Uncommon vascular and hematological causes of extradural cord compression and its clinical impact

Poster No.: C-1889
Congress: ECR 2018
Type: Educational Exhibit
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Keywords: Obstruction / Occlusion, Cerebrospinal fluid, Diagnostic procedure, MR, CT, Vascular, Musculoskeletal spine, Neuroradiology spine, Haematologic diseases
DOI: 10.1594/ecr2018/C-1889

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Learning objectives

To highlight the key imaging findings of uncommon vascular and hematological causes of extradural cord compression and to facilitate early detection and appropriate management of these critical cases of compressive myelopathy.
**Background**

Most common causes of extradural compressive myelopathy are discogenic lesions, infective pathologies and tumors of the spine. Vascular and hematological causes of compressive myelopathy are not common and often are misinterpreted as malignant lesions. Hence it is important to recognize these uncommon causes of extra-dural compression of the spinal cord, as the management strategies are completely different from the malignant lesions causing compressive myelopathy.
Findings and procedure details

Imaging findings:

Uncommon benign vascular and hematological causes of compressive myelopathy documented in this exhibit are

A) Chronic contained rupture of the aortic aneurysm:

Aortic aneurysm is one of the uncommon causes to be considered while clinical examination of the patients with back pain. Rupture of aortic aneurysm is a known complication, and if acute can result in severe pain and hemodynamic instability with poor prognosis(1,2). In few cases, the rupture can be walled with adjacent structures or inflammatory reaction thus resisting active extravasation of blood in to retroperitoneum(3). On imaging, draped aorta sign (posterior aspect of aorta draping vertebral bodies), retroperitoneal organized hematoma, and adjacent lysis of vertebral column can occur in these cases(4,5,6). In addition to all, discontinuity in the mural calcification, paraaortic soft tissue density, displacement of the adjacent structures and lack of contrast material in the hematoma are also useful in the diagnosis of the contained rupture of the aortic aneurysm (7). Most of these findings were observed in the present case(Fig 1-3).

Vertebral involvement in aortic aneurysm rupture is not very common finding. Usually the vertebra has inherent strength to tamponade the aneurysm effectively. Erosion of the vertebral bodies by an abdominal aortic aneurysm is seen in only 7% of cases. The aortic pulsations along with contained rupture resulting in perianeurysmal hematoma formation might exert constant and progressive mass effect on the vertebral column resulting in chronic remodeling and lytic destruction of the bone, though exact pathogenesis is unclear. (6,8,9,10,11).

Differential diagnosis in this setting is pyogenic infection in which soft tissue and vertebral destruction occurs similar to the chronic contained rupture; irregular margins of the destruction helps in differentiation from chronic aneurysmal rupture which shows smooth margins(12).

Even though aneurysmal rupture is chronic and contained, there is risk of re-rupture of the aneurysm, which makes the immediate treatment of aortic aneurysm essential.(13). The patient in the study (Fig 1) was treated by aortic repair with covered graft.
B) **Extra medullary hematopoiesis:**

Extramedullary hematopoiesis (EMH) refers to hematopoiesis occurring anywhere other than bone marrow(14-19) in Chronic anemias like thalassemias, sickle cell anemia, Most commonly, intra-thoracic EMH occurs in posterior mediastinum and middle and lower paravertebral areas. Very rarely these lesions can also involve or extend to the extradural space within the canal and cause spinal cord compression(20, 21, 22).

Many of the extra-medullary hematopoietic masses are incidentally detected and are asymptomatic. Rarely, para-spinal masses can result in the cord compression, pleural effusion, massive hemothorax and respiratory failure. (23, 24)

On CT, these lesions appear as isolated/ multiple lobulated paraspinal/ posterior mediastinal soft tissue masses on either side and enhances intensely on contrast administration. On MRI, these masses depending on the presence of iron, hemosiderin, meth/ deoxy hemoglobin, show low signal intensity on T2WI and intense enhancement on post contrast study(25)( Fig 4-5).

Treatment options include surgical decompression / radiation therapy. (24)

C) **Vertebral Extra-dural cavernous hemangioma causing spinal cord compression:**

Hemangioma involving vertebra are most- commonly benign lesions with no neurologic symptoms. Rarely, these lesions can be aggressive , with involvement of the vertebral body and neural arch resulting in expansion with associated extensive soft tissue component. (26).

Characteristic findings on CT are thickened vertical bony trabeculations of the vertebra resulting in corduroy / polka dot appearance. On MRI, hemangiomas typically show hyperintense signals on T1WI (due to fat) and hyperintense signals on T2WI (due to water). However, aggressive hemangiomas have reduced water and fat content and are highly vascular resulting in hypointensity on T1WI and hyperintensity on T2WI. (27, 28).

All of these characteristic features of hemangioma associated with epidural and paravertebral soft tissue component having extremely bright signal intensity on T2WI and intense enhancement are demonstrated in this study. ( Fig 6-9)
Aggressive hemangiomas are commonly misdiagnosed as metastases, lymphoma or paget's disease. However, extreme hyperintensity on T2WI, (Fig 6) osseous remodelling with corduroy appearance, absence of cortical thickening helps in differentiating hemangioma from these lesions. (29).

**D) Spinal Epidural Cavernous Angiomas are** relatively rare lesion especially at the spinal level (4% of all spinal epidural tumors)(30). Epidural cavernous angiomas have no bony involvement and are frequently located at the thoracic level with predilection for the T2-T6 segment. If present, they usually involve the posterior part of the epidural space and extend to the extraspinal space.

On MRI, these lesions usually appear hyperintense on T2 WI due to the high content of stagnant blood. Striking difference from the intra-axial cavernomas, is the lack of the low signal hemosiderin ring on both T1 & T2-WI presumably due to easier removal of blood products outside the blood-brain barrier(30). Despite epidural cavernous hemangiomas can mimic extradural meningiomas, being hyperintense signal on T2 WI and intense enhancement on Gado are the characteristic MRI findings of this vascular malformation. (Fig 10-12).

**E) Spinal Dural A-V fistula:**

Vascular malformations of the spinal cord include cavernous malformation, intradural arterio-venous malformation, perimedullary arterio-venous fistulas and dural arterio-venous fistulas. (31-35). Understanding and differentiating each of these lesions is essential for accurate diagnosis and appropriate treatment (36-40).

Spinal dural arteriovenous fistuals (AVFs) are thought to be acquired lesions, showing low flow shunts that are supplied by the dural branch of the intervertebral artery. Spinal dAVFs most commonly affect men in their 5th decade of life(30).

The nidus is composed of a network of separate vessels that converge into the medullary vein that carries shunted blood retrograde to the coronal venous plexus. Due to the valveless nature of the intrathecal venous system, increased pressure within the medullary veins subsequently leads to venous hypertension, venous congestion, producing ischemic injury to the spinal cord and progressive myelopathy [41]as seen in Fig 14 &15.
The goal of treatment for spinal dural AVFs is to eliminate the venous congestion either by interruption of the venous drainage from the fistula between the dura and the dilated coronal venous plexus or by elimination of the nidus. Success has been shown with endovascular embolization prior to surgical intervention [42].

F) Spontaneous epidural hematoma compressing the cord:

A spinal epidural hematoma is a rare but significant neurological condition. The most common site of a spontaneous spinal epidural hematoma is the cervicothoracic region or thoracolumbar region after the fourth or fifth decade (43).

Patients usually presents with sudden onset stabbing pain in the neck or back, which gradually progresses to paraparesis or quadriparesis, depending on the level of the lesion and the nerve root (44).

Precipitating factors includes anticoagulant therapy for prosthetic cardiac valves, therapeutic thrombolysis for acute myocardiac infarction, hemophilia B, factor XI deficiency, long-term aspirin using as a platelet aggregation inhibitor, and vascular malformations (45).

MRI is the diagnostic modality of choice that shows biconvex shaped epidural collection with well defined borders tapering superiorly and inferiorly. Sub acute hematoma shows high signal intensity on T1-weighted image (46). Similar findings are demonstrated (Fig 16) in a known case of Christmas factor deficiency, who presented with spontaneous epidural hematoma.

G) Spinal epidural angiolipoma

Spinal epidural angiolipomas are rare, accounting for 0.04-1.2% of spinal axis tumors and 2-3% of extradural spinal tumors (47-48). These benign tumors are composed of mature lipocytes admixed with abnormal blood vessels.

MRI is the investigation of choice in diagnosing spinal angiolipoma [48-49]. Angiolipomas are iso- or hyperintense on T1-weighted images due to fat content and variable in T2-weighted images of MRI, The degree of central hypointensity on T1-weighted images is predictive of the degree of vascularity (49,50).
In this exhibit, non-contrast T1-weighted images of MRI dorsal spine displays epidural angiolipoma with lipomatous element and multiple foci of linear and serpiginous signal voids (in both T1 and T2WI), representing high degree vascularity, compressing the spinal cord (Fig 17).

**H) Aneurysmal bone cyst (ABC) of the spine**

ABCs are benign tumour-like expanding osteolytic lesions, consisting of blood-filled spaces of variable size separated by connective tissue septa containing trabeculae or osteoid tissue and osteoclast giant cells (51). They can cause acute spinal cord compression in young patients (52).

The pathophysiology of the ABC, is unknown. Among the different theories (53), arterio-venous fistula and venous blockage is one proposed etiology. The vascular lesion cause increased pressure, expansion, erosion and resorption of surrounding bone. The malformation is also believed to cause local haemorrhage, that initiates the formation of reactive osteolytic tissue.

This exhibit CT and MRI findings in a histology-proven case of spinal ABC presenting with sudden paraplegia. Typical features of a spinal ABC at the thoracic level involving posterior neural arches with considerable extension into the posterior epidural space and cord compression is demonstrated (Fig 18).
Fig. 1: Saccular aneurysm in descending thoracic aorta with chronic contained rupture. 58yrs male presented with weakness in both lower limbs for 4 days. Pain in the back of the chest for 15days. Plain CT- Peripheral rim calcification (arrow head) in the wall of the descending thoracic aorta with large peri-aneurysmal thrombus and hematoma. CECT- Saccular aneurysmal outpouching (thick arrow) from left postero-lateral aspect of the aorta at the level of D8-9 with crescentic multiple layers of hematoma and thrombus formation.(thin arrow). Erosion of the lower dorsal vertebrae with intraspinal extension(curved arrow) causing cord compression and displacement.

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Fig. 2: Saccular aneurysm of descending thoracic aorta with chronic contained rupture. Peri-aneurysmal thrombus and hematoma showing varying phases of hemoglobin degradation products (thin arrow) in the left paravertebral region causing marked erosion of the vertebrae (thick arrow) with extradural extension.

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Fig. 3: Saccular aneurysm descending thoracic aorta with large chronic contained rupture in the paravertebral region. Extradural cord compression by peripheral contained hematoma (curved arrow) better delineated on T1W and T2W.

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Fig. 4: Extra medullary hematopoiesis: 32 yrs male -known case of Thalassemia, presented with fatigue and progressive weakness in both lower limbs for 3 weeks. CXR - Lobulated soft tissue opacities on both sides of the posterior mediastinum with retro-cardiac opacities, expansion of the ribs at costo-vertebral junctions and anterior ends of ribs (curved arrows). CT - Medullary hyperplasia with expanded hemopoietic bone marrow at the costo-vertebral junction (thin arrow) and along the ribs (curved arrow); Hepato-splenomegaly and hematopoietic tissue involving cortex of the left kidney (thick arrows).

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**Fig. 5:** Extra medullary hematopoiesis: Lobulated extradural hematopoietic soft tissue component (thin arrow) in the posterior aspect of spinal canal at mid thoracic level, compressing the spinal cord. Medullary hyperplasia (curved arrow) with expanded hemopoietic bone marrow at the costo-vertebral junction and along the ribs.

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Fig. 6: Aggressive hemangioma of D4 vertebra. 45 yrs male presented with progressive weakness for 4 weeks. Trabeculated appearance of D4 vertebral body, bilateral pedicles and right transverse process; associated lobulated extradural soft tissue component (thick arrow) in the right antero-lateral aspect of spinal canal at T4 thoracic level compressing the spinal cord. The soft tissue component extends through the right neural foramen (curved arrow) into the right paravertebral region. (arrowhead)

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**Fig. 7:** Aggressive hemangioma of D4 vertebra. Trabeculated appearance of D4 vertebral body, bilateral pedicles and right transverse process; associated lobulated extradural soft tissue component (thick arrow) in the right antero-lateral aspect of spinal canal at T4 thoracic level, compressing the spinal cord. The soft tissue component extends through the right neural foramen (curved arrow) into the right paravertebral region.(arrowhead)

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Fig. 8: Aggressive hemangioma of D4 vertebra. Trabeculated appearance of D4 vertebral body, bilateral pedicles and right transverse process; associated intensely enhancing lobulated extra-dural soft tissue component (thick arrow) in the right antero-lateral aspect of spinal canal at T4 thoracic level compressing the spinal cord. The intensely enhancing soft tissue component extends through the right neural foramen (curved arrow) into the right paravertebral region. (arrowhead)

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**Fig. 9:** Aggressive hemangioma of D4 vertebra. Trabeculated appearance of D4 vertebral body, bilateral pedicles and right transverse process; associated intensely enhancing lobulated extradural soft tissue component (thick arrow) in the right antero-lateral aspect of spinal canal at T4 thoracic level, compressing the spinal cord. The intensely enhancing soft tissue component extends through the right neural foramen (curved arrow) into the right paravertebral region. (arrowhead)

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**Fig. 10:** Spinal Epidural Cavernous Angioma 31 yrs aged female presented with low back pain with mild weakness in both lower limb - 4 months. Sagittal T1 WI and T2 WI shows extra-dural lesion in the posterior part of the spinal canal obliterating epidural space from D9 to D12 levels, compressing the cord. The lesion exhibits mixed intensity (intensity more than that of spinal cord) on T1WI (arrows) and extremely bright signal intensity on T2 WI.

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Fig. 11: Spinal Epidural Cavernous Angioma. Axial T1 WI shows extra-dural lesion in the posterior part of the spinal canal extending into right para-spinal region through the ipsilateral neural foramen. Cord is compressed and displaced anteriorly and laterally to the left by the lesion. The lesion exhibits areas of high signal intensity (arrow) on pre-contrast T1 WI in the right para-spinal region.
**Fig. 12:** Spinal Epidural Cavernous Angioma Axial Post Gado T1 WI shows the extradural and para-spinal component of the lesion showing intense enhancement.

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Fig. 13: Spinal Epidural Cavernous Angioma post operative study shows complete resection of the lesion

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Fig. 14: Spinal dural arteriovenous fistula of spinal canal. 39 yrs male presented with progressive weakness of both lower limbs. Multiple abnormal vascular channels (arrows) in the spinal canal in extra medullary and extradural compartments. Diffuse signal alteration involving the spinal cord from D7-8 to D11 levels.

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Fig. 15: Spinal dural arteriovenous fistula of spinal canal. Multiple abnormally enhancing vascular channels (arrows) in the spinal canal in extra-medullary and extra-dural compartments (thick green arrows). Diffuse signal alteration involving the spinal cord from D7-8 to D11 levels- myelopathy changes secondary to increased pressure in vascular channels.

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Fig. 16: Epidural hematoma of the spinal canal from C3 to C6 levels. 45 yrs male presented to ER with severe posterior neck pain which had occurred suddenly. Coronal T1-WI and Axial T1-WI MR image showing iso to hyperintense space occupying lesion (arrows) left lateral part of the epidural space from C3 to C6 levels. On Sagittal T2 WI, the lesion shows hyperintense signals with marginal areas of dark signal intensity -representing haemorrhage. Detailed hematological examination revealed Christmas factor deficiency in the patient -persisting for about 2 months.

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Fig. 17: Spinal epidural angiolipomas. 56 yrs aged male presented with weakness in both lower limbs. T1-weighted image of MRI dorsal spine showing hyperintense signals with multiple foci of linear (Thin arrow) and serpiginous signal voids (Thick arrow) within the epidural lesion at C4- D7 level, causing spinal cord compression; T2WI images of MRI dorsal spine showing hyperintensity with persistent signal void areas; T1W coronal images demonstrate cord compression.

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**Fig. 18:** Aneurysmal bone cyst in the posterior neural arch of D7. 15 yrs aged child presented with weakness in both lower limbs. Sagittal T1 and T2 W images show expansile cystic lesion involving posterior neural arch of D7 causing cord compression at this level. The lesion shows iso to mixed signal intensity on T1WI and bright signal with dark signal areas on T2 WI - representing haemoglobin degradation products. NECT reveals expansile lytic lesion with thin rim of cortex in the lamina of D7 vertebra.

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Conclusion

Imaging plays a pivotal role in recognition and early diagnosis of these uncommon vascular and hematological causes of compressive myelopathy. Most of these lesions have characteristic imaging findings and pattern. Nevertheless in few instances these can mimic common spinal pathologies. In these scenarios differences must be sought which are crucial in planning the management, thereby reducing the morbidity and mortality of these patients.
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