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Rare Case of Diffuse Spinal Arachnoiditis Following a Complicated Vertebral Artery Dissection

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Introduction

Spinal arachnoiditis (SA) is an extremely rare and delayed complication of intracranial subarachnoid hemorrhage (SAH). SA is an inflammatory process leading to chronic fibrosis of the spinal cord. Possible pathophysiology is a two-staged disease of initial inflammatory reaction secondary to SAH, followed by a "free interval phase" prior to delayed adhesive phase (i.e. SA). The clinical course can be complicated and is the cause of major morbidity.

- 25 case reports since first described by Nelson in 1943. Ours is the 26th case reported.
  - Majority of cases occur in females (86%) between ages of 22 and 69 years old.[8]
  - High predilection of SA at level of thoracic spine[8]
  - 18/25 had SAH after aneurysm rupture.
  - 16/18 involved posterior circulation arteries: 13 PICA*, 2 PCom*
  - Our case involves the vertebral artery (VA)
  - SA can arise anytime from 1 month to 12 years after SAH.
- SA complications include arachnoid cysts[1-4], syringomyelia[2,5,6], and spinal cord compression (e.g. cauda equina syndrome).[7]
- Little is still known about underlying inflammatory pathogenesis and clinical pattern.

PICA = posterior inferior cerebellar artery, PCom = posterior communicating artery, VA = vertebral artery

Clinical Case Report

DAY 0-1

- **47-year-old female** complains of having the "worst headache" of her life and a grade 3 SAH.
- **Past medical history**: benign paroxysmal positional vertigo (BPPV)
- **Diffuse Subtraction Angiography (DSA)**:
  - Ruptured dissection/dissecting fusiform aneurysm of **V4 segment of left VA**
  - Intact 1.7 x 1 mm aneurysm at proximal **A1 segment of left ACA**
- **CT scan**: diffuse SAH in the basal cisterns and posterior fossa extending through the foramen magnum.
- **VA dissection** was treated with flow diversion using pipeline embolization (PE).

TWO WEEKS LATER

- Patient ruptured her **A1 ACA aneurysm** and re-ruptured/re-bled from her **VA dissection**.
- **A1 ACA aneurysm** was treated with PE. **VA dissection** was re-treated with second PE.
- **Hospital course complications**:
  - Polymicrobial ventriculitis – treated with PICC-line broad spectrum antibiotics.

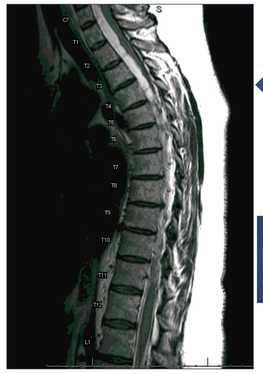
SEVERAL MONTHS LATER

- New onset of paraparesis and left-sided weakness, dizziness, positional nausea, and gait ataxia.
- **MRI** *(fig. A, B)* showed thecal dural thickening from the cervicomedullary junction → C4 and from T5 extending circumferentially → sacrum.
- **MRI** *(fig. A)* also showed various degrees of cord compression along inferior surface of cerebellum, anteriorly within cervical and thoracic canals, and T4-T6 syrinx with diffuse spinal edema.
- Ventricular shunt and decompression surgery of T5-T7 were performed.

LATEST FOLLOW-UP

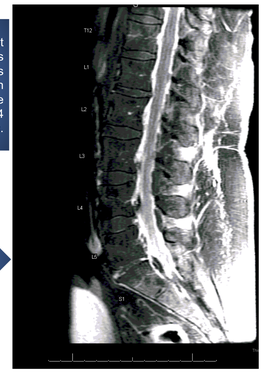
- Patient had full-strength with moderate gait instability and was weaning off of her thoracolumbosacral orthosis back brace. She currently ambulates with a cane.

Radiologic Findings



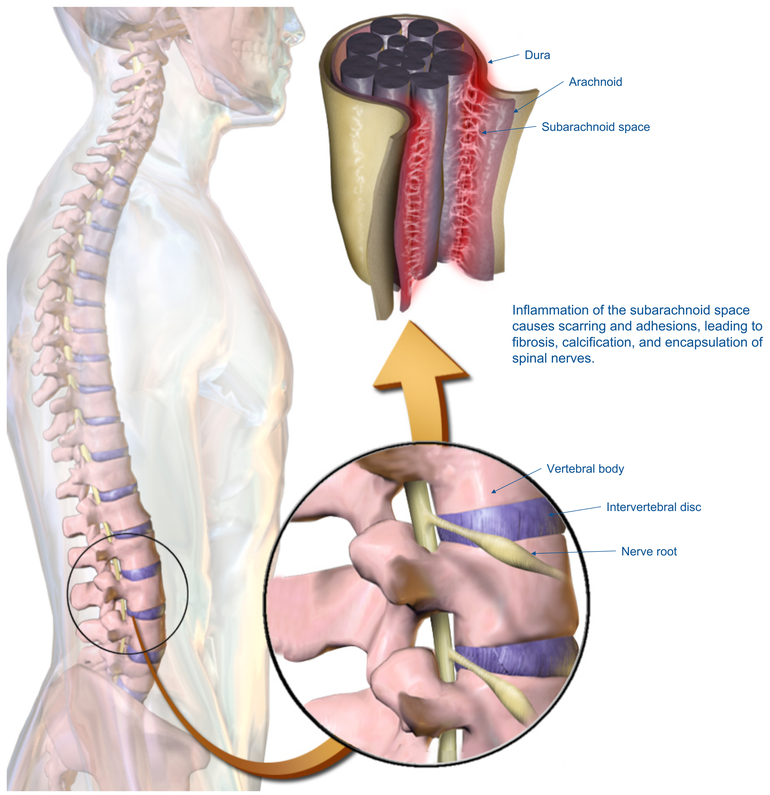
MRI thoracic spine w/o contrast shows multiple loculated collections within thecal sac causing various degrees of cord compression within cervical and thoracic canals. There is cord edema extending from T4 → through the conus.

MRI lumbar spine w/o contrast shows similar findings as shown in the cervical and thoracic canals with cord compression and edema.



Pathophysiology

- Hemolysis in ruptured aneurysm incites chronic irritation of leptomeninges.
- Inflammatory reaction in the intrathecal compartment may lead to arachnoiditis.
- Nerve root injury and spinal syrinx can subsequently develop.
- Chronic inflammation leading to arachnoid cysts in pia-arachnoid space and fibrotic SA have been widely described in literature.
- Increase in procollagen propeptides in the CSF after SAH has been demonstrated in a time-dependent manner, suggestive of fibroproliferative reaction.[11,12]



Discussion

Our unique case presentation:

- Patient developed SA with extensive, multiple localizations, involving segments from the posterior fossa, extending from cervical area to the sacrum.
- One other case had comparable diffuse arachnoiditis of thoracic and lumbar spine.9
- Involvement of cervicomedullary junction, the highest-involved region currently reported in literature. Previously reported highest-involved region was C2.[10]
- Unique symptoms of ataxia, nausea, and vomiting.
- First reported SA from a ruptured VA aneurysm.
- Re-rupture of aneurysm complication.
- Polymicrobial ventriculitis and encephalomalacia in the left and right PICA territories
- Unclear if these complications contributed to development of diffuse, severe SA
- Emphasis of potential inflammatory role succeeding SAH and leading to SA development, highlighting the need for closer surveillance of patients with posterior fossa/circulation SAH.[11]

Conclusions

Clinical course of SA after SAH can be complicated and is the cause of major morbidity. Patients showing signs and symptoms of SA following SAH, especially those involving the posterior circulation, should be followed closely as time of onset ranges from days to years after SAH. There may be an increased risk of SA after SAH of ruptured posterior circulation aneurysms. Chronic inflammation is widely believed to play a major role in SA pathophysiology. Measuring inflammatory markers and procollagen peptides in CSF after SAH may help determine degree of inflammation and subsequent risk of SA.

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