

# Horner's Syndrome: A Case Report and Review of the Pathophysiology and Clinical Features

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## ABSTRACT

*Tumours of the superior sulcus of the lung, commonly referred to as Pancoast tumours, present with characteristic clinical symptoms and signs. An interesting case of a patient who presented with such a tumour is presented. The pathophysiology, clinical features and approach to management are reviewed.*

**Keywords:** Horner's syndrome, Jamaica, Pancoast tumour, superior sulcus tumour

# El Síndrome de Horner: Reporte de un Caso y Revisión de la Fisiopatología y las Características Clínicas

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## RESUMEN

*Los tumores del sulcus pulmonar superior, comúnmente conocidos como tumores de Pancoast, presentan signos y síntomas clínicos característicos. Se presenta el interesante caso de un paciente que acudió con un tumor de este tipo. Se pasa revista a la patofisiología, las características clínicas, y el enfoque del tratamiento.*

**Palabras claves:** Síndrome de Horner, Jamaica, tumor de Pancoast, tumor del *sulcus* superior

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## INTRODUCTION

Tumours of the superior sulcus of the lung are typically bronchogenic in origin. As such, there is a strong causal association with smoking. These tumours may invade the ribs, subclavian vessels, brachial plexus, stellate ganglion and adjacent vertebral bodies resulting in characteristic clinical features including Horner's syndrome.

A male patient who presented to the Emergency Department at the University Hospital of the West Indies with features of Horner's syndrome is described. He was subsequently diagnosed with a Pancoast tumour. A discussion of the patient's presentation ensues. The clinical features of Pancoast tumours and their pathophysiologic basis are reviewed.

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## CASE REPORT

A 59-year old male with no known chronic medical illnesses presented with a five-day history of constipation and not being able to walk. He reported no history of trauma, back pain, viral type symptoms, or nausea but gave a six-month history of shortness of breath. He denied cough, haemoptysis, palpitations, or chest pain. Of note, he had a 30-pack-year smoking history and reported significant weight loss and anorexia over several months.

On examination, the patient was tachycardic at 102 beats/minute, tachypnoeic at 32 breaths/minute and had an oxygen saturation of 84% on room air. He had right-sided ptosis and obvious anhidrosis on the right side of his face (Horner's syndrome) with distended neck veins on the right and fullness in the right supraclavicular region (Fig. 1). His right pupil was constricted (Fig. 2). Cranial nerve examination was otherwise unremarkable. Respiratory examination revealed decreased chest expansion on the right, with tracheal deviation to the left, dullness to percussion and absent breath sounds in the right upper-mid zones. He was fully alert with a Glasgow coma scale of 15/15. Grade 5 power was present in the left upper limb and grade 4 in the



Fig. 1: Right sided ptosis, distended right-sided neck veins and right supraclavicular fullness are evident.

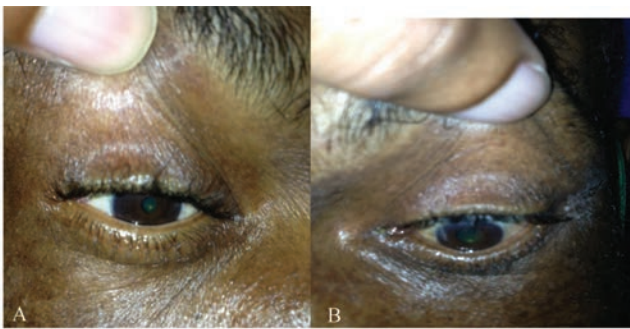


Fig. 2: The right (A) pupil is constricted compared to the left (B) due to miosis of the right pupil as a result of Horner's syndrome.

right. There was grade zero power in both lower limbs and a sensory level (light touch) at T4 dermatome on the left and C7 on the right with no appreciation of pinprick below T7 bilaterally. Reflexes were decreased at both ankles and tone was decreased in both lower limbs. His abdominal examination was normal.

The computed tomography (CT) scan of the chest revealed a soft tissue density in the right upper hemithorax extending to the chest wall (Fig. 3A). Associated destructive lytic bony changes were noted at the T2 and T3 vertebral levels (Fig. 3B). Similar density material was observed within the spinal canal at these levels. Bony changes include lytic destructive lesions of the right first rib, lytic destructive changes of the right second rib. Lytic destructive changes were also observed in the right third rib transverse process, pedicle, spinous process, both laminae and the vertebral body of T3, and in the right fourth rib and transverse process (of T4). A small right pleural effusion was seen. There was

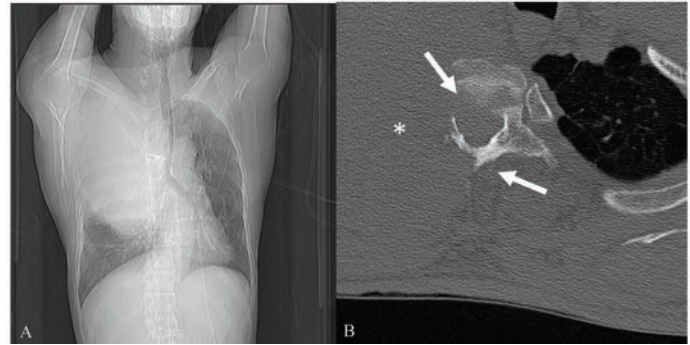


Fig. 3: Large soft tissue mass is present in the upper right hemithorax (A). The second thoracic vertebra (B). The lung tissue on the right is completely replaced by the tumour (asterisk) which has invaded and destroyed the right pedicle and spinous process (arrows).

evidence of cervical spondylosis most marked at C5/C6 and C6/C7. The patient was evaluated by the neurosurgical team on call. He was referred for outpatient evaluation and to be considered for hospice care. However, he demised at home before a lung biopsy or further work-up could be performed. No postmortem was performed.

## DISCUSSION

Horner's syndrome is characterized by ipsilateral blepharoptosis, miosis and anhidrosis. In patients with lung tumours, this results from invasion of the preganglionic neurons exiting the ventral spinal roots. These fibres arch over the apex of the lung and ascend in the cervical sympathetic chain to the superior cervical ganglion. Involvement of these preganglionic fibres in apical lung tumours produces the features of Horner's syndrome, all of which were evident in the index case. Fullness of the right supraclavicular region was due to the tumour itself, and distension of the veins in this area resulted from compression of the right subclavian vein. The patient also had invasion of the vertebral bodies causing back pain, inability to walk and a sensory level which was subsequently seen on CT.

Preganglionic Horner's syndrome is the most common variety seen. Two other types, central and postganglionic, are recognized. Central Horner's syndrome is uncommon. Most commonly it results from infarction of the vascular territories of the posterior inferior cerebellar artery or distal vertebral artery territory. These patients will have additional brainstem, spinal cord or hypothalamic signs and symptoms such as dysphagia, ipsilateral facial anaesthesia, contralateral anaesthesia of trunk/limbs, cerebellar ataxia and rotary nystagmus depending on the actual lesion. Postganglionic Horner's syndrome may result from lesions involving the internal carotid artery, skull base or cavernous sinus/orbital apex. Third, fifth and sixth cranial nerve palsies suggest lesions involving the cavernous sinus. The most important cause of postganglionic Horner's syndrome is dissection of the carotid artery. It is possible to distinguish central and preganglionic Horner's syndrome from postganglionic Horner's syndrome.

ner's syndrome on examination of the eyes. Normal persons experience pupillary dilation on instillation of a drop of 10% cocaine. This does not occur in patients with Horner's syndrome. Subsequent instillation of 1% hydroxyamphetamine produces pupillary dilation in patients with central and preganglionic Horner's syndrome as the postganglionic fibres are intact. This will not occur in postganglionic Horner's syndrome (1).

Pancoast initially described the clinical and radiological characteristics of superior sulcus tumours in 1924 (2). Initially thought to arise from epithelial cell rests in the fifth branchial cleft, they were subsequently shown to be primarily due to bronchogenic tumours (3). These tumours typically arise in the superior sulcus of the lung and may invade the ribs, subclavian vessels, brachial plexus, stellate ganglion and adjacent vertebral bodies. In one case series, rib involvement was evident on chest X-ray or CT scan in 51.6% of the patients and most commonly affected the first and/or second rib (4). The first thoracic nerve root was the commonest nerve root involved in up to 94% of the cases (4). Sixty-eight per cent of patients had involvement of the T2 nerve root. The index patient did not have radiological evidence of T1 vertebral involvement, however, the T2–T4 vertebral bodies showed infiltration by tumour (4). Bronchogenic cancers are

the commonest cause of a Pancoast tumour; as such, a history of smoking is commonly found, as in this case. Tumours are often seen on chest X-ray and further characterized on CT imaging. Definitive diagnosis is usually *via* percutaneous needle biopsy. Complete blood count, liver function tests, electrolytes, CT abdomen, bone scan and mediastinoscopy are important to assist with staging which determines whether management is surgical or medical. A magnetic resonance imaging of the brain is advised if symptoms suggest cerebral metastases. Patients such as ours with metastatic disease may benefit from chemotherapy for palliation.

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