

Intra Medullary Capillary Hemangioma

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Citation

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Abstract

Intramedullary capillary hemangioma is a rare tumour, Capillary hemangionas are benign vascular lesion commonly encountered in the cutaneous tissue. They are rare in the nervous system. Capillary hemangiomas have been well documented as occurring in the dura and spinal nerve roots (1, 2). Only eight cases are known to have been described as occurring in the spinal cord (3). These lesions are histologically characterized by a lobular architecture. In early lesions lobules are highly cellular and composed of mitotically active plump endothelial cells(4). The lobules are surrounded by a pseudocapsule and consist of small vessels lined by single endothelial layer. The clinico-pathological features revealing extramedullary hematopoiesis and syringomyelia is highlighted. The objective of this paper is to review the capillary hemangioma and its differential diagnosis and add to the few numbers that have been published

CASE PRESENTATION

A 64 year old Caucasian male presented to our unit with a two year history of mid thoracic back pain, progressive paraparesis and paresthesia in his hands. There was associated radicular pain to both nipple areas. The back pain was constant with no relief by rest and worsening of symptoms at night. The pins and needles paresthesia with burning sensations in hands became more distressing three months prior to presentation.

His past medical history revealed he has background gouty arthritis, hypertensive cardiomyopathy and ischemic heart disease. He had a previous cardiac bypass graft. His social history revealed that he has been a pipe smoker for the last twenty years.

Clinically he had truncal weakness with preserved abdominal reflexes. He had 4/5 power in his lower limbs, hypotonia and brisk deep tendon reflexes with non-sustained ankle clonus – the plantar response were extensor bilaterally. There were patchy sensory loss to pick below C6 level with preservation of perianal sensation. His joint position sense was impaired in the left lower limb and he had significant hyperesthesia and allodynia over L5 and S1 dermatomes of the right side.

Plain X-rays of his thoracic spine showed marked anterior osteophyte at T7, T8, T10 and T12 – Magnetic resonance T1W image revealed a isodense lesion at T5 dorsally on T2W images it was slightly hyperdense.

The lesion enhanced markedly with gadolinium significant central cord changes are noted up to C5 level. (Figure 1a ,b &c).

Figure 1

Figure 1a

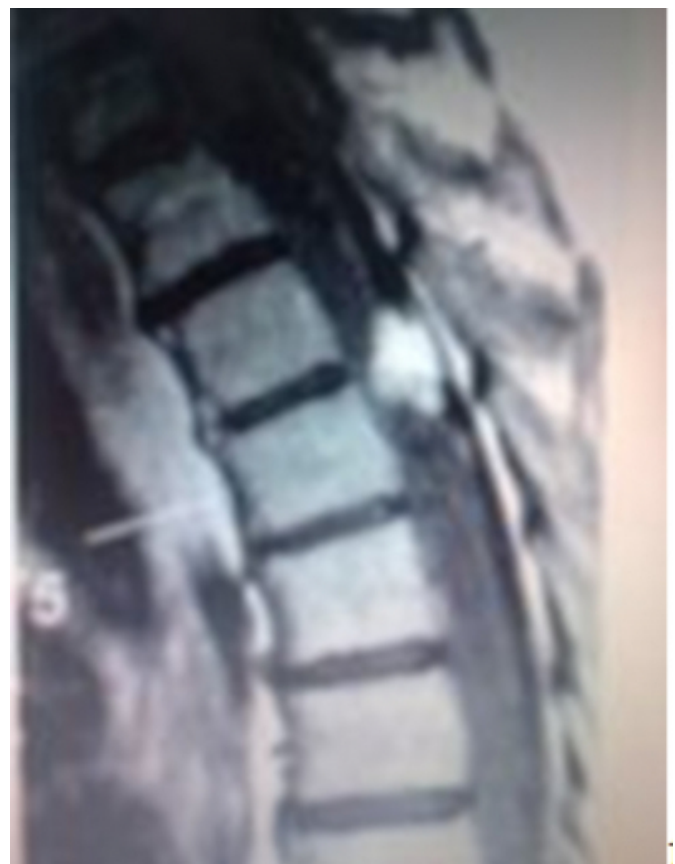


Figure 2

Figure 1b

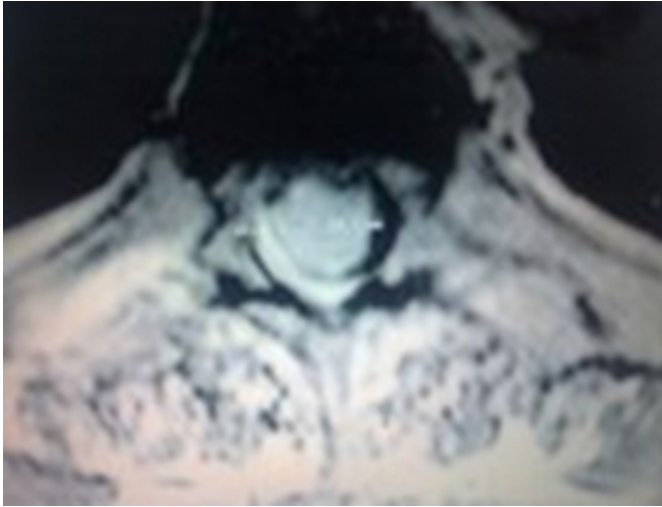


Figure 3

Figure 1c



He underwent T3,T4 and T5 laminectomy with right T4, T5 –hemi facetectomy. The reddish vascular tumour arose from the dorsal spinal cord with significant attachment to the right T5 sensory root. It had multiple arterial feeders and large drainage veins. It was removed in toto.

Post operatively his back pain and radiculopathy resolved completely. The paraparesis improved significantly. His paresthesias however remained unchanged. At 6 months follow-up the burning sensation in his legs had slightly worsened and were controlled with carbamazepine.

Histology revealed a haemorrhagic tissue. Microscopy shows homogeneous non fibrillary background with an intimate association of stromal cells and capillaries. It had a lobulated growth pattern with small capillaries and larger

feeder type vessels. Foci of extramedullary hematopoiesis were present comprising megakaryocyte and myeloid precursor forms (fig 2a ,2b).

Figure 4

Figure 2a

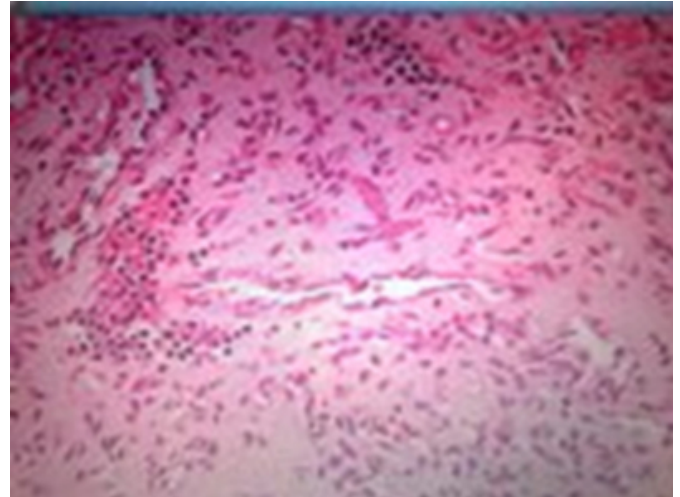
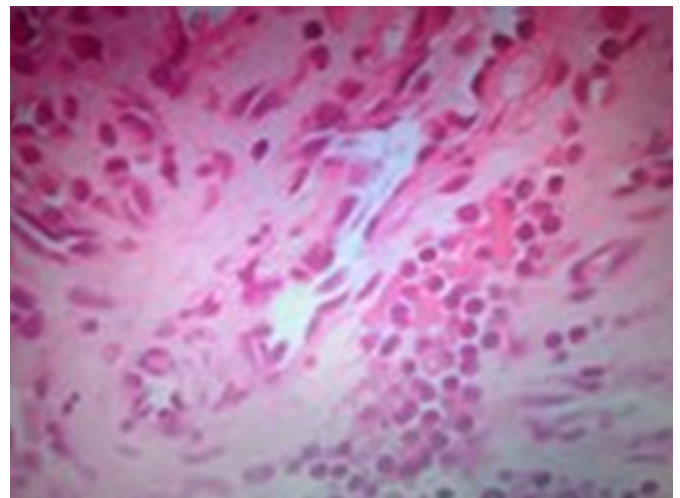


Figure 5

Figure 2b



DISCUSSION

This case illustrates an unusual cause of thoracic radiculopathy and progressive myelopathy. The duration of symptomatology is in agreement with the slow progression of the disease. The natural history of capillary hemangioma is yet to be fully documented. Capillary hemangioma unlike cavernous hemangioma do not present with acute symptoms referable to bleeding (3). They are commonly found in men older than 50 years in contrast to cavernous hemangiomas that are seen in adult woman (5). With the previous myocardial disease of this patient presenting with chest

pain, the T5 radiculopathy of the patient was initially ascribed to have arisen from his heart. The progressive myelopathy was a pointer to the presence of thoracic cord pathology. More compounding was the associated paresthesia in the patients upper limbs. The patchy sensory loss below C6 dermatome suggested a suspended sensory impairment that can be attributed to a syringomyelia or myelomalacia due to venous hypertension in the spinal cord.

Magnetic resonance imaging has greatly increased the detection of spinal cord vascular lesions. The intramedullary cavernous hemangiomas have diagnostic MRI features(6). Capillary hemangiomas in this case enhanced markedly with gadolinium. This is in agreement with those documented in the cauda equina and spinal nerve roots (1, 2)

The differential diagnosis will include hemangiopericytoma, capillary hemangioblastoma, intra medullary vascular meningioma. Intramedullary angiolipomas have been described and although they show to be hyperintense on T1W. there may be significant enhancement with gadolinium (5).

At surgery there was a distinct at plane between tumour and spinal cord. parenchyma. The lesion was subpia and attached to the sensory nerve root but easily separated from the nerve. The multiple feeding vessels and high vascularity of lesion precluded piece meal removal.

Histologic characteristics of lobular architecture with a pseudocapsule is illustrated by this case. The lobules consist of small round vessels lined by single layered endothelium. No areas of hemosiderin or calcification is found. The foci of extramedullary hematopoiesis comprising occasional megakaryocytes and myeloid precursor forms reveals an interesting pathologic entity. In the four cases described by Roncaroli et al none had this feature (3) Capillary hemangioma of the spinal nerve root have however been shown to have this feature (2). Erythroblasts as well as megakaryocytes were described (2). No mitosis was seen in the case illustration.

Capillary hemangiomas can be distinguished from other vascular lesions of the spinal cord. The hemangiopericytoma has close histologic resemblance but it has pericellular deposition of reticulum fibres and contain foamy Neuron Specific Enolase (NSE) reactive stromal cells (8).

Angiolipomas can be a close differential diagnosis. They are histologically distinguished by presence of mature adipocytes. The vascular portions of angiolipomas contains

abnormal blood vessels that vary from capillaries to sinusoidal or venular to arterial in size with irregularly thickened walls. These walls contain mildly pleomorphic endothelial and adventitial cells (7)

Capillary hemangioblastoma may show extramedullary hematopoiesis and may have features of lobularity. They however do not possess a fibrous pseudocapsule and contain vacuolated stroma cells reactive for NSE and protein S – 100 (8).

Surgical outcome was satisfactory. Spinal cord and nerve root decompression resulted in improvement of myelopathy and resolution of radiculopathy respectively residual sensory complaints are not unusual (3). At 6 months the paresthesia and burning sensation in the lower limbs required 200mg of carbamazepine to control. Financial reasons precluded a post operative MRI.

CONCLUSION

Though rare, capillary hemangiomas of the spinal cord are certainly distinct a pathological entity. The exact natural history is unknown but they certainly have a long duration of symptomatology and do leave residual changes within the spinal cord which remain symptomatic. Our case illustrates these.

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