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An Interesting Case of Type 3 Spinal Cord Arteriovenous Malformation in a Pregnant Female Presenting As Compressive Myelopathy

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ABSTRACT

Arteriovenous malformation is described as an abnormal connection between arteries and veins bypassing the capillaries. Ten percent of the AV malformations of central nervous system are seen in the brain stem and spinal cord. There are 4 types of spinal AV malformations depending on the anatomic location. Type 3 or juvenile AVMs are seen in both extramedullary and intramedullary region. Type 3 AVM's are infrequently reported in the literature. This is a case of a 24 year old pregnant female who presented with a compressive myelopathy and diagnosed to have a type 3 spinal AV malformation on magnetic resonance imaging.

Keywords: Type 3 Spinal Cord, pregnant, myelopathy.

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INTRODUCTION

Arteriovenous malformation is described as an abnormal connection between arteries and veins bypassing the capillaries. AV malformations of the central nervous system are more commonly seen in the supratentorial region. Ten percent of the AVMs of CNS are seen in the brain stem and spinal cord. The prevalence of AVMs range from 1 - 4% of the general population. Spinal AVMs present with symptoms of back pain, lower limb weakness, disturbances of gait, bladder and bowel incontinence [1]. We present a case of type 3 spinal AVM in a pregnant female who presented with compressive myelopathy.

Case Presentation

A 24 year old female, primigravida at 12 weeks of gestation, presented to our medical outpatient department with complaints of dull aching pain over the neck since 3 months which was gradually progressive and was associated with progressive weakness, initially of the left upper limb followed by left lower limb, right lower limb and right upper limb in that sequence. Patient noticed negative sensory symptoms over the left half of the body and face. Patient had history of urinary incontinence since the past few days prior to presentation.

Patient was tachypneic on examination with a reduced single breath count. Nervous system examination showed following deficits

- Onion skin pattern of sensory loss over left side of face
- Weakness of left sternocleidomastoid
- Neck and trunk muscle weakness
- Upper motor neuron type of weakness of left upper and lower limb
- Sensory loss over left half of body

Examination of other systems was unremarkable.

Investigations

Magnetic resonance imaging of the cervico-medullary junction with angiography was suggestive of a spinal arteriovenous malformation (type 3).

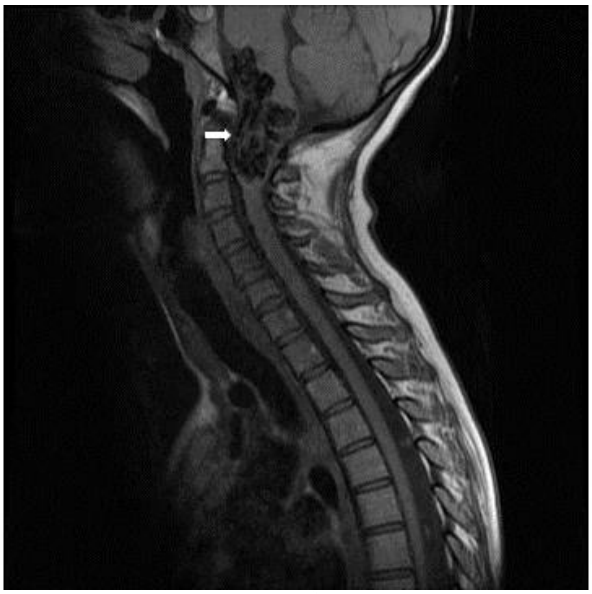

Figure 1

T1WI – SAG image :
Arrow: showing tortuous linear hypointense areas in the intramedullary and extra medullary portions of cervical cord at the level of cervicomedullary junction.

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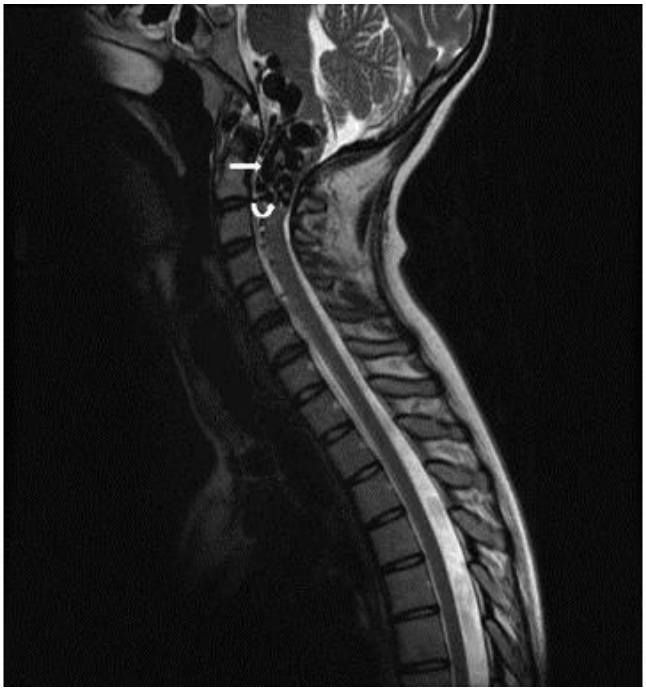

Figure 2

Cervical spine T2WI – SAG image-
Arrow: multiple flow voids in the extra medullary portions of cervical cord at the level of cervicomedullary junction.
Curved arrow: Intramedullary flow voids in cervical cord at the level of cervicomedullary junction

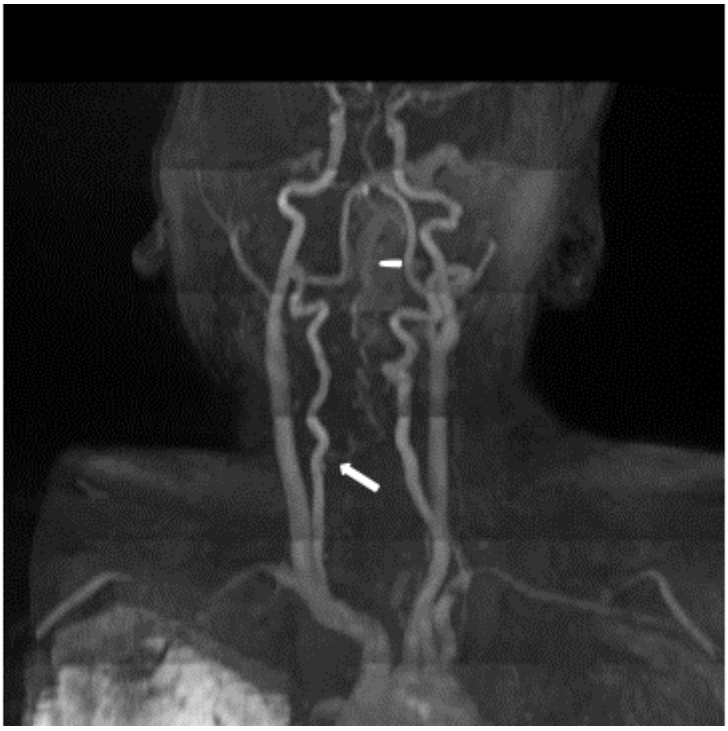

Figure 3

3D Reconstructed image of MR angiogram- showing
Arrow: Flow related enhancement in the form of tortuous arterial feeder from right vertebral artery (V2 segment)
Bullet: Draining veins

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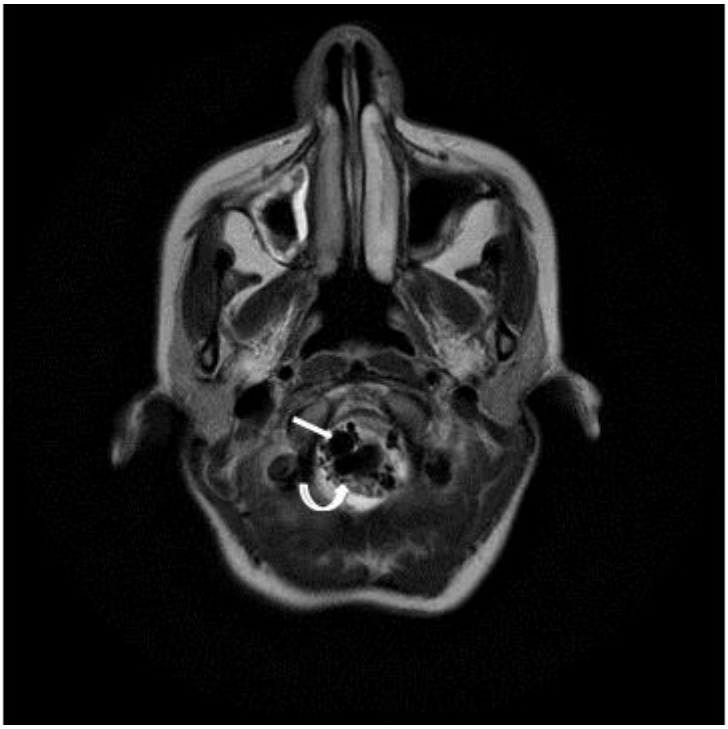

Figure 4

Axial T2WI :
Arrow: multiple flow voids in the extra medullary portions of cervical cord at the level of cervicomedullary junction.
Curved arrow: Intramedullary flow voids in cervical cord at the level of cervicomedullary junction

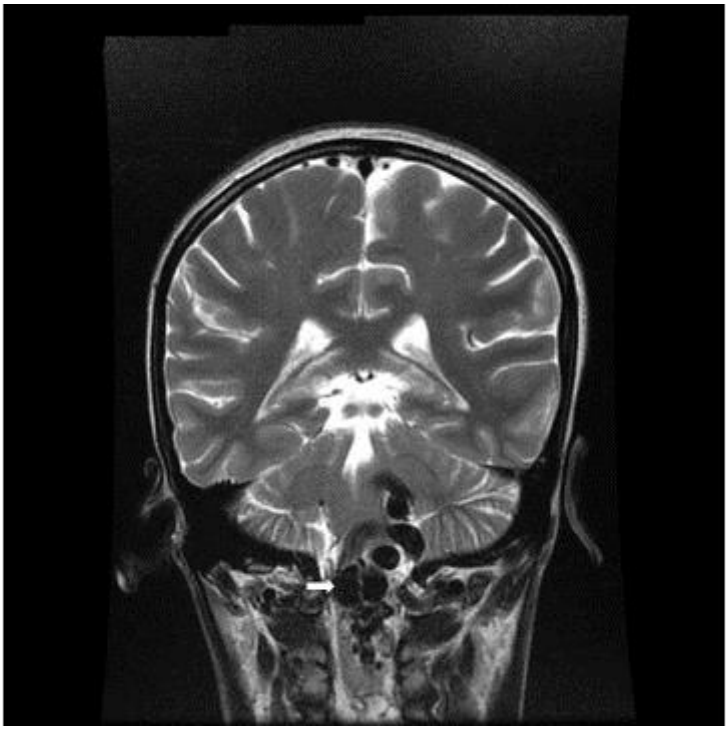

Figure 5

Coronal T2WI – Arrow: multiple flow voids in the extra medullary portions of cervical cord at the level of cervicomedullary junction.

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DISCUSSION

Spinal AVMs become symptomatic due to various pathophysiologic mechanisms such as subarachnoid hemorrhage, vascular steal causing ischemia, mass effect due to the abnormal vessels and associated aneurysms. Spinal AVMs are divided into four subtypes by Anson and Spetzler [2].

- Type 1 or dural AVM is the most common of all the spinal AVMs with 70% of all the spinal vascular malformations falling into this category.
- Type 2 or glomus AVM account for 20% of the spinal vascular malformations and are located in the intramedullary region.
- Type 3 or juvenile AVMs are seen in both extramedullary and intramedullary region.
- Type 4 or pial AVMs are seen in the intradural extramedullary region.

Type 3 AVMs which is diagnosed in our patient are relatively rare as seen in studies published by Malis LI et al and Ommaya ak et al [2,3].

Bradley A et al have done a pooled analysis of case reports of Type 3 spinal AVMs and have opined that Type 3 AVMs have the youngest age of presentation among the spinal AVMs. Other findings of this study include a slightly male predilection and higher prevalence of aneurysms compared to other spinal AVMs [4].

Hemorrhage from the Type 3 spinal AVM lesions is the most common cause of symptomatic presentation. The annual hemorrhage rate in an analysis by Gross and Du is 2.1%. Presence of associated aneurysms are considered as an additional risk factor for hemorrhage [5].

The treatment goal in a spinal AVM is to eliminate the shunt by endovascular surgery. Embolization with liquid embolic materials is done for most of the spinal AVM. Due to the presence of both intramedullary and extramedullary involvement in Type 3 spinal AVM, partial embolization is attempted in few cases with good outcomes and reduced hemorrhage rates during follow up. Securing of associated aneurysms also result in lower hemorrhage rates [6].

Outcome

Patient was transferred to a higher center specialized in management of spinal AV malformations.

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