

Cerebral venous thrombosis presenting as multiple lower cranial nerve palsies

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Abstract

Cerebral venous thrombosis (CVT) is a well-recognized entity, but its clinical presentation is varied and often mimics many neurological disorders, making it a diagnostic challenge. Cerebral venous thrombosis has a wide spectrum of signs and symptoms, which may evolve suddenly or over weeks. It mimics many neurological conditions such as meningitis, encephalopathy, idiopathic intracranial hypertension, and stroke. Cerebral venous thrombosis presenting as multiple lower cranial nerve palsies, are rarely reported. We describe a pregnant lady who presented with sensorineural deafness of the right ear and paralysis of the 9th, 10th, and 12th cranial nerves on the right side. She was diagnosed to have thrombosis of the right transverse sinus and sigmoid sinus with extension to the jugular vein and confluence of sinuses. She improved with anticoagulant treatment.

Keywords: Cerebral venous thrombosis, lower cranial nerve palsies, magnetic resonance imaging

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Introduction

The clinical picture of cerebral venous thrombosis is variable, and a wide spectrum of neurological symptoms has been described. Several cranial nerve syndromes in CVT have been identified and attributed to extension of thrombosis into contiguous venous tributaries, which leads to direct pressure and subsequent paralysis of the nerves lying in proximity to the clot.^[1] Lower cranial nerve palsies in CVT is very rare, and we describe here a patient with paralysis of the 8th, 9th, 10th, and 12th cranial nerve palsies due to CVT.

Case Report

A 34-year-old pregnant lady in last trimester presented with headache of two months duration and difficulty in speaking, swallowing, and tinnitus of two weeks

duration. Headache was mainly in the occipital region radiating to the retroauricular region and the back of the neck. It was initially associated with vomiting. Six weeks after the onset of headache, she developed slurring of speech, hoarseness, nasal twang of voice and difficulty in swallowing with nasal regurgitation. She had also difficulty in moving the tongue side-to-side and hearing with right ear.

On examination, the lady was pale, and her speech was dysarthric. The first seven cranial nerves were normal. Fundi were bilaterally normal. Sensorinural deafness was present on the right side. There was arching of the palate to the left side, and palatal movements were decreased on right side. Gag reflex was absent on the right side. The trapezius and the sternocleidomastoid muscles had normal strength. There was weakness of the tongue with deviation to the right side. Rest of the neurological examination was normal. ENT examination did not reveal any focus of infection.

Her routine blood examinations showed hemoglobin of 10 gms, total count of 7000 cells/cumm, and an ESR

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of 76 mm/hr. Her routine urine examination, liver, and renal function tests were normal. Anti-nuclear antibody and anti-phospholipid antibodies were negative. Serum homocystine and C-reactive protein values were normal. Coagulation profile including prothrombin time, activated partial thromboplastin time was normal. Her blood culture and sensitivity were negative. Pure tone audiometry showed moderate sensorineural deafness of right ear. Magnetic Resonance Imaging (MRI) of brain with Magnetic Resonance Venogram was done and showed a thrombus in the right internal jugular vein and sigmoid sinus [Figures 1 and 2]. The thrombus was extending to right transverse sinus and confluence of sinuses [Figure 3]. The patient was treated with heparin in the hospital for a total of 4 week and subsequently switched over to oral anticoagulants. She had an uneventful twin delivery. Her neurologic signs and symptoms slowly improved over one week. At the time of discharge, she had residual sensorineural deafness.

Discussion

Thrombosis of the cerebral venous sinuses is characterized by the classical features of headache, seizures, and various focal neurological deficits including cranial nerve palsies. There are only few reported cases of cerebral venous thrombosis affecting the 8th cranial nerve. Kim *et al.* has described cerebral venous thrombosis mimicking acute unilateral vestibulopathy and has stressed the importance of recognizing this entity while evaluating a patient with headache and vertigo.^[2] Fonseca *et al.* has reported reversible bilateral sensorineural hearing loss in a woman with cerebral venous thrombosis whose hearing loss reversed with treatment of intracranial hypertension.^[3] There are different explanation for VIII nerve dysfunction in CVT. Increased pressure in the subarachnoid space may be transmitted to the endolymphatic system via cochlear aqueduct and the perilymph. This results in increased fluid pressure in the internal ear, causing cochlear dysfunction. Increased intracranial pressure may also disturb VIII nerve function by causing increased pressure in its meningeal sheath.^[3] However, CVT has also been referred as a direct cause of hypoacusis, in the absence of increased intracranial pressure. Cochlear venous blood is collected by cochlear vein and drains through labyrinthine vein into the inferior petrosal sinus or directly into transverse sinus. Thrombosis of transverse sinus or internal jugular vein can produce a venous overload and congestion of inner ear structures. Damage is due to ischemia secondary to rise in cochlear pressure, resulting from back-pressure effect of extension of thrombus to the cochlear or labyrinthine veins.^[2]

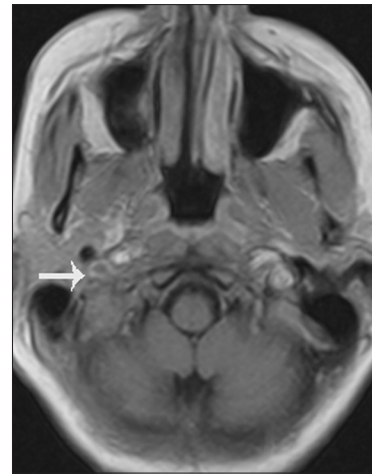


Figure 1: Axial T1-weighted image of base of skull shows focal thrombus within the most superior part of the right internal jugular vein (white arrow)

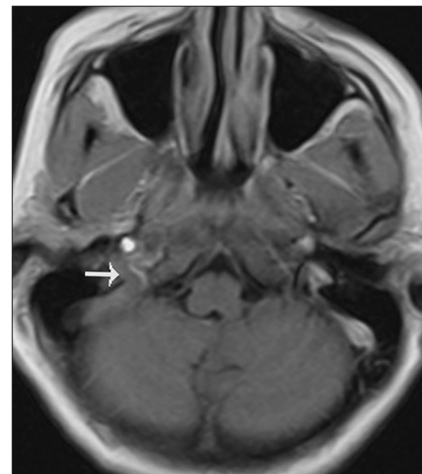


Figure 2: Axial T1-weighted contrast image shows thrombus in the right sigmoid sinus and internal jugular (white arrow)



Figure 3: MR venography shows a decreased signal in the right transverse sinus and absent signal in the right sigmoid sinus and internal jugular vein (white arrows)

Jugular foramen courses anterolaterally as it exits the skull base, and usually the right jugular foramen is larger than the left in two third of the cases. This is presumably related with dominance of right cerebral sinus drainage. Each foramen is separated into an anteromedial portion (the pars nervosa) and a postolateral portion (pars vascularis) by a 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.^[4] Involvement of jugular foramen region producing lower cranial nerve deficit is characterized by several eponymous syndromes like Vernet's syndrome, Collet Sicard syndrome, Villaret syndrome, Avellis's syndrome, Schmidt's syndrome, Tapia's syndrome, and Jackson's syndrome.

Collet Sicard Syndrome (CSS) named after Frederic Collet and Jean Sicard refers to ipsilateral dysfunction of cranial nerves 9-12 due to the pathological process located near the jugular foramen. Usual causes are trauma, tumor, infections, and malignant infiltration. The current literature has only three reported cases of CSS as a result of venous thrombosis. Tom *et al.* has reported a case of Collet Sicard Syndrome with initial sparing of the accessory nerve due to thrombosis of the sigmoid jugular complex.^[5] A possible explanation for the initial sparing of the 11th nerve is that it might have been due to the duality of innervation of these muscles supplied by the spinal branch of CN XI and C2-4 cervical spinal branches.^[6] Our case differs from all these, in that our patient had 8th nerve involvement in addition to 9th, 10th, and 12th cranial nerves. Such a combination of cranial nerve palsies in cerebral venous thrombosis has not been reported so far.

While investigating the cause of lower cranial nerve palsies, one must consider causes like trauma, malignancies, infections, and thrombotic states. MRI imaging of brain with an MR venogram is the most sensitive examination in detecting cerebral venous thrombosis. Pregnancy is an acquired prothrombotic condition known to produce CVT. In our case, a search was made for a focus of infection and other inflammatory and hematological conditions known to produce CVT, which was negative.

In the management of CVT during pregnancy, in addition to supportive care, seizure control and measures to lower intracranial pressure, selection of anticoagulant

is important as oral anticoagulants are associated with fetal embryopathy and bleeding in the fetus. Recent recommendation favors the use of full-dose low molecular weight heparin (LMWH) over unfractionated heparin (UFH), both in the acute phase and subsequent prophylaxis of CVT during pregnancy.^[7] LMWH or a vitamin K antagonist should be continued for at least 6 weeks postpartum (for a total minimum duration of therapy of 6 months) with a target INR of 2.0 to 3.0. CVT is not a contraindication for future pregnancies. Considering the additional risk that pregnancy confers to women with a history of CVT, prophylaxis with LMWH during future pregnancies and the postpartum period can be beneficial.^[7]

Conclusion

When a patient presents with multiple cranial nerve palsies in the setting of a hypercoagulable state like pregnancy, cerebral venous thrombosis has to be kept as an important differential diagnosis. Imaging plays a key role in diagnosing CVT. Successful management mandates anticipation, prompt recognition, and an optimal treatment of CVT.

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