Opalski Syndrome

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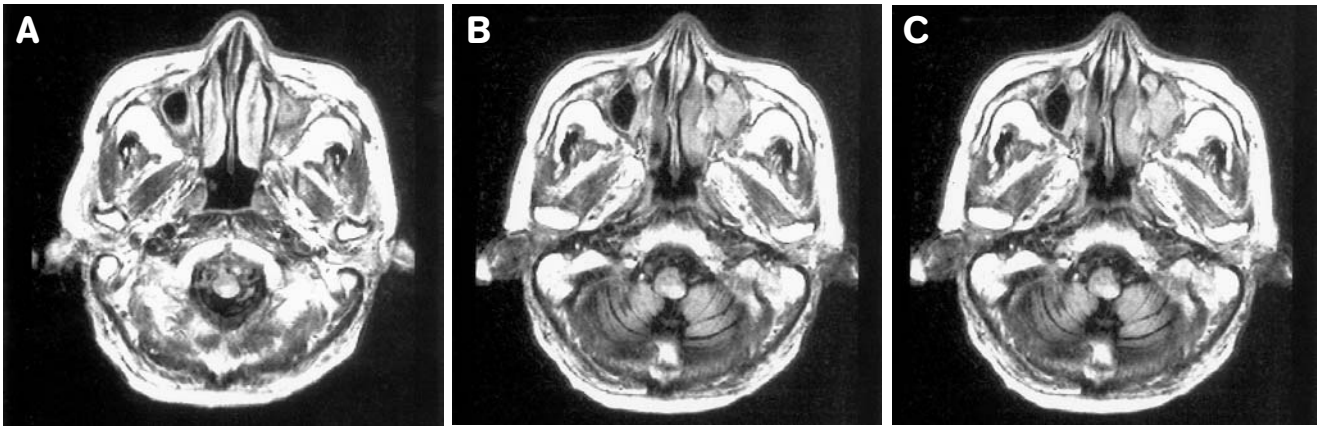

Figure 1. Axial FLAIR (fluid-attenuated inversion recovery) demonstrates hyperintensity lesion at left upper cervical (A) and left posterolateral lower medulla (B). Diffusion-weighted imaging shows hyperintensity, favors acute infarction (C).

A 76-year-old male heavy smoker with diabetes mellitus and hypertension for more than 10 years suffered from sudden onset of left limbs weakness and numbness, which were associated with dizziness, easy chocking, dysarthria, hiccup, constipation and urinary retention. He was brought to an outside hospital at first and transferred to our hospital two days later. There was no recent neck trauma or chiropractic treatment. He could not raise his left limbs initially. When the limb weakness was improved later, wild movements on attempted motion followed. They consisted of flinging, violent, jerky components and would hit himself or adjacent objects inevitably. Neurological examinations on admission revealed left Horner's sign, impaired left gag reflex, decreased pin-prick and thermal sensation on left face and right limbs, hyperesthesia on left limbs and trunk, mild left hemiparesis with severe dysmetria and truncal ataxia. The joint position sensation was impaired on both legs and left upper limb.

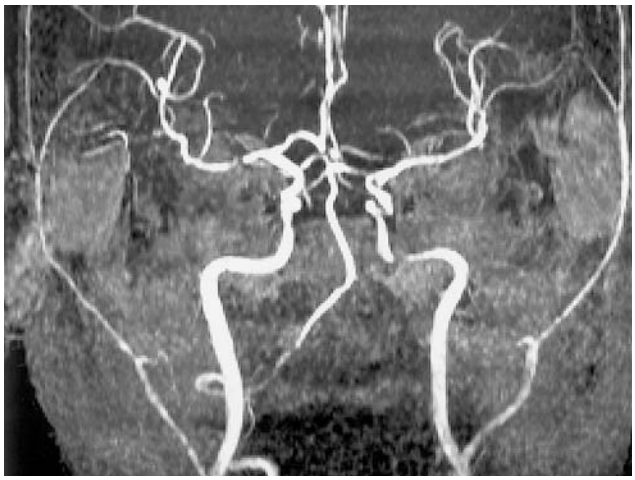

Figure 2. MR angiography reveals severe stenosis of left vertebral artery.

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Brain MRI showed acute infarction at left posterolateral aspect of lower medulla oblongata and contiguous upper cervical cord (Fig. 1). MR angiography showed severe stenosis of left intracranial vertebral artery (Fig. 2). Color Doppler scan disclosed high resistant flow in left cervical vertebral artery with reversed flow in left intracranial vertebral artery. Transthoracic echocardiography showed left ventricular hypertrophy without thrombus in the atrium and ventricle.

Opalski syndrome is relatively rare and easily be missed. It is characterized by lateral medullary syndromes with ipsilateral hemiplegia that was first described by Opalski in 1946[1]. The lesion is located lower than that of Wallenberg syndrome and involves the corticospinal fibers caudal to the pyramidal decussation. Only one case of Opalski syndrome with MRI findings was reported[2].

One interesting finding was the occurrence of wild motion of the proximal ipsilateral arm in this patient. On attempted movement, the arm would fling itself over the head in a grossly ataxic jerky fashion mimicking hemiballism. However, it disappeared completely at rest. Currier et al. reported 5 cases of Wallenberg syndrome with similar pictures, one of them received autopsy[3]. It was a cerebellar sign but more violent than usual. It was suggested that the slightly more caudal location involves the cuneate nuclei, the crossing fibers of the sensory decussation, and the spinocerebellar pathways and results in this special depiction which probably has localizing value[3].

Although this patient complained of left limbs numbness, loss of pain sensation was found on the right limbs and left limbs showed hyperesthesia. Similar picture was noted by Kim JS[4], who studied 174 patients with Wallenberg syndrome and found 12 (6.7%) had ipsilateral symptoms in the limbs/body, in addition to typical lateral medullary syndrome. Those sensory symptoms were generally described as numbness or tightness, predominantly affecting the upper extremities, especially distal fingers. In addition, those patients were more frequently associated with ipsilateral hemiparesis and their lesions were usually located at more caudal part of the medulla extending dorsomedially. The involvement of ipsilateral dorsal column or decussating lemniscal fibers was suspected.

Opalski syndrome is a variant of Wallenberg syndrome, and is differentiated by a concomitant ipsilateral limbs weakness. Conventional MRI and MR angiography are the first choice to visualize the lesion of acute infarction caused by ipsilateral vertebral artery occlusion.

References:

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