COLLET-SICARD SYNDROME SECONDARY TO THE GLOMUS JUGULARE PARAGANGLIOMA

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ABSTRACT

Collet-Sicard syndrome is a constellation of cranial nerve IX,X,XI and XII palsies secondary to either neoplastic or non-neoplastic lesions at jugular foramen. We present here a case of a 71-year-old male who presented with dysphagia, left sided facial palsy and change in voice and was later diagnosed as Collet-Sicard syndrome secondary to the glomus jugulare paraganglioma.

Introduction

Collet-Sicard syndrome (CSS) also called as condylar jugular syndrome is a group of cranial nerve palsies i-e cranial nerve (CN) IX, X, XI and XII; which occurs secondary to the either neoplastic or non-neoplastic skull base lesions at the jugular foramen and the hypoglossal canal.[1,2] Traumatic skull base fractures are the most common non-neoplastic cause of Collet-Sicard syndrome; other non-neoplastic causes include osteomyelitis and Paget's disease. Whereas, amongst the neoplastic causes includes glomus jugulare tumor, schwannoma and metastases.[3]

Case Report

A 71-year-old male came to us with the history of hoarseness of voice, dysphagia and left sided facial palsy for 1 year. On clinical examination, there was a visible swelling along the left side of neck with signs of left 7th, 9th, 10th, 11th and 12th cranial nerves palsies like drooping of ipsilateral angle of mouth (CN VII), dysphagia and absent gag reflux (CN IX), hoarseness of voice (CN X), weakness of trapezius and sterno-cleidomastoid muscles (CN XI) and deviation of tongue towards left side (CN XII). Subsequently, contrast enhanced CT scan was performed which

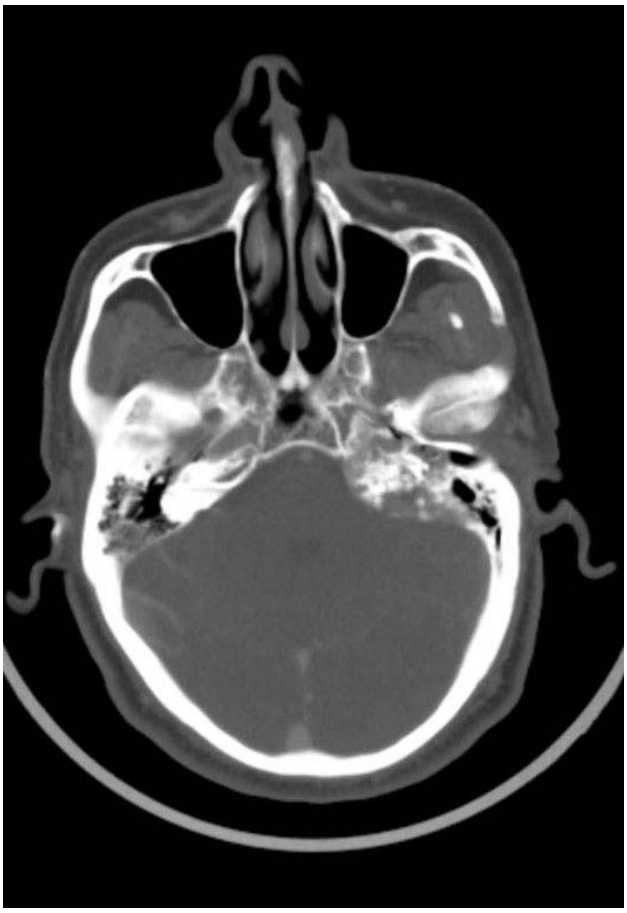

Figure 1a. CT scan axial view (bone window) showing destructive lesion involving the left mastoid air cells and petrous temporal bone.

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revealed a large soft tissue density mass causing destruction of the left petrous temporal bone, jugular foramen and facial nerve canal and is extending into the middle ear. Inferiorly the mass extends up to the left hypoglossal canal (Fig.1a,b,c). Later, contrast enhanced MRI was done which demonstrates the involvement of the left facial and vestibulocochlear nerves (Fig. 2a) and left glossopharyngeal and vagus nerves (Fig. 2b). Note the avidly enhancing tumor is also encasing the left internal carotid (Fig. 2c,d). On

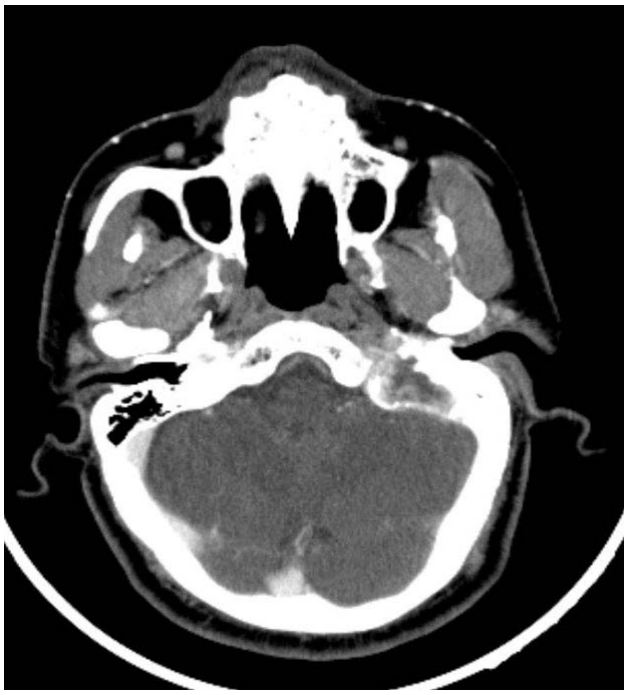

Figure 1b: Contrast enhanced CT scan axial view showing soft tissue mass in the left petrous bone with extension into the hypoglossal canal.

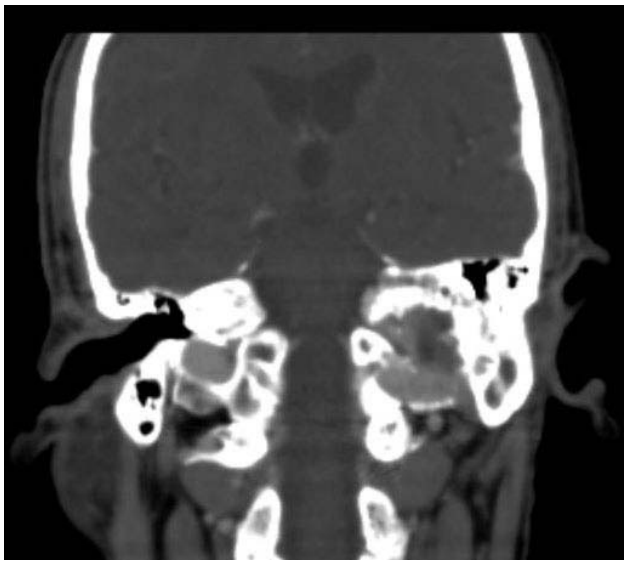

Figure 1c: CT scan coronal view (bone window) showing large soft tissue density mass involving the left jugular foramen, mastoid and petrous parts of temporal bone and extending inferiorly up to the hypoglossal canal.

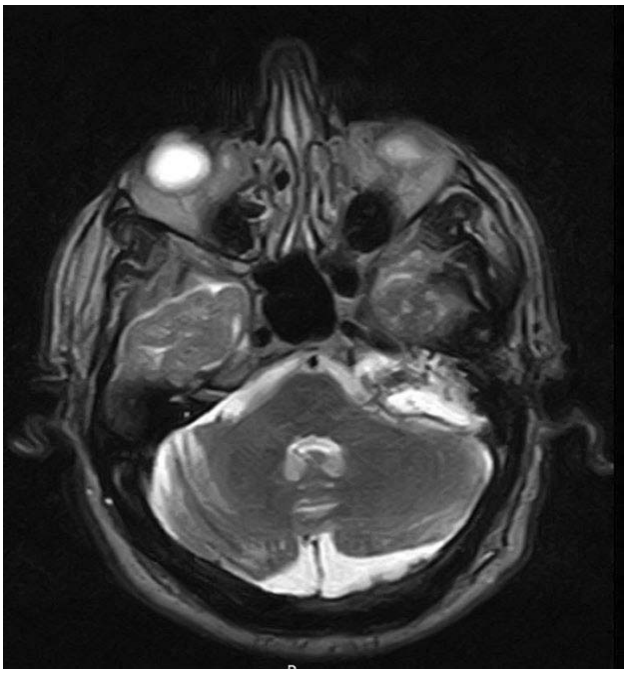

Figure 2a: MRI brain axial T2WI showing heterogeneous mass involving the left CN 7 and 8

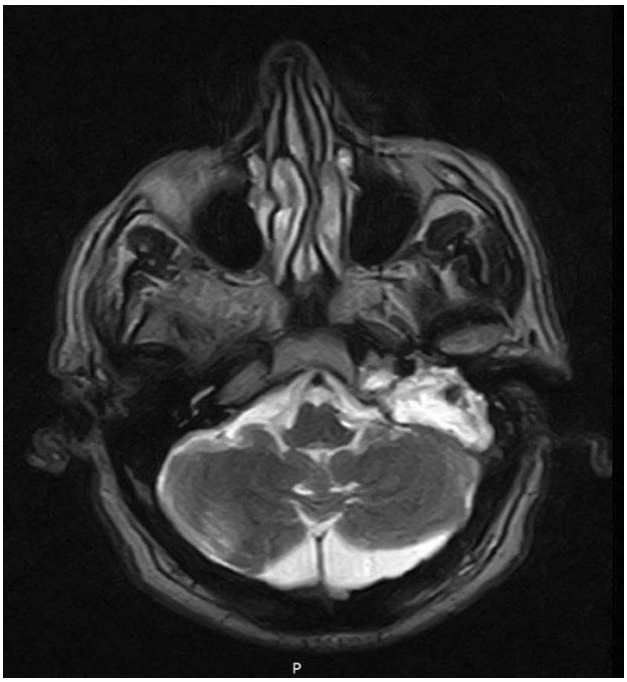

Figure 2b: MRI brain axial T2WI showing heterogeneous mass involving the left CN 9 and 10. Note the encasement of the internal carotid artery.

the basis of these clinical and radiological findings diagnosis of Collet-Sicard syndrome secondary to the left sided glomus jugulare paraganglioma was made.

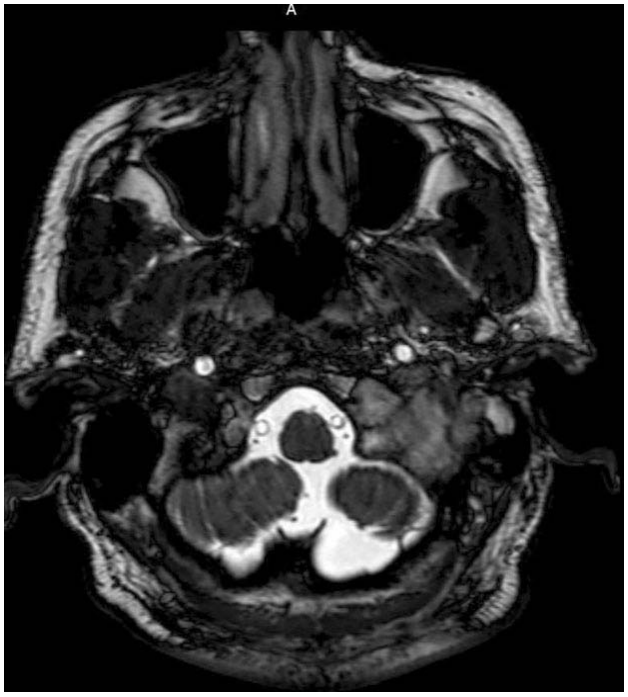

Figure 2c: MRI axial FIESTA showing heterogeneous mass involving the left CN 12

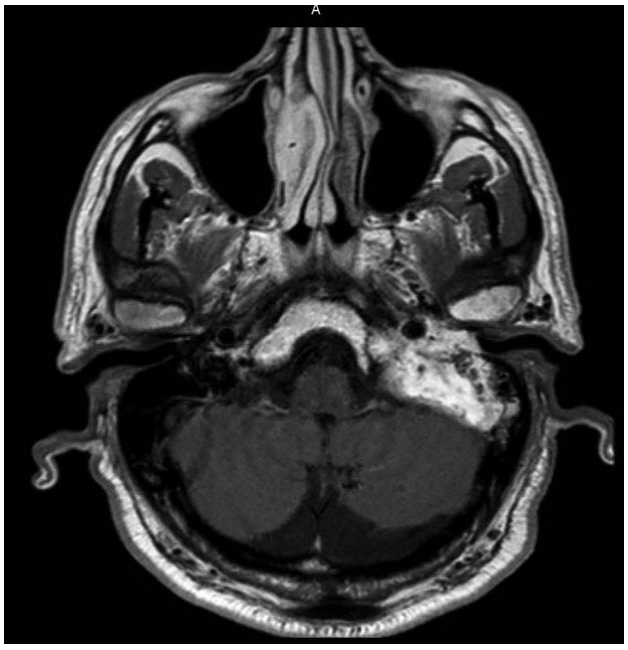

Figure 2d: MRI axial T1 post contrast showing avidly enhancing tumor with extension into the left hypoglossal canal and encasement of the left internal carotid artery.

Discussion

The CSS was first described in 1915, by Frederic Collet who reported this rare entity secondary to the gunshot injury to the world war I soldier with subsequent new additions byJean A. Sicard.[1,4] CSS presents with signs and symptoms of CN 9, 10, 11 and 12 palsies like displacement of palate, dysphonia, vocal cord paralysis, dysphagia, atrophy and paresis of tongue muscles and weakness of sternocleidomastoid and trapezius muscles.[2]

Paragangliomas also called glomus tumors are uncommon tumors which originates from paraganglia and account for <1% of head and neck tumors.[3] These tumors are named according to the site of their origin; like, glomus tympanicum are those tumors arising from the middle ear and mastoid; whereas, glomus jugulare are those originating from jugular foramen.[5] Paragangliomas most commonly occur in 5th to 7th decade with significant female predominance.[6] Glomus jugulare paragangliomas causes destruction in the region of jugular foramen, posterior inferior part of petrous temporal bone and then the mastoid and adjacent occipital bones[2] which leads to the involvement and subsequent paralysis of CN 9,10 and 11 resulting in Vernet syndrome (also called jugular foramen syndrome); with further extension of the tumor; there is also involvement of the CN 12 resulting in CSS.[7,8]

Radiologically, CT scan shows soft tissue density enhancing mass with erosion and destruction of the bone giving it moth eaten appearance. On MRI, it shows classical “salt and pepper” appearance; with salt being the blood products from hemorrhage or slow flow and pepper being the vascular flow voids. Intense blush is seen within the tumor on angiography.[2,9] Glomus jugulare tumors greater than 1.5 cm can also be imaged with Indium-111 labelled octreotide due to the presence of somatostatin receptors on these tumors.

Surgery is the treatment of choice; however, angioembolisation is typically performed a couple of days prior to the surgery.[9,10] Completely resected tumors have excellent prognosis; however, in irresectable or residual lesions radiotherapy can be performed.[2]

Conclusion

CSS is an uncommon entity with complex clinical presentations which can result in its delayed diagnosis. In the absence of trauma, neoplastic or metastatic disease of the temporal bone should be excluded in all the suspected cases of CSS.

Conflict of interest: None

Ethical review board: Ethical approval for this case report was given exemption from the institutional ethical review board.

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