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Rare Variant of Lateral Medullary Syndrome; Opalski Syndrome with Cerebellar Infarction

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Abstract

Presentation

A 36-year-old female presented with sudden onset neck pain, vertigo, nausea, facial numbness and gait disturbance. Classic features of lateral medullary syndrome (LMS) were elicited; Right Horner's, reduced right-sided facial pain and temperature sensation, left upper and lower limb numbness, dysphonia, dysarthria and dysphagia. Additionally, there was right hemi-paresis.

Diagnosis

CT angiogram showed a poorly enhancing right vertebral artery and MRI showed oedema of the right postero-lateral medulla and inferior cerebellar peduncle consistent with vertebral artery dissection and acute infarction.

Treatment

She was admitted to the stroke observation unit. We commenced high dose aspirin in combination with multidisciplinary team input, risk factor modification and supportive therapies.

Conclusion

This was Opalski syndrome, caused by right vertebral artery dissection extending caudally from the posterior inferior cerebellar artery (PICA). This resulted in ischemia of the lateral medulla, inferior cerebellar peduncle, and corticospinal fibres after the pyramidal decussation, resulting in Wallenberg's syndrome, cerebellar ataxia and ipsilateral hemiparesis.

Introduction

Lateral medullary syndrome (Wallenberg syndrome) is a well-known neurological disorder caused by ischemia of the vertebrobasilar vascular system. The implicated lateral medullary structures include the nucleus ambiguous, the spinal trigeminal nucleus, cranial trigeminal tract, cerebellum/inferior cerebellar peduncle, hypothalamo-spinal fibres and vestibular nuclei. Classically it is manifested by vertigo, diplopia, dysarthria, Horner's syndrome, numbness (ipsilateral face and contralateral limb), and no limb weakness. Opalski syndrome, a rare variant of LMS, is associated with ipsilateral hemiparesis due to the involvement of corticospinal fibers caudal to the pyramidal decussation. Babinski-Nageotte syndrome is another variant which has contralateral hemiparesis because pyramidal tract is affected before decussation.

Case Report

A 36-year-old female, parity 2 gravidity 2, presented with a sudden onset of vertigo, visual disturbance, nausea, gait disturbance, and right facial discomfort. She reported recurrent attacks of right sided neck pain worse when moving head to the left side for few days. On examination, her blood pressure was 168/95, her pulse was 92 beats per minute, and her cardiovascular and respiratory examination were normal. Neurological exam revealed right Horner's syndrome, Nystagmus was noted on right lateral gaze with rapid ocular movement to the right, right-sided ataxia, reduced pinprick and temperature sensations on right side of her face and her left side of body, dysphonia and decreased gag reflex. In addition, there was an evidence of right (ipsilateral) hypotonia and hyporeflexia with hemi-sensory loss. CT angiogram showed a poorly enhancing, small calibre right vertebral artery [Figure 1].

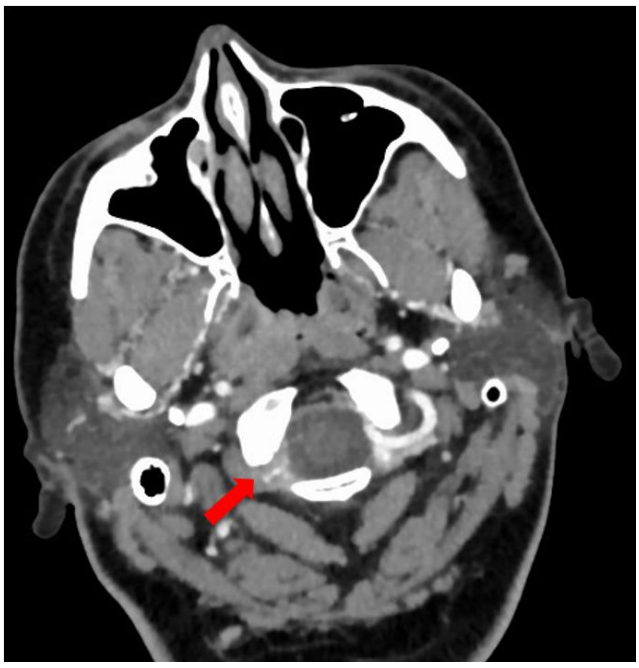

Figure 1. Asymmetrical vertebral arteries, with a small calibre right vertebral artery which is poorly enhancing. This is in-keeping with right vertebral artery dissection.

Diffusion-weighted and T2-weighted brain MRI revealed Oedema of the right posterior lateral medulla involving inferior cerebellar peduncle, in-keeping with infarction [Figure 2].

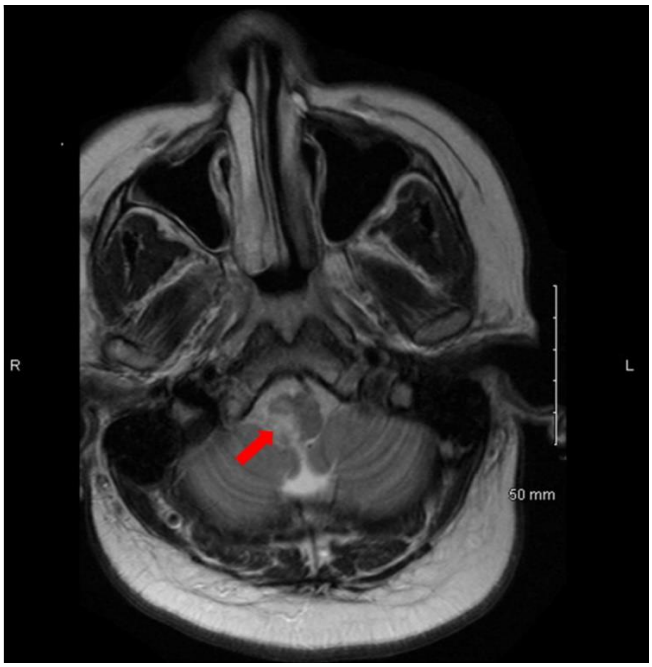

Figure 2. Oedema of the right posterior lateral medulla involving inferior cerebellar peduncle, in-keeping with infarction.

She was admitted to the stroke unit and treated with high dose aspirin. She underwent rehabilitation by the multidisciplinary team with medical risk factor modification. After 10 days, she was discharged to a rehabilitation hospital for further therapy, at this point her right sided power and tone had returned, and she had developed right sided hyper-reflexia with upgoing Babinski response.

Discussion

Opalski syndrome was described by Adam Opalski in 1949, who reported two patients with mild hemiperesis, with hyperreflexia and Babinski's sign on the same side, along with features of LMS.[1] He believed the weakness resulted from involvement of corticospinal tracts extending caudally to the pyramidal decussation. Dhamoon et al. later reported a case with the lateral medullary syndrome and severe ipsilateral weakness following vertebral artery occlusion.[2] Subsequently he reported an autopsy case describing this as a perfusion failure due to severe atherosclerosis and thrombosis in the proximal and distal right vertebral arteries. In our patient, CT angiography revealed severe stenosis of the proximal and distal parts of the right vertebral artery so we may say it was the extensive vertebral artery dissection extending caudally to involve the inferior cerebellar peduncle and the corticospinal fibres after the pyramidal dissection.

Prognosis of vertebral artery dissection depends mainly on the severity of the resulting stroke syndrome. Management comprises antiplatelet agents and anticoagulants; evidence demonstrates equal results between these two modalities.[3] Endovascular and surgical treatments are reserved for patients with concomitant complications while intra-arterial thrombolytics in acute ischemic events presenting within 4.5hrs of symptoms have been utilized safetly.[4]

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