Acute rare Presentation of Dural Arteriovenous Malformation (AVF) in young patient

MIK Omer, MBBS, MRCP(UK), Internal Medicine Resident in King Abdullah Bin Abdulaziz University Hospital- Princess Nora Bin Abdurrahman University

Background:

Dural AVF represent 70 % of all spinal cord vascular malformations. The most common clinical presentation is progressive, step-wise myeloradiculopathy, resulting in ascending symmetrical weakness and sensory changes. Minority of patients can present with rapidly progressive myelopathy due to venous thrombosis. Symptoms may initially fluctuate, but eventually a permanent and progressive paraparesis with sensory disturbances and sphincter dysfunction occurs. [1]. Quick accurate diagnosis and proper management can be helpful in stabilizing, even ameliorating, the neurologic deficits [2,3].

Case Summary:

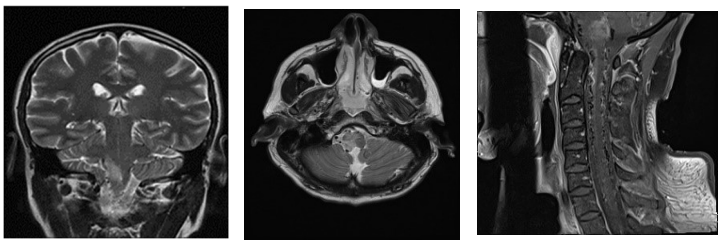
30-year-old male, medically free, but he was suffering from recurrent severe headache for 2 months prior to admission. He presented with 10-day history of dizziness & vomiting and 3-day history of numbness and clumsiness of his limbs. Apart from neck pain, he had no other symptom

Examination: power was preserved in upper limbs while lower limbs examination revealed distal power of 1-2/5 and proximal power of 4/5. The rest of CNS examination was unremarkable
Investigations including inflammatory marker, autoimmune screening and serum immunoglobulin levels were normal. CSF analysis showed normal albumin & Glucose, negative oligoclonal band and negative PCR for mycobacterium Tuberculosis.

The patient was admitted in the medical floor with an impression of demyelinating CNS disease for work up. Over subsequent 2 days after admission, his weakness markedly worsen with power in distal lower limbs reached 0-1/5 and proximally 2/5, while in the upper limbs it was 4/5. He had complete urine retention, difficulty in swallowing, and poor control of his anal sphincter. Based on the above, the impression was acute quadreparesis with swallowing difficulties and sphincters disturbance.
The patient was shifted to the ICU for close monitoring and regular Forced Vital Capacity measurement. His FVC & NIF were reducing & his saturation was not promising; accordingly, he was intubated and hooked to MV with sedation.

MRI brain and cervical spine was suggestive of Dural AV fistula malformation; accordingly, the neurologist impression was vascular myelopathy with dural AV fistula and the patients was referred to other facility with specialized interventional vascular neurology/neuro surgery center for further management. Interventional procedure was done on the 29th /September/2020. Report confirmed presence of High-grade arteriovenous fistula at the craniocervical junction with a large draining vein which drain retrogradely in to the anterior & posterior spinal veins.

Successful balloon assisted liquid embolic agent embolization of the arteriovenous fistula through branches of right occipital artery was done with complete obliteration of the fistula.



MRI report

There is medulla and upper cervical spinal cord expansile T2 hyperintense signal changes with coating hemosiderin staining and delayed enhancement in post contrast images suggestive of edematous changes. There are multiple abnormal enlarged small arteries in the craniocervical junction and upper cervical spinal cord with large draining vein seen in the right lateral aspect of the medulla, suggestive of dural arteriovenous fistula, which was confirmed lately with diagnostic and therapeutic cerebral angiography

Discussion:

After 4 months the patient reported mild improvement in the power of his upper limbs and persistent anal sphincter incontinence. Follow-up MRI 2 months after the intervention revealed Interval resolution of the previous medulla and cervical cord T2 hyper intense changes, and the previous posterior fossa serpiginous signal voids with residual small signal voids draining to straight sinus

Discussion

Although the common presentation of the Dural AVF take a form of step-wise pattern, the unusual presentation, like acute quadreparesis with swallowing difficulties and sphincters disturbance should be taken in account for rapid diagnosis and intervention that could improve the outcome.

Reference:

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