Case report - Opalski’s syndrome A rare variant of lateral medullary syndrome in TAKAYASUS ARTERITIS

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Abstract:
we present an interesting case of Opalski syndrome which is an unusual manifestation of Lateral Medullary Syndrome in addition this patient also has features suggestive of Takayasus arteritis. A 42 year female, known diabetic presented with left upper limb claudication and recurrent transient ischemic attacks for the past 3 months, with recent sudden onset of left hemiparesis, left 9th 10th cranial nerve palsy, sensory loss in right half of the body and left half of the face with left cerebellar signs. She had absent pulse in left upper limb and significant BP variation in both upper limbs (BP-right upper limb 11070 mm of Hg, left upper limb not recordable) normal pulse and BP in the lower limbs showing some of the clinical features suggestive of Takayasus arteritis. Her CT scan brain showed left cerebellar infarct, MRI brain revealed acute infarct in left medulla, with rostral spinal cord extension, cerebellar vermis, right occipital lobe, right thalamus, left thalamus, CT was normal, CT Angiogram showed left Subclavian artery occlusion at its origin from the aorta, with faint reformation of distal axillary and brachial artery from the lateral thoracic artery. Patient underwent medical management and improved well and regular follows up.

Keyword: Takayasu’s arteritis, Lateral medullary syndrome, Opalski syndrome, CT angiogram

Introduction:
Wallenberg described the lateral medullary syndrome (LMS), the most common form of brainstem stroke. Wallenberg’s original patient had an occlusion of the posterior inferior cerebellar artery (PICA), but LMS is most often due to ischemia in the PICA distribution because of vertebral artery occlusion (Figure 1).
Figure-1: Cross section of the medulla illustrating the site of the lesion following thrombosis of the posterior inferior cerebellar artery.

Typical manifestations include vertigo, nausea, vomiting, nystagmus, hoarseness, dysphagia, dysphonia, singultus, ipsilateral hemiataxia, numbness of the ipsilateral face and contralateral body. Occipital headache or pain in the back of the neck may occur at the onset; prominent pain raises the possibility of vertebral artery dissection. Lateral Medullary Syndrome is often found in patients with vertebrobasilar vascular lesions. On rare occasions, the syndrome is associated with ipsilateral hemiparesis which is known as Opalski syndrome. Ipsilateral hemiparesis is due to inferior extension of the infarct to the lateral funiculus of the rostral spinal cord or may be contralateral hemiparesis due to inferior extension of the zone of ischemia to the medullary pyramid. An unusual manifestation of Lateral medullary syndrome is like an ipsilateral upper motor neuron facial palsy due to the involvement of Dejerine's aberrant pyramidal tract. LMS is usually ischemic; it has also been described with aneurysm, vasculitis, abscess, hematoma, arteriovenous malformation, demyelinating disease, and metastatic neoplasm. Takayasu’s Arteritis (TA) also known as pulseless disease or non-specific aorta arteritis is a form of large vessel granulomatous vasculitis of unknown cause that chiefly affects the aorta and its major branches, left common carotid, brachiocephalic and left Subclavian artery. Young women are more frequently affected. It can present as pulseless upper extremities (arm, wrist, hand), arm claudication. This is because the granulomatous inflammation leads to arterial stenosis, thrombosis, and aneurysm. these changes are seen in arterial studies such as MRA, CT angiogram, and digital subtraction angiography as occlusion or narrowing of the vessels. Stroke is a common complication and often the first manifestation of T.A, with an incidence of 10-20%.

Case report:

A 42 year female, known diabetic admitted with previous history of left upper limb claudication and recurrent transient ischemic attacks involving vertebrobasilar territory for the past 3 months and now presented with weakness of left upper & lower limb with bulbar symptoms. On examination she had left sided hemiparesis, left 9th 10th cranial nerve palsy, left Cerebellar signs, Horner’s syndrome, diminished pain and temperature on right half of the body and left half of the face, the other sensations being normal. She had absent pulse in left upper limb and significant BP variation in both upper limbs (BP-right upper limb; 110/70 mm of Hg, left upper limb not recordable). On investigation basic blood reports revealed raised ESR, vasculitic workup, chest X-ray PA view, carotid Doppler study were normal.HIV, VDRL were non-reactive. ECHO showed diastolic dysfunction.MRI Brain showed acute infarct involving the left half of medulla, cerebellar vermis, right occipital lobe right thalamus and rostral spinal cord (Figure 2 & 3).
Figure -2: T2W MRI showing hyperintense lesions in cervical cord, left medulla and left cerebellum.

Figure -3: MRI FLAIR showing hyperintense lesion in cervical cord, right occipital lobe and right thalamus.

CT Angiogram showed occlusion of left Subclavian artery at the origin with faint reforma tion of distal axillary and brachial artery from the lateral thoracic artery (Figure-4).

Figure-4: CT Angiogram showing occlusion of left Subclavian artery at the origin with faint reformation of distal axillary and brachial artery from the lateral thoracic artery.

Patient underwent medical management with aspirin, atrovastatin, corticosteroids and improved well on regular follow-up.

Discussion:
An unusual Manifestation of Lateral Medullary syndrome is

1 Ipsilateral hemiparesis\textsuperscript{2, 3, 4, 5} due to inferior extension of the infarct to the lateral funiculus of the rostral spinal cord is known as Opalski syndrome or may be contra lateral hemi paresis due to inferior extension of the zone of ischemia to the medullary pyramid.

2 Ipsilateral upper motor neuron facial palsy due to involvement of Dejerine's aberrant pyramidal tract.

3 Ocular motor abnormalities\textsuperscript{6} are common, including skew deviation with ipsilateral hypotropia, ocular tilt reaction, and bizarre environmental tilt illusions, including world inversion (floor on ceiling phenomenon), ipsilateral gaze deviation with impaired contralateral pursuit, saccadic abnormalities, seesaw nystagmus, and eyelid nystagmus. Ocular abnormalities and facial weakness are common and do not imply extension of the lesion beyond the lateral medulla.

4 There is impaired sensation\textsuperscript{7} of the ipsilateral arm and leg due to inferior extension to the gracile and cuneate nuclei.

5 Other unusual manifestations include ipsilateral loss of taste, or contra lateral facial hypalgesia mild unilateral, proximal arm ataxia\textsuperscript{8}, neurotrophic ulceration of the face; inability to sneeze; and weakness of the sternocleidomastoid and which are reported in literature.
6 Takayasu’s arteritis (TA) is a chronic, idiopathic, inflammatory disease primarily affecting aorta and its branches. It mainly affects young females in the age group of 10-30 years.

7 TA has been described worldwide; it occurs most commonly in Japan, China, India, and Southeast Asia; they presented as various system involvements including vascular, central nervous system, musculoskeletal and, cardiac system.

American College of Rheumatology Classification Criteria for Takayasu’s

1 Onset before age 40 yr
2 Limb claudication
3 Decreased brachial artery pulse
4 Unequal arm blood pressure (>10 mm Hg)
5 Subclavian or aortic Bruit
6 Angiographic evidence of narrowing or occlusion of aorta or its primary branches, or large limarteritis

*The presence of three or more of the six criteria is sensitive (91%) and specific (98%) for the diagnosis of Takayasu’s arteritis. Twenty percent of TA patients have self-limited disease. The rest have relapsing-remitting course or requiring chronic corticosteroid or immunosuppressives and regular follow up is needed in this type of patients.

Conclusion:
This middle aged female diagnosed to have Takayasu’s arteritis based on clinical and radiological investigation, developed Opalski syndrome which is a rare variant of lateral medullary syndrome.

TA should be suspected in young female who present with stroke, as this is fairly responsive to medical management such as corticosteroid or immunosuppressives and regular follow up is needed in this type of patients.

Reference:
1 Gil Polo, A. Castrillo Sanz Opalski syndrome: A variant of lateral-medullary syndrome. j.nrleng.2012; 02:006-8


4 Dhamoon SK, Iqbal J, Collins GH. Ipsilateral hemiplegia and the Wallenberg syndrome. Arch Neurol, 1984; 41:179-180

5 Shinichiro Nakamura, MD. Opalski syndrome Ipsilateral hemiplegia due to a lateral-medullary infarction Neuroimages Neurology, 2010; 75:1658 -1663


7 Kim JS. Sensory symptoms in ipsilateral limbs/body due to lateral medullary infarction. Neurology 2001; 57:1230-1234

9 Syed Ahmed Zaki, Unusual presentation of Takayasu's arteritis as posterior reversible encephalopathy syndrome Ann Indian Acad Neurol 2011; 14:214-6