodgkins disease, TB, leukemia, mediastinal tumour	Herpes zoster
Syringomyelia	Neuroblastoma and lesions of middle ear	Idiopathic
Demylinating disease		
Pituitary and basal skull tumours		

Ipsilateral Horner's syndrome with contralateral hemiparesis and contra lateral hypesthesia occurs in tumors and hemorrhage of hypothalamus (Austin and Lessell 1991). Contra lateral ataxic hemiparesis, hypoesthesia, dysphasia with vertical gaze palsies reflects involvement of thalamus (Rosetti *et al*. 2003). A combination of Horner's syndrome with contra lateral trochlear nerve paresis is suggestive of lesions of dorsal mesencephlon (Guy *et al*.1989). Pontine lesions have additional unilateral or bilateral abducens nerve palsies (Kellen *et al*.1988).

Sacco *et al*.1993, observed Horner's syndrome in 91%, ipsilateral ataxia in 85% of 33 patients of lateral medullary infarction. Vertigo, dyspahgia, nystagmus and facial weakness were less frequently found. The presenting symptoms of case one of the present study were vertigo and dysphagia. Signs of Horner's syndrome were a part of lateral medullary syndrome in the present study. Wallenberg syndrome is caused by occlusion of either posterior inferior cerebellar artery or vertebral artery. Kim 2003, in his study on 130 patients of lateral medullary infarction found that the pathogenesis was large vessel infarction in 50%, arterial dissection in 15%, small vessel infarction in 13% and cardiac embolism in 5%.

Spinal cord lesions like trauma, inflammatory disease, infectious myelitis, vascular malformation, syringomyelia do present with central Horner's syndrome, loss of touch and motor function on the same side but loss of temperature and pain sensation on the opposite side (Kobayashi *et al*.2003).

Ipsilateral shoulder pain, paresthesias along medial side arm, forearm radiating to 4th and 5th digit accompanied by weakness and or atrophy of hand muscles with preganglionic or second order horner's syndrome is a classical presentation called the Pancoast syndrome. Tumours involving the apex of the lung, plasmacytoma, non Hodgkins lymphoma, lymphomatoid granulomatosis, metastasis lead to Pancoast syndrome. Sympathetic chain schwannomas, mediastinal tumors and cysts also cause preganglionic Horner's syndrome (Sivashakthi and Miller 2015). Signs of lung disease are evident among patients. Horner's syndrome, an unusual presentation of Hodgkin's disease has been described by Simon *et al*. (1985), who described occulosympathetic damage due to mediastinal compression which is in accordance to case 2 of the present

study. Brachial plexus injuries, fracture of first rib, surgical procedures around neck like internal jugular catheterisation, CABG, pacemaker insertion lung or mediastinal surgery have also been attributed to iatrogenic Horners syndrome (Sivashakthi and Miller 2015).

Presenting symptoms and signs are different in post ganglionic Horner's syndrome. Unilateral head & neck pain with cerebral ischemia symptoms, miosis and ptosis (incomplete Horner's syndrome) is the presentation in patients affected by lesions of internal carotid artery either dissection or aneurysm. Lack of anhidrosis in incomplete Horner's syndrome has been explained that fibres supplying sweat glands of face travel along external carotid plexus. Horner's syndrome along with abducens nerve, facial nerve palsy and sensorineural neural loss is observed in basal skull fractures.

Kim (2013) reported a rare case of Horner's syndrome due to stenosis of vertebral artery associated with nasal bone and left medial orbital wall fractures post sequeal to blunt trauma. The present case is in accordance to the study by (Martin and Fuent 2014) who also reported a similar case of Horner's syndrome due to vertebral artery stenosis.

Out of the 450 patients Maloney *et al*.(1980) observed 270 (60%) cases had an identifiable cause, of which 34(13%) had first order neuronal/central lesion, 120 cases (44%) had a second order (preganglionic) cause and 116 cases (43%) had third order /postganglionic cause.

Imaging modalities for Horner's syndrome

There are no clear imaging guidelines for diagnosing Horner's syndrome. Traditional modalities such as chest radiographs, flexion extension views of cervical spine and carotid Doppler ultrasound are still being advocated to diagnose second-order neuron Horner's syndrome such as Pancoast syndrome or aortic dissections. But on digital radiography 12- 90% of bronchogenic carcinomas are missed on chest radiographs especially if lesion is present in upper lobe or is peripheral (Shah *et al*.2003). Carotid Doppler US is the best modality used to assess stenosis or pathologies on carotids, distal cervical and intracranial vasculature but eventually cross-sectional imaging or conventional angiography is still required in almost all patients to confirm diagnosis (Davagnanam *et al*. 2013).

A simple protocol combining clinical and imaging techniques is being emphasized for early diagnosis of Horner's syndrome in table 2.

Table 2 Investigations for Horner's Syndrome

Horner's Syndrome	Proposed investigation
With localising clinical signs	MRI Brain with or without cervical and upper thoracic spine MRI (First order neuron) (Second & Third order neuron) CT angiogram MRI if iodinated contrast contraindication
Lacking localising signs but associated with trauma, malignancy and of acute onset	CT angiogram (urgent) MRI /CMERA if iodinated contrast contraindication
Non acute onset with no localising signs or trauma, malignancy	CT angiogram (within 6 weeks) MRI if iodinated contrast contraindication

1. First order lesions: Horner's syndrome due to first order neuron lesions with concurrent central nervous system signs is best imaged with MRI. If there are no brain or brainstem signs then imaging of cervical and thoracic spine with MRI should also be included.
2. Second order neuron and third order neuron lesion perpetuating Horner's syndrome but with no clinical signs. A CT angiogram emphasizing the orbits, circle of willis down to the aortic arch at about T4-T5 level is advised to patients with acute onset, trauma, malignancy on the day of clinical assessment to avoid delay. If iodinated contrast is contraindicated, fat suppressed MRI of brain with CMERA (Arterial phase contrast enhanced MR Angiography) is advocated (Reed *et al*.2008).
3. Performed in a timely manner without delays, the algorithm targets specific anatomical sites and pathologies of oculosympathetic pathways. In institutions lacking MRI; CT with contrast is arguably effective in assessment of head and neck areas (Ahmed and Branstetter 2008).

CONCLUSION

In the present study, the first case is Central Horner's syndrome which is a part of lateral medullary syndrome secondary to vertebral artery stenosis without carotid artery dissection and cerebral ischemia. There are limited reports of vertebral artery involvement as a cause of Horner's syndrome. The second case is Preganglionic Horner's syndrome due to Hodgkin's lymphoma (second-order neuron) and is explainable due to mediastinal compression, though an unusual presentation of Hodgkin's lymphoma.

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References

1. Moon J Y, Lee YH. Oculosympathetic paresis. N Z Med J. 2015 Jul 3; 128 (1417): 55-6.
2. Kong YX, Wright G, Pesudovs K, Day JO, Wainer Z and Weisinger HS. Horners syndrome. Clin Exp Optom. 2007; 90(5): 336-344.
3. Davagnanam I, Fraser CL, Miszkiel K, Daniel CS and Plant GT. Adult Horner's syndrome: a combined clinical, pharmacological, and imaging algorithm. Eye. 2013;27:291-98
4. Sivasshakthi K and Miller NR. Horner syndrome: clinical perspectives. Eye and Brain 2015;7:35-46.
5. Patel S and Ilsen PF. Acquired Horner's syndrome: clinical review. Optometry. 2003 Apr; 74(4): 245-56.
6. Kim CH. Horner's syndrome due to vertebral artery stenosis. J Craniofac Surg. 2013 Nov; 26(6): 2047-9. Doi: 10.1097/SCS.0b013e3182a41cb5.
7. Austin CP, Lessel S. Horners syndrome from hypothalamic infarction. Arch Neurol. 1991; 48:332-334.
8. Rosetti AO, Reichhart MD, Bogousslavsky j. Central Horner's syndrome with contralateral ataxic hemiparesis: a diencephalic alternate syndrome. Neurology.2003; 61:334-338.
9. Guy JR, Day AL, Mickle JP, Lee AG.Contralateral trochlear nerve paresis and ipsilateral Horner's syndrome. Am Opthalmol. 1989:107:73-76.
10. Kellen RI, Burde RM, Hodges FJ 3rd, Roper-Hall G. Central bilateral sixth nerve palsy associated with a

unilateral preganglionic Horner's syndrome. *J Clin Neuroopthalmol*. 1988;8:179-184.
11. Kim JS. Pure Lateral medullary infaction: clinical-radiological correlation of 130 acute, consecutive patients. Brain.2003;126:1864-1872.
12. Kobayashi N, Asamoto S, Doi H, Sugiyama H. Brown-Sequard syndrome produced by cervical disc herniation: report of two cases and review of the literature. Spine J.2003;6:530-533.
13. Simon SR, Dorighi JA, Branda RF, Ershler WB. Horner's syndrome: an unusual presentation of Hodgkin's disease. Med peadiatr oncol.1985;13(6): 390-1.
14. Martinmoro JG and Dela Fuente BP. Vertebral artery stenosis is a very unlikely cause of Horner's syndrome. *J Craniofac Surg*. 2014 Jul;25(4):1570. Doi:10. 1097/ SCS.0000000000000785.
15. Maloney WF, Younge BR, Moyer N. Evaluation of the causes and accuracy of pharmacologic localization in Horner syndrome. *Am J Ophthalmol* 1980; 90:394-402.
16. Reed DL, Garcon E, Smoker WR, Kardon R. Horner's syndrome: clinical and radiologic evaluation. Neuroimaging Clin N Am. 2008; 18(2)369-85.
17. Ahmed A Bransstetter BF. Ct verses MRI: still a tough decision. Otolaryngol Clin North Am.2008; 41(1): 1-22.

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