Extradural spinal cavernous malformation: a review and presentation of a typical case

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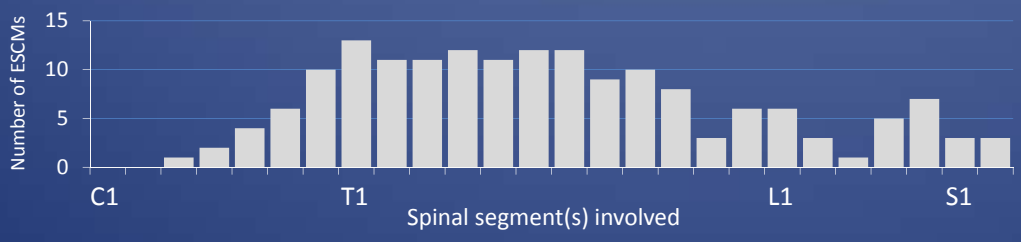
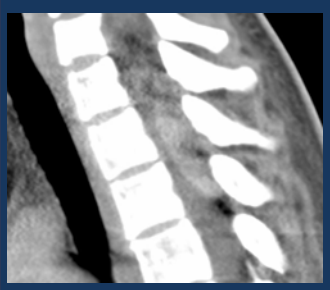
Background: Purely extradural spinal cavernous malformations (ESCM) are rare, but the number of cases reported seems to be increasing.

Methods: A 44-year-old woman presented with progressive cervicothoracic back pain, lower limb paraesthesia and hypertonia. MRI showed a well-circumscribed, convex lesion in the T1-4 extradural space causing cord compression and encroaching into the left T2/3 neural foramen. Selective spinal angiography demonstrated a vascular blush at the T2/3 level corresponding with the mass. A feeding vessel arising from the base of the T1 spinous process was identified and successfully catheterised, but embolisation was precluded by unfavourable anatomy of the posterior spinal artery. At hemilaminectomy, the lesion was found to be an ESCM. The patient went on to make a full recovery. We reviewed 71 cases of ESCM reported in the last decade.

Results: Incidence of ESCM is unclear; the largest case series reported on 9 patients but most published accounts were single case studies. Patients presented with back pain (33%), myelopathy (56%) and/or radiculopathy (39%). Onset of symptoms was usually insidious over months to years but 30% presented with sudden-onset neurological symptoms, often due to spontaneous haemorrhage. Mean age at diagnosis was 44 (range 2-74, SD 19.6) with a 1:1 sex ratio. Lesions were found at all levels except C1-2 but a strong predilection for the dorsal thoracic spine (68%) was shown. The lesion extended into an intravertebral foramen in at least 24 cases (34%). MRI is the investigation of choice and angiography of ESCM has rarely been described. Most (87%) were hypo- to isointense on T1 MRI while hyperintensity on T2 (91%) and avid (89%), usually homogenous gadolinium update was almost universal. Perilesional haemosiderin, characteristic of intracranial and intramedullary cavernous malformation, was rarely seen. In many cases, meningioma or nerve sheath tumour was misdiagnosed prior to surgery. All patients underwent total (95%) or subtotal (5%) microsurgical resection with excellent results; all improved (23%) or recovered fully (77%) after surgery. Those who presented acutely did worse; 38% had residual deficits.

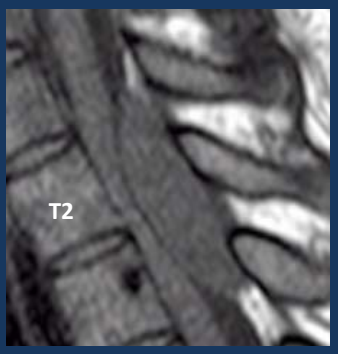




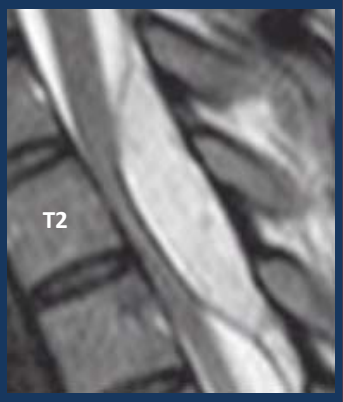
Slight blush on sagittal CT angiogram

Conclusion: Although uncommon, ESCMs are an important differential when investigating extramedullary lesions of the spinal canal because of their propensity to bleed, either spontaneously or during an unwary approach to an ESCM mimicking other pathology. Angiography may have a role in the diagnosis of ESCM and some lesions may be amenable to embolisation to definitively treat the lesion or facilitate subsequent surgery. Outcomes following microsurgical resection are good.

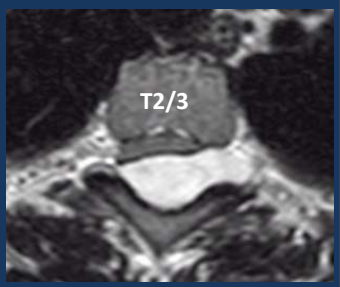
MRI features



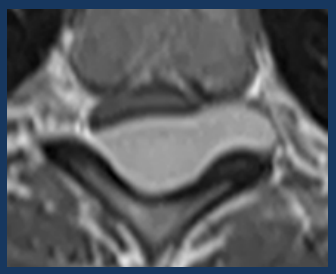
T1 weighted sagittal



T2 weighted sagittal



Left foraminal extension at T2/3



Axial T1 post Gd contrast