

A Case Report of Collet-Sicard Syndrome Resulting From IJV Thrombosis

K.C Ranjit, M.K Surag, P.K Mukundan, Dr. Ajith. M. K, C. Jayakrishnan

Department of Internal Medicine/Pariyaram Medical College/KUHS/South India

Abstract: A middle aged lady presented with fever, generalized tiredness and dysphagia. Neurological evaluation revealed weakness of pharyngeal muscles, uvula, palate with absent gag reflex and a weak sternocleidomastoid muscle. Involvement of 9,10,11,12 cranial nerves in the absence of Horner's syndrome confirmed the diagnosis of Collet-Sicard syndrome. MR imaging studies revealed a Sigmoid Sinus Thrombosis with proximal IJV involvement.

Keywords: Collet-Sicard syndrome, Villaret syndrome, IJV thrombosis.

I. Introduction

Collet-Sicard Syndrome is a rare condition associated with unilateral palsy of 9th, 10th, 11th, 12th cranial nerves. It is distinguished from the commoner Villaret syndrome by the sparing of sympathetic trunk in the former. We are reporting the case of a 41 yr old female with no previously diagnosed comorbidities who presented with fever of 1 month duration and tiredness with difficulty in swallowing since 3 days.

The veins of head and neck are less susceptible to thromboembolism due to the relatively valveless nature and the assistance of gravity in emptying the veins in upright position[1]. Hence, thrombosis of Internal Jugular vein is considered rare. Here we describe a case of IJV-Sigmoid sinus thrombosis resulting in Collet-Sicard syndrome.

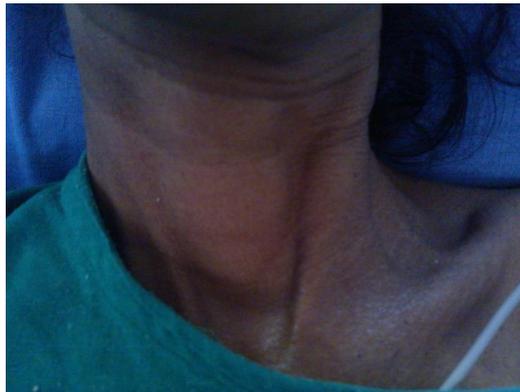
II. Case Report

A 41 yr old lady evaluated from elsewhere as a case of pyrexia of unknown origin presented to us with 3 days history of generalized tiredness and difficulty in swallowing. There was no history of cough, ear discharge, seizures, loss of consciousness or altered sensorium. On examination, patient had deviation of tongue towards right side (**Figure 1**), weakness of the soft palate during phonation and the base of uvula pulled towards left side. Patient also had a weak cough along with a weak gag reflex on the right side. She was unable to raise her head off the bed (**Figure 2**). Rest of the neurological examination was found to be normal. There was no Horner's syndrome. All these features showed involvement of lower cranial nerves namely 9, 10, 11, 12 cranial nerves without features of Horner's syndrome. Hence a clinical diagnosis of Collet-Sicard Syndrome was made. This syndrome was distinguished from Villaret syndrome by lack of sympathetic involvement. After MR imaging studies of brain including MR venogram, the condition was diagnosed as Sigmoid sinus thrombosis with proximal IJV involvement (**Figure 3**). Causes for the thrombosis were sought and relevant things to be ruled out was infection and malignancy as the patient was having prolonged fever, weight loss, tiredness. Routine blood examination revealed a raised ESR with a neutrophilic predominance to the total leucocyte count, but the exact foci of infection could not be located even after an extensive search. No evidence of vasculitis. She was started on IV antibiotics, antifungals and heparin for anticoagulation. In spite of all these measures she had a downhill course and finally succumbed to the disease with Multi-organ dysfunction syndrome.

III. Figures



Figure (1) showing deviation of tongue due to the weakness of the 12th cranial nerve



Figure(2) showing weakness of the Rt. sternocleidomastoid muscle from involvement of the Rt. 11th cranial nerve



Figure(3) MR Venogram showing sigmoid sinus thrombosis with proximal IJV involvement

IV. Discussion

Thrombosis of the sigmoid-jugular complex is frequently asymptomatic because the venous drainage system of the brain has sufficient alternative routes or pathways. Blockage of the cerebral venous drainage through the jugular foramen can cause raised intracranial pressure, which can lead to epileptic seizures[2].

In our case, CN IX–XII were involved which suggested a pathological process located near the jugular foramen. This foramen courses anterolaterally as it exits the skullbase and consists of a smaller anteromedial portion (the pars nervosa) and a larger posterolateral portion (the pars vascularis) that are separated by a complete or incomplete fibrous or bony septum. The pars nervosa contains CN IX– XI, the inferior petrosal sinus, and the meningeal branch of the ascending pharyngeal artery. The pars vascularis contains the sigmoid-jugular complex. The jugular foramen varies greatly in size, but averages 15mm in length and 10mm in width [3]. In terms of imaging, the most sensitive examination was MRI in combination with magnetic resonance venography [4], hence an MRI with MR venogram was done. T1 weighted and T2 weighted MRI showed hyperintense signal from the IJV thrombosis.

Involvement of the jugular foramen region leading to lower CN involvement is characterised by several eponymous syndromes, where because of their close anatomical relationship multiple CN involvement is the rule. CSS appears to be the most likely syndrome related to this presentation.

Frederic Collet and Jean Sicard provided the original descriptions of CSS based on posttraumatic cases during World War I in which the location of the lesions was demonstrated via radiographic studies as a result of the presence of metallic fragments [5,6].

In this case the cause of thrombosis of the IJV remains unknown. Nearly 20%–25% of cerebral venous thrombosis have no known aetiology [7]. With a presentation of pyrexia and negative screening for vasculitis, thrombophilia and connective tissue disease, we concluded that the most likely cause of IJV thrombosis was infection.

V. Conclusion

Despite the fact that this is a rare disease, any patient presenting with multiple lower cranial nerve palsies, the diagnosis of Collet-Sicard syndrome has to be considered. Early recognition and appropriate treatment may result in significant clinical improvement and outcome. A multidisciplinary approach to the management of the CSS patient is important in achieving successful functional recovery.

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