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# An unusual case of remitting and relapsing intraspinal epidural hamartoma with hemorrhage

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***Abstract***

Spinal epidural hematoma is a rare disease and is most commonly idiopathic. Other common causes are vascular malformations, coagulopathy, hypertension, surgery, and trauma. Epidural vascular hamartoma associated with hemorrhage is a less known entity. We report an unusual case of epidural vascular hamartoma associated with hemorrhage resulting in back pain and neurological deficits. A distinctive feature of this case was waxing and waning signs and symptoms with evidence of radiological resolution of the lesion in the phase of remission.

*Keywords:* Spinal, epidural, hematoma, vascular hamartoma

## INTRODUCTION

Spinal epidural hematoma is a well-recognized but rare disease. Vascular malformations are a well-known cause of spinal epidural hemorrhage. However, epidural vascular hamartoma associated with hemorrhage resulting in compressive myelopathy has never been described in literature. We report a case of remitting and relapsing case of epidural vascular hamartoma with hemorrhage. Recurrent back pain and neurological deficits with symptomatic improvement and radiological resolution of the epidural lesion during the phase of remission was the unique feature of this case. This case report includes the neuroimaging, intraoperative and pathologic findings of the case with literature review of spinal epidural hematoma.

## CASE REPORT

A 25-year-old male presented with history of sudden onset back pain, which was gradually progressive, associated with imbalance in walking, weakness in bilateral lower limbs and paresthesia in bilateral lower limbs up to upper abdomen below nipple region. There was no history of trauma. No history of bladder/ bowel dysfunction. General and systemic examination were within normal limits. Tone and power in bilateral upper and lower limbs were normal. Bilateral triceps and ankle jerks were hypoactive. Sensory system evaluation revealed 80% sensation below T5 level. MR imaging revealed an extradural lesion at D2-D3 level displacing the cord anteriorly and causing mass effect on the spinal cord associated with compressive myelopathy. The lesion appeared predominantly hyperintense on T2W with interspersed areas of hypointensity [Figure 1A and 1B] and isointense on T1W sequence [Figure 1C and 1D]. There was no evidence of any diffusion restriction or post contrast enhancement [Figure 1E and 1F]. The imaging findings were non-conclusive and there was evidence of gradual spontaneous recovery in the signs and symptoms after 48-72 hours of admission.

MR imaging was repeated after 3 days that revealed significant interval reduction in the size of the lesion as compared to the previous scan. A thin linear T2 hyperintense signal was seen in the posterior epidural space at D2-D3 level [Figure 2A and 2B] with no significant mass effect on the cord. No obvious signal alteration was noted on T1W sequence [Figure 2C]. A thin linear susceptibility signal was detected on SWI sequence [Figure 2D] with no evidence of diffusion restriction or post contrast enhancement [Figure 2E and 2F]. Cord appeared normal in morphology and signal intensity.

In view of significant reduction in the size of the lesion and spontaneous resolution of symptoms, surgery was deferred, and the patient was discharged and advised follow up MR imaging after 3 months. Radiological evidence of the complete resolution of the lesion was likely to

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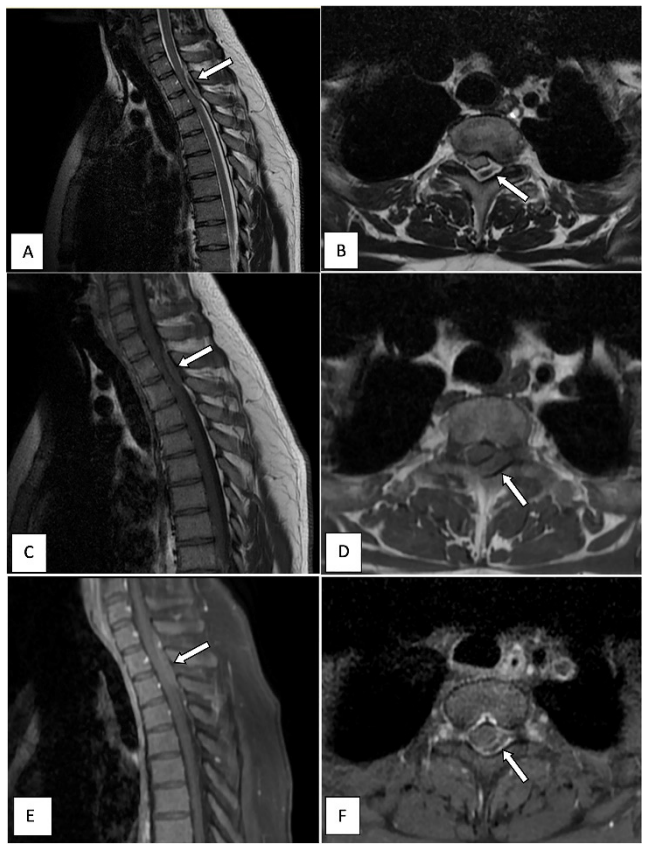


Figure 1. 25-year-old male with a spinal epidural lesion presented with severe back pain and weakness in bilateral lower limbs. T2W sagittal (A) and axial (B) images demonstrate an epidural hyperintense lesion at D2-D3 level, with interspersed hypointense areas (white arrows) causing mass effect on the cord. The lesion appears isointense on T1W sagittal (C) and axial (D) images (white arrows). No evidence of any enhancement noted on post contrast images (E and F) as depicted by white arrows.

represent epidural hematoma. However, the cause of the hemorrhage could not be found.

After a few months, the patient again developed back pain and weakness in bilateral lower limbs and paresthesia that did not resolve spontaneously as it happened in the previous episode. Patient was admitted to our center for further evaluation and management. General and systemic examination were within normal limits. Bilateral lower limbs revealed grade III spasticity. Power in bilateral lower limbs was significantly reduced. Bilateral biceps jerks were hypoactive. Bilateral ankle and knee jerks were hyperactive and there was extensor response on plantar reflex. Sensory system evaluation revealed reduced sensation below T4 level. MR imaging of spine revealed a well-defined altered signal intensity lesion in the posterior epidural space extending from the level of lower border of D1 to upper border of D3 vertebral body, displacing the spinal cord towards right side. The lesion appeared predominantly hyperintense on T2W sequence [Figure 3A and 3C] and isointense on T1W sequence [Figure 3B]. Multiple T2 hypointense foci with susceptibility signals were seen within the lesion [Figure 3D]. Diffusion weighted imaging revealed restricted diffusion with ADC values in the range of 0.4 x 0.5 x 10-3 mm2 / sec [Figure 3E]. No evidence of any abnormal enhancement detected on dynamic contrast enhanced imaging [Figure 3F and 3G]. Focal area of T2 hyperintensity was noted in the cord at D2 level. He was diagnosed to be having D1-D3 left extradural lesion with myelopathy resulting in paraparesis and bilateral lower limb spasticity.

He underwent left D1-D3 minimally invasive hematoma evacuation using METRx tube system under general anaesthesia. He tolerated the procedure well and made uneventful recovery. Histopathological evaluation revealed bone bits with vascular channels of various caliber and evidence of fresh and old hemorrhage to suggest underlying vascular hamartoma [Figure 4].

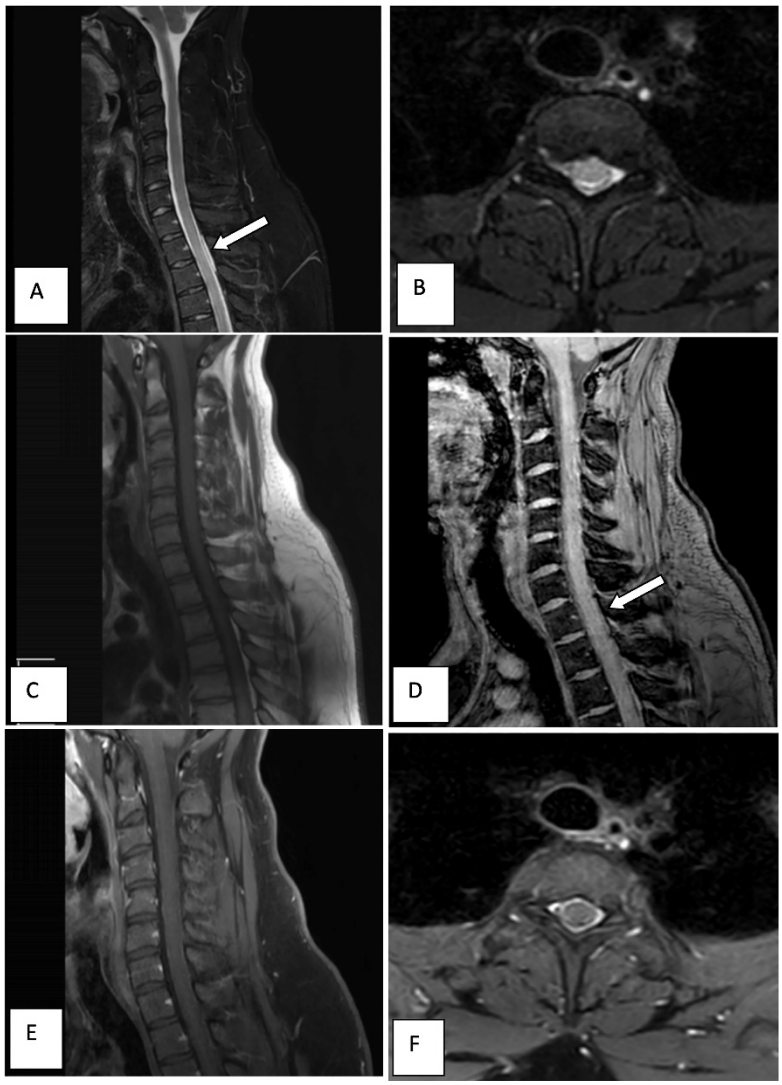


Figure 2. MR imaging repeated after 3 days revealed significant reduction in the size of the previously noted epidural lesion. A thin linear streak of hyperintensity is noted on T2W sagittal image at D2-D3 level (A, white arrow) with no mass effect on the spinal cord (B). No obvious signal abnormality detected on T1W image (C). Thin linear susceptibility signals with negative phase can be seen on SWI sequence at D1-D3 level (D, white arrow). No evidence of any abnormal enhancement noted on post contrast images (E and F).

## DISCUSSION

Epidural hematomas are most commonly idiopathic. The valveless state of the epidural venous plexus is thought to predispose it to rupture with sudden changes in pressure that can be affected by position, CSF pressure, and thoraco-abdominal pressures.[1,2] This is thought to be due to transmission of raised intra-abdominal or intrathoracic pressure from apparently mild trauma or straining.[3,4] Symptoms are often triggered by bending, sneezing, voiding, straining or turning in bed.[1] Beatty and Winston suggested that an arterial source of bleeding originating from the extensive network of epidural arteries better explains the precipitous neurological deterioration seen clinically.[3,5] An arterial source and microscopic subperiosteal vascular malformations have also been suggested.[4] Vascular malformations and vascular tumors can be a cause of bleeding in some cases. Small cryptic vascular malformations can be a plausible cause of many episodes of bleeding that can be obscured by resultant hemorrhage.[6] It was histologically proven by Solero and Fornari that 10% of spontaneous spinal epidural hematomas have arteriovenous

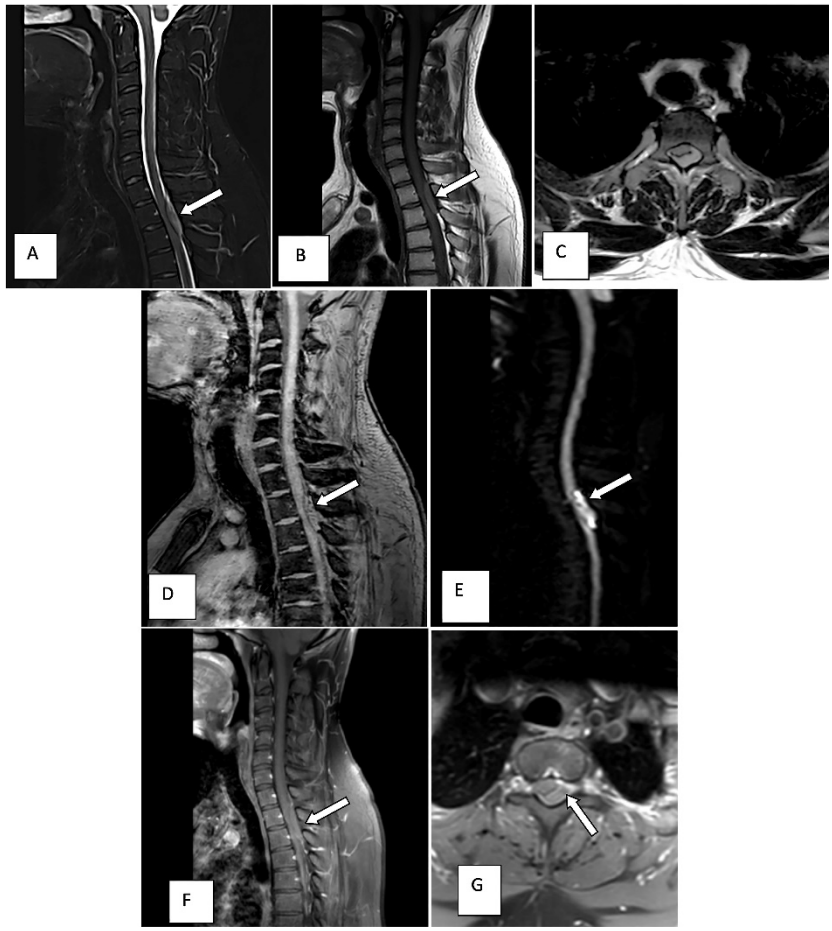


Figure 3. MR imaging repeated after 10 months for similar symptoms revealed an epidural lesion at D2-D3 level. T2W sagittal (A) and axial (C) images demonstrate an epidural hyperintense lesion at D2-D3 level, with interspersed hypointense areas (white arrows) causing mass effect on the cord. The lesion appears isointense on T1W sagittal (B) image (white arrow). Few susceptibility signals are seen within the lesion on SWI sequence (D, white arrow). The lesion displays restricted diffusion (E, white arrow). No evidence of any enhancement noted on post contrast images (F and G) as depicted by white arrows.

malformations.[7] It has also been postulated that, if all the clots were always saved and examined histopathologically, vascular anomalies would be found more frequently.[7] Additional causes include coagulopathy, hypertension, trauma and iatrogenic causes from interventional pain management procedures.[2,3] Gundry and Heithoff suggested that spinal epidural hemorrhage can result from rupture of fragile epidural veins by an adjacent herniated disc.[1,8]

Relapsing and remitting epidural lesion in our case was vascular hamartoma with hemorrhagic changes. Mass effect due to the epidural vascular hamartoma and associated hemorrhage was the cause of back pain and other neurological symptoms. Sudden change in intrathecal pressure from apparently mild trauma or straining must have resulted in rupture of the aberrant thin-walled vasculature in the hamartoma. Evolutionary changes in the hemorrhagic contents with longitudinal spreading of the hemorrhagic contents within the epidural space would be the most plausible cause of reduction in the size of the lesion and spontaneous resolution of symptoms. However, recurrent hemorrhage due to the underlying vascular hamartoma, resulted in relapse of symptoms. Due to neurological deficits and further deterioration, surgical intervention was planned. Evacuation of the hamartoma and the hemorrhagic contents resulted in uneventful recovery and the patient was discharged.

Few cases of remitting and relapsing epidural hemorrhage with underlying coagulopathy have been reported.[6] However, epidural hemorrhage

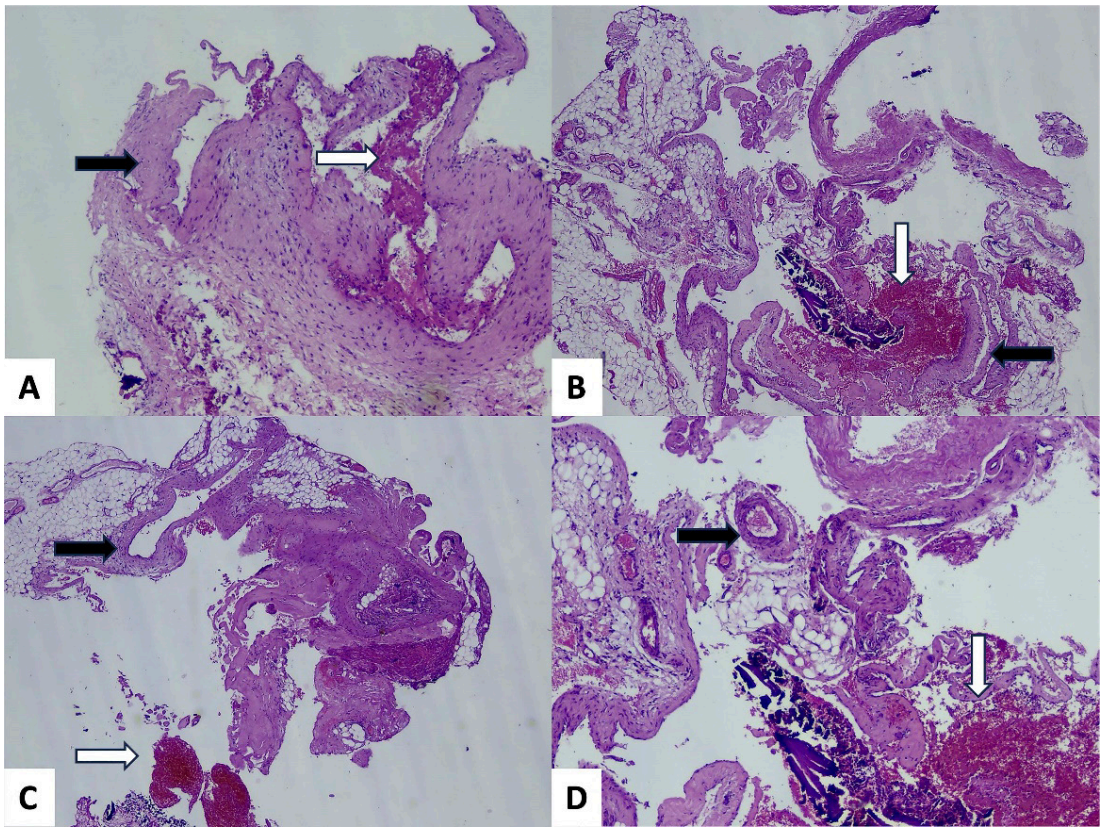


Figure 4. H&E images showing bone bits with vascular channels of various caliber (black arrows) and evidence of fresh and old hemorrhage (white arrows).

associated with vascular hamartoma, presenting with waxing and waning symptoms has never been reported, to the best of our knowledge.

In conclusion, spinal epidural hematoma is a relatively rare condition. Vascular malformations are well known cause of epidural hematomas. To the best of our knowledge this is the first case report of epidural hamartoma with hemorrhage causing myelopathy. The unique feature of our case is the remitting and relapsing nature of the disease. It is recommended that in patients with radiological evidence of epidural hemorrhage, prompt surgical evacuation should be considered followed by histopathological evaluation.

## DISCLOSURE

Conflicts of interest: None

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