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A CASE SERIES ON HORNER SYNDROME AS UNCOMMON PRESENTATION OF COMMON DISEASES

Rahul Gupta[1], Ramji Sharma[2], Ashish Gupta[3], Mayank Gupta[4]

[1] Associate Professor, Department of Neurology, SMS Medical College, Jaipur
[2] Associate Professor, Department of General Medicine, SMS Medical College, Jaipur
[3] Associate Consultant, Department of Neurology, Jeevan Rekha Super speciality Hospital, Jaipur
[4] Associate Professor, Department of General Medicine, NIMS Medical College, Jaipur

Corresponding Author

Mayank Gupta

Associate Professor, Department of General Medicine, NIMS Medical College, Jaipur

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ABSTRACT

Horner´s syndrome (HS) results froma blockage of the sympathetic innervation to the eye at any point along its trajectory. It presents as ptosis, miosis, facial anhidrosis and enophthalmos. It can also occur as a part of Pancoast syndrome which also includes C8T1 roots. We describe three cases of common diseases presenting with HS; first case is a 51 year old male with mild neck pain who got operated for C6-C7 disc herniation and later proved to be Pancoast tumour in left lung apex. Second case is a 16 year old girl presenting with low grade fever, weight loss and left hand wasting diagnosed as left apical pulmonary tuberculosis. Third case is a young male who presented with right upper limb pain and later found out to have apical hydatid cyst. All these three cases had features of HS on examination.

KEYWORDS: Tuberculosis, Echinococcosis, Pancoast, Horner Syndrome.

INTRODUCTION

Horner syndrome (HS) was characterized first in humans by Johann Friedrich Horner in 1869[1]. HS results from interruption of the oculo-sympathetic pathway at anywhere along its course between the hypothalamus and the orbit[2]. HS is characterized by the triad of ipsilateral eyelid ptosis, miosis and facial anhidrosis. HS occurs as a part of Pancoast syndrome which involves brachial plexus. It was first described by Pancoast in 1924[3] in which an apical pulmonary tumour invades the C8 and T1 extraspinal nerves, the sympathetic chain, stellate ganglion and adjacent structures. Herniated cervical disc (HCD) is a common cause of spinal cord compression, but it is a rare cause of HS. Not many cases have been described in very few literatures with HCD[2] and T1-T2 disc herniation[4,5], but none have been described due to C7/T1 disc herniation. Few cases have been published in the literature due to tuberculosis[6-11] and hydatid cysts[12]among other possible causes of HS. Here we describe three cases with diseases which are otherwise common but rarely manifest with HS.

CASE 1

A 51-year-old male presented with a mild neck pain for 2 years. There was no history of trauma, arthritis. On admission, neurological examination revealed diminished sensation to pain and temperature on the left side restricted to T1-2 dermatome with a left complete Horner Syndrome (HS), including slight lid drop, miosis and facial anhidrosis. There was no motor weakness and deep tendon reflexes were normal. There was no bladder or bowel dysfunction. Magnetic resonance imaging of the cervical spine revealed a central and left-

sided extradural C7–T1 disc herniation, almost obliterating the right side of the spinal cord. The underlying cord did not show any high signal intensity on T2-weighted images.

CASE 2

A 16-year old girl presented with thinning of left hand and inner aspect of forearm which she noticed for last 2 months with associated sensory loss over the inner aspect of forearm. She also reported appetite and weight loss along with fever and night sweats for last 3 months. She denied any cough, haemoptysis, or breathlessness. On examination she had normal vitals except for low grade fever of 38.5^{o}C. her cranial nerve examination was unremarkable except for left incomplete ptosis and miosis of left eye consistent with left partial HS. She also had wasting of small muscles of hand with reduced bulk over left thenar eminence and oblique atrophy over medial aspect of forearm with sensory loss over C8-T1 dermatomal distribution with preserved reflexes. All these features were consistent with Pancoast syndrome and as she had constitutional symptoms, a malignant etiology was sought. The laboratory reported a haemoglobin of g/dl, MCV fl, MCH pg, erythrocyte sedimentation rate of mm/1 hour. She underwent MRI of the brain with gadolinium which ruled out brain lesions. The chest X-ray showed a rounded, radiopaque image with poorly defined edges in the right lung apex. A non-contrast CT scan of the chest was requested, which showed a patchy areas of consolidation with cavitation with centrilobular nodular opacities showing tree in bud pattern in left upper lobe consistent with tuberculosis. MRI brachial plexus confirmed CT findings. An ELISA test for human immunodeficiency virus was performed, the result of which was negative, as was the Mantoux test. Sputum microscopic examination (Ziehl-Neelsen) was repeatedly positive for acid-fast bacilli. With the diagnosis of Horner's syndrome as part of Pancoast syndrome secondary to pulmonary tuberculosis, treatment with isoniazid, rifampicin, pyrazinamide, and ethambutol was started and institutional discharge was granted.

CASE 3

A 35-year-old male presented with non-radiating, dull aching back pain over right scapular region and right arm for last 5 years. This was associated with numbness of inner aspect of the arm and later developed weakness of right hand resulting in difficulty performing fine motor tasks. This was also associated with decreased sweating over right side of the face and right arm and trunk when he noticed dry shirt over the same areas after exercise. He also had drooping of right eyelid for the same period of time but it was not significant enough to affect his vision. There was no history of head trauma, fever, vomiting, headache nor did he had difficulty in swallowing or breathing, nasal regurgitation. There was no weakness of lower limbs, difficulty in balancing, dizziness, vertigo, double vision or blurring of vision. Examination remarkable for right eyelid ptosis and miosis with normal extra-ocular movements. Trapezius and sternocleidomastoid muscles were weak with power of 3/5 without anytongue atrophy or fasciculations. Rest of the cranial nerve examination was normal. Motor bulk and tone were normal in all 4 limbs. Power (MRC Grading) in right shoulder abduction and adduction, flexion, extension was 4/5, elbow flexion and extension 4+/5, supination and pronation 4+/5, wrist extension and flexion 3/5, hand grip was weak, card test positive with normal deep tendon reflexes. Left upper limb and both lower limb examination was normal and bilateral plantar reflexes were flexor. Sensory examination showed absent pain touch and temperature sensation over right C8-T1 dermatome. Cerebellum and spine were normal. There was no neck rigidity and Kernig’s sign was absent. Chest skiagram showed a well-defined mass in right apical region. CECT thorax was donefor suspected malignancy which revealed a well-defined hypodense low attenuation area with enhancing internal septations in right lung upper lobe indenting the trachea suggestive of hydatid cyst was reported. USG thorax revealed a large well defined cystic lesion with multiple daughter cysts in right upper hemithorax which confirmed the diagnosis of Hydatid Cyst. Serological testing was positive for Echinococcus IgG enzyme-linkedimmunosorbent assay (ELISA), following which patient was referred to thoracic surgery department and underwent a surgical exploration. Near-total

excision was possible as cystwall was partly adherent to the C8-T1roots and the obtained pathologicalspecimen showed a lobulated cystic mass of 10 cm × 9 cm × 3 cm with the presence of daughter cysts that corroborated our diagnosis of hydatid cyst.

DISCUSSION

The oculo-sympathetic pathway begins at the posterior lateral aspect of the hypothalamus with a first-order neuron and extends down the spinal cord from C8 to T2through the brain stem. Second-order (preganglionic) neuronexit the spinal cord via the ventral roots and enter the paravertebral sympathetic chain which is located in the intermediolateral gray substance of the spinal cord at the level C8-T2 (cilio-spinal centre of Budge-Waller). The preganglionic pathway ascends in the cervical sympathetic chain to the superior cervical ganglion and passes over the apex of the lung. The third-order (postganglionic) neuron follow the carotid plexus into the skull, join with the ophthalmic nerve, and enter the orbit, which is superior cervical ganglion located at the level of C2-C3. HS may occur as a consequence of injury anywhere along this pathway [13,14].

HS is often classified as central (first-order neuron), preganglionic (second-order neuron) or postganglionic (third-order neuron) based on localization of the oculo-sympathetic pathway interruption. The preganglionic type is mostly caused by a tumour or trauma.

Tuberculosis is a disease that is prevalent in low and middle income countries like India and can affect any part of the body and present itself in various ways. We describe the case of apical pulmonary tuberculosis that presented as Horner's syndrome as a part of Pancoast syndrome. An analysis of 216 and 100 patients with Horner's syndrome, done by Giles and Henderson[6] and Keans[7] respectively, showed that none were due to tuberculosis. The first and only case of Horner's syndrome associated with tuberculosis reported in the English literature was in 1988 by Ismail et al.[7]Tuberculosis can cause it in an unusual way, due to involvement of both the preganglionic and postganglionic portions of the superior cervical sympathetic ganglion. Involvement of the post-ganglionic portion can be caused by retropharyngeal tubercular abscesses[9] and by cervical lymph node tuberculosis[10,11]. Tuberculosis of the pulmonary apex that generates involvement of the pre-ganglionic portion, as in this case, is rare. There are only four cases described in the literature of Horner's syndrome produced by isolated tuberculosis of the pulmonary apex[8,15-17]. This case highlights that tuberculosis needs to be considered among the etiological diagnoses of Horner's syndrome, in which the epidemiology and accompanying symptoms of the patient are of the utmost importance in the etiological orientation.

Cervical disc herniationis commonly encountered neurosurgical problem in routine neurosurgical practice. There are few reported cases of cervical disc herniation, presenting with Horner's syndrome[2,18-21](Table 1). Of all the previously reported cases, 3 cases had disc herniation at C5/6 level, 2 at C4/5 level and 1 at C6/7 level. Only two cases of cervical disc herniation at C7/T1 have been reported but none had HS. Our case is the first case with disc herniation at C7/T1 level presenting with HS.

TABLE 1. Few reported cases of cervical disc herniation, presenting with Horner's syndrome

Sl. No.	Case (Publication Year)	Patient Age (years)/ Sex (M/F)	Level of Herniation	History of prior trauma (time to IDH)	Other clinical Findings	Diagnostic finding	Surgical Approach
1	Marega (1959)	41 M	C5/6	No	Quadriparesis	Myelography: complete blockage	Posterior laminectomy
2	Dürig et al (1977)	52 F	C5/6	Yes (8 years)	-	Myelography: complete blockage	Posterior laminectomy

3	Sprick et al (1991)	49 F	C6/7	Yes (6 years)			Anterior discectomy, with fusion
4	Ma et al (2012)	41 M	C4/5	No	Numbness below right side of the T8 dermatome.	MRI: upward migrated and large left paramedian disc herniation with severe unilateral spinal cord compression at the C4-C5 level	Microsurgical anterior approach
5	Meng et al (2016)	51 F	C5/6	No	progressive right hemiparesis, numbness in the lower trunk and limbs on the left side, mild neck pain	MRI: large central and right-sided extradural C5–6 disc herniation	Anterior cervical corpectomy (C5) and reconstruction
6	Baudracco et al (2017)	45 F	C4/5		Severe nuchal pain	MRI: central disc osteophyte, paracentral disc extrusion at C4/5 with cranial extension; OP: PLL tear, ossification of PLL with adhesion to dura	Anterior discectomy, with fusion

According to Woischneck et al[22], disc herniation at the level of C7/T1 is so rare, that malignancies must be included in the differential diagnosis of pathologies at the cervicothoracic junction. In their case, their patient was predisposed to IDH due to age-related degenerative changes of the spine. In his case he hypothesized that in their patient, cervicothoracic disc herniation resulted from mechanical overload of the cervicothoracic

junction between the cervical spondylosis and the rigid axis of the thoracic spine, due to axial forces on the spine caused by trauma. Gunasekaran et al[23] reported a case with C7/T1 herniation and hypothesised that a pre-existing OPLL that led to adhesion, erosion and subsequent dural defect creation. This may have allowed herniation of the native disc into that intradural space through the defect.

Echinococcal cysts are slowly enlarging, generally remain asymptomatic until their expanding size or their space-occupying effect in an involved organ elicits symptoms. These infections are most prevalent in those areas where livestock is raised in association with dogs. Pulmonary hydatid cysts are usually asymptomatic and clinical features are only evident when they are large enough to exert mechanical compression or are complicated due to their rupture[12]. Our patient presented with HS as a part of Pancoast syndrome. The majority of patients have a longstanding history of contact with domestic animals (dogs) and/or farming animals (sheep). Our patient had history of rearing pet dogs for last 7-8 years and used to look after them by himself suggesting frequent contact with faeces. Hydatid cyst manifesting as Pancoast tumour is a are complication of the infection[24,25].

CONCLUSION

In conclusion, this case series highlights the importance of considering a wide range of potential causes for Horner's syndrome (HS), including common infections such as tuberculosis, and hydatid cysts. The cases presented emphasize the need for careful differential diagnosis and re-evaluation, particularly when a patient's symptoms do not resolve as expected with initial treatment, as seen in the case of cervical disc herniation. Furthermore, the discussion underscores the relevance of epidemiological factors, such as tuberculosis prevalence in low- and middle-income countries, and the patient's history, such as contact with animals in the case of hydatid cysts; as they can disrupt the oculo-sympathetic pathway, leading to the clinical manifestation of HS. These factors should guide clinicians in identifying the underlying cause of HS, ensuring that appropriate and timely interventions are provided.

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