Case Report

Collet Siccard Syndrome without skull base metastasis: A rare presentation of bronchogenic adenocarcinoma

Shobha Sudarshan Shetty*, Ramaprakasha S, Ivor D'sa, Glaxon Alex and Sandheep George Villoth

Department of General Medicine, K. S. Hegde Medical Academy, Derlakatte, Mangalore, India.

*Correspondence Info:
Dr. Shobha Sudarshan Shetty,
Associate Professor,
Department of General Medicine,
K.S.Hegde Medical Academy, Derlakatte, Mangalore, India.
Email: shobhashetty22@gmail.com

Abstract
A 54 year old female on evaluation of dysarthria was found to have left IX, X and XII cranial nerve palsy. General physical examination revealed bilateral cervical level V multiple lymph nodes which were firm to hard and non-tender; respiratory system examination suggested left sided mild pleural effusion. MRI brain and cervical spine did not show any evidence of meningeal enhancement or skull base metastasis. Bronchoscopy revealed a mass lesion in left main bronchus. Her cranial nerve symptoms were attributed to Collet-Sicard syndrome because of the lymph node metastasis from lung cancer. It is a rare case of Collet-Sicard syndrome resulting from lymph node metastasis of adenocarcinoma of lung and is important as a differential diagnosis of lower cranial nerve palsy.

Keywords: Adenocarcinoma of lung, Collet Siccard Syndrome, Lower cranial nerve palsies, Skull base metastasis

1. Introduction
Metastatic tumors involving the central nervous system (CNS) are common and occur in 20 to 40 % of patients with a systemic malignancy. Common primary sources include lung, breast, and skin malignancies, and less frequently gastrointestinal, renal, prostate, testicular, and ovarian cancers. Metastatic involvement of the cranial base may also be seen in 4% of patients with systemic malignancies. Collet-Sicard syndrome is caused by a lesion that involves the lower 4 cranial nerves.

2. Case report
A 54 year old female was admitted with history of mild slurring of speech since two months which became worse since two days. There was no history of headache, diplopia, difficulty in swallowing, stridor, limb weakness or bladder and bowel symptoms. She also gave history of left shoulder pain radiating to left arm since two months. General physical examination revealed bilateral cervical level V multiple lymph nodes, firm to hard and non-tender. Neurological examination revealed diminished gag reflex with decreased palatal and pharyngeal sensations on the left side. There was weakness of the left half of the tongue. She was noted to have uvular deviation to the right from a paretic left palate, a decreased left gag response, left tongue deviation. There were no sensory deficits or motor weakness in the limbs, and no meningeal signs as well. Respiratory system examination revealed dull note with diminished breath sound intensity in left infrascapular area.

Clinical impression was left IX, X and XII cranial nerve palsy with cervical lymphadenopathy. Investigations
revealed normal complete hemogram, ESR, blood sugar, liver and renal function tests. Viral markers for HIV, HBsAg and HCV were nonreactive; Mantoux test was negative. Chest X-ray revealed prominent left hilum, homogenous opacity with irregular borders in left para-cardiac region and blunting of left costo-phrenic angle. Ultrasonography (US) neck suggested bilateral level 5 and 7 supraclavicular multiple lymph nodes, with the largest measuring 3.5 × 2 cms. Excisional biopsy of lymph node suggested metastatic adenocarcinoma. Meanwhile, MRI of brain and cervical spine was normal, and there was no evidence of meningeal enhancement or skull base metastasis. Bronchoscopy revealed nodular growth in the left main bronchus.

Fig: 1. Left sided hypoglossal nerve palsy due to pressure effect by enlarged lymphnodes

Fig: 2. Bronchoscopy picture showing nodular growth in left main bronchus

Fig: 3 Histopathology of lymphnode showing sinusoids filled with tumour cells(X100, H&E) and pleomorphic tumour cells arranged in tubules (X400, H&E)
3. Discussion

Cranial neuropathy in a patient with systemic cancer may be due to skull base metastasis, meningeal carcinomatosis or at times due to contiguous spread from a head and neck malignancy.\textsuperscript{2,3} In some instances, the presentation may be confusing and mimic a cerebrovascular, infectious, or metabolic affliction. Greenberg \textit{et al.}\textsuperscript{4} described cranial dysfunction syndromes caused by skull base metastases. These include Collet-Siccard syndrome,\textsuperscript{5} which is a palsy of the lower four cranial nerves; Villaret syndrome,\textsuperscript{6} which is a palsy of the lower four cranial nerves with ipsilateral Horner syndrome; Occipital condyle syndrome,\textsuperscript{7} which consists of unilateral occipital pain with ipsilateral 12\textsuperscript{th} nerve paresis; Jugular foramen syndrome\textsuperscript{8} with dysphagia and neck pain; Parasellar syndrome\textsuperscript{9} with extra ocular palsy and isolated 3rd, 6th or 7th cranial nerve palsies.

Collet-Siccard syndrome results in the paralysis of vocal cords, palate, trapezius and sternocleidomastoid muscles. Clinically, it results in anaesthesia of the larynx, pharynx and soft palate. It is caused by lesions at the base of the skull affecting the lower cranial nerves. It is associated with various etiologies of tumoral and other origins. The differential diagnosis includes non-tumoral factors causing Collet-Siccard syndrome like fractures at the base of the skull, aneurysms, inflammatory processes (osteomyelitis, Paget’s disease) or other alterations such as diabetes mellitus or porphyrias. However, considering a potential tumor cause in the differential diagnosis is important. The patient described above had presented with Level 5 and 7 cervical lymph node metastases; this presentation in the absence of skull base metastasis along with Collet-Siccard syndrome has not been described before. The patient died before any potential treatment benefit could be assessed. Carcinomatous adenopathies and reticulosis such as Hodgkin’s disease have also been reported to be associated with lower cranial nerve palsy.\textsuperscript{10}
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