

Lower motor neuron facial palsy in cerebral venous sinus thrombosis

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ABSTRACT

With advances in the neuro-imaging modalities, diverse manifestations of the cerebral venous sinus thrombosis (CVT) are being recognized. There are very few reports of isolated cranial nerve palsies in CVT. In this case report, we describe a patient of lower motor neuron facial palsy with CVT who was successfully treated with anticoagulation, highlighting the atypical manifestation of the disease.

Key words: Anticoagulation, cranial nerve diseases, cerebral venous sinus thrombosis, facial paralysis, magnetic resonance venography, sinus thrombosis

Introduction

Before the advent of the imaging studies, cerebral venous sinus thrombosis (CVT) was thought to be a rare and severe disease characterized by headache, papilledema, seizures, focal deficits, progressive coma and death, mostly diagnosed during the autopsy. However with advent of magnetic resonance imaging (MRI) and venography (MRV) there is recent surge in literature describing various less recognized manifestations of the CVT. It is more common than previously assumed, the spectrum of its clinical presentation is extremely wide and outcome with anticoagulation is usually favorable. Because of its diverse etiology, manifestations and unpredictable course, it remains a challenge for the clinicians. In this report we describe a patient with CVT presenting with unilateral lower motor neuron (LMN) facial palsy.

Case Report

A 30-year-old lady congenitally deaf and mute presented with history of headache since one month and right

LMN facial palsy since last 20 days. Using the sign language, she indicated that at the onset headache was predominantly over the anterior part of head and within few days it became diffuse. It was pressing in character, present throughout the day and associated with vomiting on increased severity. Ten days after the onset of headache she noticed difficulty in closing her right eyelid and three days later deviation of angle of mouth to left. There was no history of fever, ear discharge, weakness of limbs, seizures, diplopia, blurring of vision, ataxia, tinnitus or recent worsening of deafness. Physical examination showed presence of pallor, no organomegaly, external ear, mastoid and eardrum were normal. She was conscious, alert and oriented. Her visual acuity was 6/9 on both sides. Examination of the optic fundi revealed presence of bilateral papilledema. Eye movements and pupillary reactions were normal. She had right LMN facial palsy with absent taste and tearing ipsilaterally with bilateral sensory neural deafness as evidenced by tuning fork tests. Rest of the cranial nerves, motor, sensory and cerebellar examinations were normal. Clinically she was diagnosed as a case of raised intracranial pressure (ICP) with right LMN facial palsy with congenital deaf mutism.

Investigations revealed hemoglobin of 7.1 gm%, microcytic hypochromic blood picture with normal white blood cells and platelets. Serum biochemistry and electrolytes were normal. Rheumatoid factor and antinuclear antibodies were negative. Serum antiphospholipid antibodies, homocysteine levels, angiotensin converting enzyme levels and chest X-ray were normal. Computerized

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tomography (CT) scan of the brain showed posterior empty delta sign in the superior sagittal sinus. Temporal bone and middle ear cavity were normal. MRI and MRV [Figure 1] showed thrombosis of superior and right lateral sinus, with no parenchymal lesions. Lumbar puncture was normal except with raised opening pressure. Audiometry showed bilateral profound sensory neural hearing loss. Brain stem auditory evoked responses were absent bilaterally. She was diagnosed as a case of CVT (superior and right lateral sinus) with ipsilateral LMN facial palsy with congenital deaf-mutism. She was started on parenteral followed by oral anticoagulation (prothrombin time, international normalization ratio maintained at about two), hematinics, anti edema measures. Over the next one month her symptoms of headache and facial palsy improved and at 4 months after her symptom onset, she was asymptomatic.

Discussion

Our patient presented with features of raised ICP with unilateral LMN facial palsy localizing to suprageniculate part (taste and tearing abnormalities) of the nerve. There were no seizures, altered sensorium or other focal deficits for the clinical suspicion of the CVT. Imaging with CT, MRI and MRV showed superior sagittal and right lateral sinus thrombosis. The temporal profile of the facial palsy, localization of nerve involvement and absence findings associated with other causes of LMN facial palsy (clinically and investigation wise) suggested that facial palsy was probably secondary to the CVT. Her congenital deaf mutism is unrelated to her present illness.

The most common form of facial paralysis is idiopathic (Bell's palsy), which has an annual incidence

of 23-25 patients per 100,000 populations.^[1,2] Bousser and Barnett have separated five groups of clinical presentations of CVT: Isolated intracranial hypertension, focal cerebral signs, cavernous sinus thrombosis, subacute encephalopathy and unusual presentations (such as cranial nerve palsies).^[3] CT scan findings in CVT are described as direct signs (cord sign, dense triangle sign, delta or empty triangle sign) and indirect signs (contrast enhancement of the falx and tentorium, white matter hypodensities, hemorrhagic infarcts.). MRI and MRV offer major advantage in the diagnosis, being noninvasive and depicting parenchymal and venous sinus pathology better.^[3] With the advent of these modalities diverse manifestations of CVT have been recognized. Apart from the few anecdotal case reports, cranial nerve symptoms alone have not been reported to be caused by CVT.^[4]

Cranial nerve palsy as an isolated manifestation of CVT has been attributed to the elevated intracranial pressure, extension of thrombosis to venous channels, or direct pressure from the clot itself.^[5,6] Kuehnen *et al.* have reported 5 patients who were initially evaluated for etiologies of single/multiple cranial nerve palsies finally turning out to be the cases of thrombosis of the ipsilateral transverse and sigmoid sinus on evaluation.^[4] Lateral sinus (transverse plus sigmoid portion) drains blood from the cerebellum, brainstem and posterior portions of the cerebral hemispheres, veins from cranial nerves in the posterior fossa, the middle ear and diploic veins. According to Kuehnen *et al.* thrombosis of the lateral sinus can produce venous congestion and dilatation of the cranial nerve veins.^[4] This causes reversible compromised oxygen or glucose consumption within the cranial nerve tissue due to edema and backpressure, and due to this cranial nerve palsies will develop.

Straub J *et al.* have described a 17-year-old lady with LMN facial palsy with ipsilateral transverse sinus thrombosis.^[6] They evaluated the patient with transcranial magnetic stimulation and concluded that the facial palsy was due to the transient neuropraxia in the intracranial segment of the nerve. They explained the neuropraxia due to the leakage of the fluids and ions into the endoneurial space of the nerve due to elevated venous transmural pressure in the nerve's satellite vein, which ultimately drains to ipsilateral transverse sinus. This causes impairment of the saltatory current flow, with reversible slowing of the conduction or even conduction block. Our patient presented with thrombosis of the superior and right lateral sinus with no parenchymal lesions with ipsilateral LMN facial palsy, which may be explained by either or both of the above mechanisms. We could not do transmagnetic stimulation studies.



Figure 1: Sagittal view of the magnetic resonance venography of the brain showing superior sagittal sinus (white straight arrow) and right lateral sinus (black curved arrow) thrombosis

Conclusion

Our patient presented with the features of right LMN facial palsy with raised ICP and on subsequent imaging studies, we made the diagnosis of CVT. Because of the site of involvement of the facial nerve, temporal course of the facial weakness and the absence of other causes of facial palsy, we suggest it is probably related to the CVT. The present case highlights the atypical presentation of CVT, the role of imaging in the diagnosis of CVT and looks in to the literature regarding the pathogenesis of the nerve dysfunction.

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