COLLET-SICARD SYNDROME: CASE REPORT OF A RARE PRESENTATION OF METASTATIC GASTRIC ADENOCARCINOMA.

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ABSTRACT

Background: Collet-Sicard is a rare syndrome that involves paralysis of unilateral lower cranial nerves (IX, X, XI and XII) due to lesions at the base of the skull. It is associated with various neoplastic and non-neoplastic aetiologies.
Case Report: We report the case of an adult who presented with an 8 months history of dysphagia, dysphonia, supraclavicular lymphadenopathy, unilateral facial and shoulder weakness. A diagnosis of metastatic adenocarcinoma was confirmed by tissue biopsy. This is a case report of an unusual presentation of Collet-Sicard syndrome secondary to metastatic gastric carcinoma. Because of delay in reporting to hospital and diagnosis, the patient died while being worked-up for radiotherapy and surgery.
Conclusion: Delay in diagnosis of Collet-Siccard syndrome is common, and this condition should be considered in patients who present with a constellation of lower cranial nerve palsies.

Keywords: Collet-Sicard syndrome, cranial, nerve, carcinoma, metastasis.

BACKGROUND

Collet-Sicard Syndrome was first described by a French pathologist called Frederic Collet in 1915. It was initially referred to as "glossolaryngoscapulopharyngeal hemiplegia". Sicard in 1917 later independently described the same condition as the syndrome of the condyloposterior-lacerated foramen.Hence, the name 'Collet-Sicard' syndrome is used to describe the disorder.[1,2] It is characterized by complete set of unilateral lower cranial nerve palsies [i.e. IX, X, XI, XII]; giving rise to paralysis of the vocal cord, palate, trapezius muscle, sternocleidomastoid muscle; as well as lost of sensation in the larynx, pharynx and soft palate.[3] It can be caused by trauma, neoplasm, vascular aneurysms and dissections, jugular vein pathologies, infections and diabetes mellitus.[3–7] The syndrome may be observed prior to the diagnosis of the primary conditions. Hence, we present this case of Collet-Sicard syndrome as a result of metastatic malignant lesion from the stomach.

CASE PRESENTATION

We report a 47 year old male, who presented with complaints of inability to swallow (dysphagia), dysphonia, dysarthria, and weakness of the left side of the face with deviation of the mouth to the right, difficulty raising his left upper limb above his shoulder, weight loss and left sided neck swelling. These symptoms began 8 months prior to presentation, and subsequently progressed insidiously. He later developed chronic cough

with associated haemoptysis. There was no history of trauma to the head or neck, and no past medical history of note. He was initially seen at a peripheral hospital were a diagnosis of primary neuromuscular disorder was made. On physical examination, the patient had significant weight loss, with enlarged, hard and fixed left supraclavicular lymph nodes. He had a lower motor neuron left VII cranial nerve palsy**.** In addition, there was deviation of the tongue to the left side with associated atrophy and fasciculation (cranial nerve XII) (Figure 1). Moreover, there was loss of gag reflex on the palate (cranial nerves IX and X) with associated weakness and wasting of the left trapezius and sternocleidomastoid muscles on the same side(cranial nerve XI) **(Figure 2).** A **c**linical assessment of Collet-Sicard syndrome was made and the patient was evaluated as follows.

INVESTIGATIONS

- **Computerised tomography (CT) scan**: Serial axial section of the head and neck showed enhancing necrotic lymph nodes involving the left supraclavicular region. Axial section of the chest and upper abdomen revealed multiple nodular lesions in both lung fields and heterogeneous enhancing soft tissue mass of the posterior wall of the stomach**.** There was no obvious lesion observed in the brain.
- **Fine Needle Aspiration Cytology** (FNAC) of the cervical lymph nodes was suggestive of metastatic malignant adenocarcinoma from the stomach; this was confirmed by subsequent excisional biopsy of the lymph nodes.

Wereceived consent from the patient to write this case report and to take pictures.

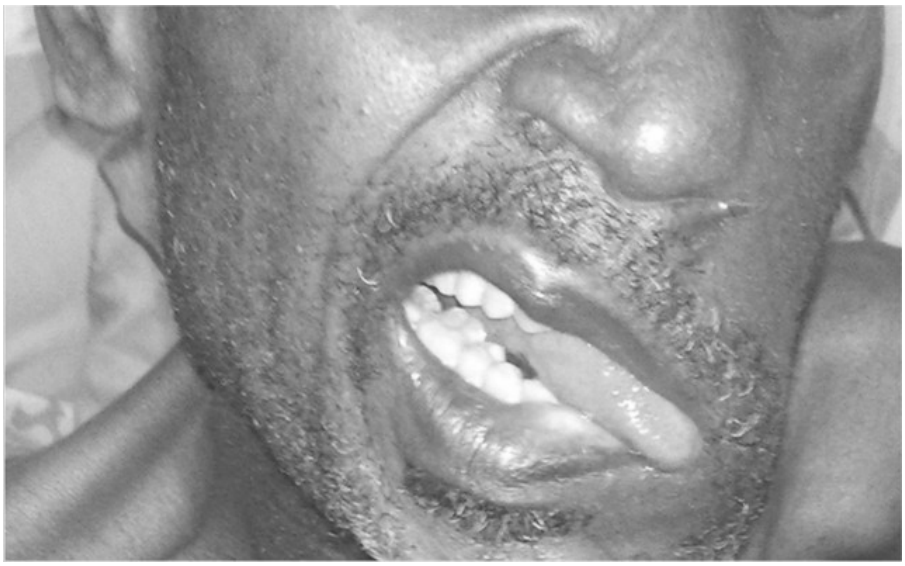

FIGURE 1: Left side lower motor neuron facial nerve palsy associated with left side hypoglossal nerve palsy.

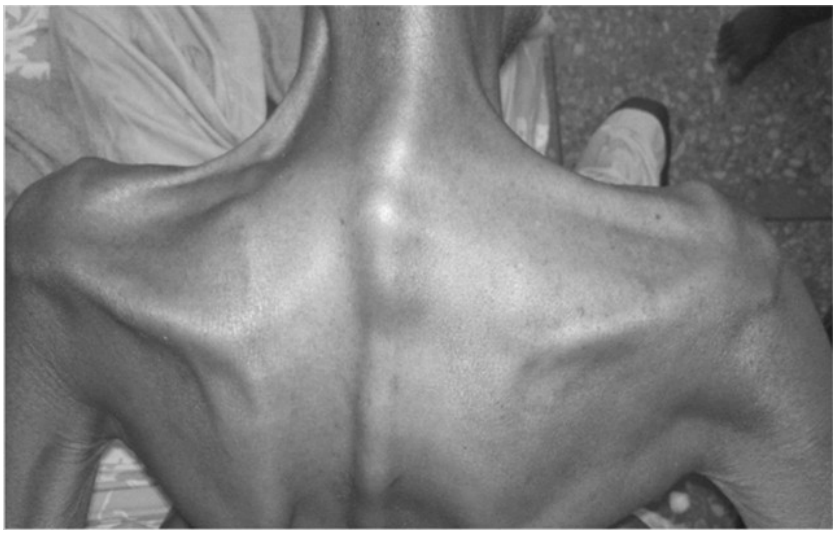

FIGURE 2: Generalised weight lost with atrophy of the left sternocleidomastoid muscle and scapular winging.

DISCUSSION

The description of our patient fits this rare clinical syndrome of Collet-Sicard, which is associated with a broad range of symptoms due to IX, X, XI and XII cranial nerve palsy without any features of cerebral ischaemic change.

Various causes of Collet-Sicard syndrome have been reported in the literature, which include: fractures and dissection of skull-base,[3] neoplasms,[2,7] vascular events like s*igmoid-jugular complex* thrombosis,[5] and infections.[8] Collet-Sicard syndrome may sometimes mimic *neuralgic amyotophy.*[9] T*he aetiology of* Collet-Sicard syndrome *can broadly be categorized into neoplastic (primary or metastatic),*[4,10] *or non-neoplastic causes due to trauma, infections, vascular and metabolic conditions.*[3,11]

Our patient had Collet-Sicard syndrome from metastasis. Primary presentation of metastatic cancer as Collet-Sicard syndrome have been reported in the literature,[2,4,6,10] However, primary presentation of gastric carcinoma as Collet-Sicard syndrome is quite rare in the literature. Metastasis of abdominal cancers to the jugular complex area is usually due to haematogenous spread from the primary site, but may also result from retrograde dissemination via the Batson's plexus – a valve-less venous plexus connecting the abdominal structures with dural and epidural veins.[2,6]

Collet-Sicard syndrome typically presents with unilateral paralysis of the lower four cranial nerves and c*linical presentation is influenced by the extent of cranial nerves involvement. Clinical features mainly include dysphonia*, dysphagia, dysarthria, tongue deviation, atrophy and fasciculation; weakness of the shoulder, wasting of trapezius and sternocleidomastoid muscles and winged scapula on the side of lesion. Our patient presented with all these features. Although he had a history of chronic cough with haemoptysis, our evaluation for tuberculosis was negative, a common chronic infection in the tropics. Collet-Sicard syndrome may also occur from infections.[8]Other clinical features of Collet-Sicard syndrome may include obstructive urinary symptoms, breast swelling, bone pains, recurrent infections, tetany or multi-organ affection.

An important differential is the Villaret's syndrome, which has sympathetic dysfunction (Horner's syndrome) in addition to the cranial nerves dysfunction, due to granulomatous lesion in the posterior retroparotid space..[11,12]

Our patient had no meningeal signs, since lower cranial neuropathy in a patient with

systemic cancer may also be secondary to meningeal carcinomatosis.[13]

Our patient had a CT-scan done, as Magnetic Resonance Imaging (MRI) was unavailable at that time. MRI is better than CT-scan in evaluating patients with this condition and may be able to detect lesions that are missed by CT-scan. Lucato et al. reported the use of Multi-detector-row computed tomography in the diagnosis of Collet-Sicard syndrome resulting from vascular abnormalities of the internal carotid artery;[14] while Stasolla et al. recently reported that high resolution contrast CT may suffice in the diagnosis, which may be useful in resource poor settings like ours.[15]

CONCLUSION

- AlthoughCollet-Sicard syndrome is considered a rare clinical entity; it may be a primary presentation of metastatic cancers.
- Its incidence may probably be higher than commonly estimated because of the number of undiagnosed cases.
- Delay in diagnosis of this condition may result in a fatal outcome, as occurred in our patient.
- It should therefore be considered in patients presenting with clinical features of multiple lower cranial nerve palsies.

REFERENCES

1. Collet F. Sur un nouveau syndrome paralytique pharyngo-larynge par blessure de guerre (hemiplegie glosso-laryngo-scapulo-pharyngee). Lyon Med. 1915;124:121–9.
2. Laigle-Donadey F, Taillibert S, Martin-Duverneuil N, Hildebrand J, Delattre J-Y. Skull-base metastases. J Neurooncol. 2005;75(1):63–9.
3. Kwon HC, Cho DK, Jang YY, Lee SJ, Hyun JK, Kim TU. Collet-sicard syndrome in a patient with jefferson fracture. Ann Rehabil Med. 2011;35(6):934–8.
4. Basu S, Nair N. Relapse of cervical cancer presenting as symptoms of Collet-Sicard syndrome with metastatic subcutaneous and adrenal deposits. Lancet Oncol. 2006;7(7):610.
5. Handley TPB, Miah MS, Majumdar S, Hussain SSM. Collet-sicard syndrome from thrombosis of the sigmoid-jugular complex: a case report and review of the literature. Int J Otolaryngol. 2010;2010.
6. Hayward D, Morgan C, Emami B, Biller J, Prabhu VC. Jugular foramen syndrome as initial presentation of metastatic lung cancer. J Neurol Surg reports. 2012;73(1):14–8.
7. Verma V, Singh MP, Gupta AK, Singh TP, Ameesh MM, Jaiswal N. Jugular Foramen Schwannoma Presenting as Collet-Sicard Syndrome – A Rare Entity. JIACM. 2011;12(3):227–229.
8. Sibai TA, Ben-galim PJ, Eicher SA, Reitman CA. Infectious Collet-Sicard syndrome in the differential diagnosis of cerebrovascular accident : a case of head-to-neck dissociation with skull-based osteomyelitis. Spine J. 2009;9(4):e6–e10.
9. Larson W, Beydoun A, Albers J, Wald J. Collet – Sicard syndrome mimicking neuralgic amyotrophy. Muscle Nerve. 1997;20:1173–1177.
10. Villatoro R, Romero C, Rueda A. Collet-Sicard syndrome as an initial presentation of prostate cancer: a case report. J Med Case Rep. 2011;5(1):315.
11. Caldemeyer S, Mathews V, Azzarelli B, Smith R. The Jugular Foramen : A Review of Anatomy , Masses , and Imaging Characteristics. RadioGraphics. 1996;17:1123–1139.
12. Boon P, Reuck J De, Velde E Van De. Villaret ' s syndrome due to thrombosis of the jugular vein. Clin Neurol Neurosurg. 1990;92:337–341.
13. Bone I, Hadley DM. Syndromes of the orbital fissure, cavernous sinus, cerebello- pontine angle, and skull base. J Neurol Neurosurg Psychiatry. 2005;76 Suppl 3(Suppl III):iii29–iii38.
14. Lucato LT, Passos RBD, Campos CR, Conforto AB, McKinney AM. Neurological picture. Multidetector-row computed tomography in the diagnosis of Collet-Sicard syndrome. J Neurol Neurosurg Psychiatry. 2008;79(5):521.
15. Stasolla A, Bykowski J, Chung R, Mafee MF. Jugular fossa tumors: Role of imaging. Oper Tech Otolaryngol. 2014;25(1):66–73.