Hemangioblastoma of the spinal cord

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Abstract A hemangioblastoma was shown at the thoracic spinal cord. Myelography showed an intramedullary lesion at the middle and lower thoracic spinal cord causing marked narrowing of the subarachnoid space and few contrast medium passing cephaladly. MR imaging revealed an intensely enhanced intramedullary nodule at T8–9 level with numerous foci of high velocity signal loss and an extensive hydrosyringomyelia of the thoracic spinal cord. *(MJS 1999 ; 2 : 80 – 84)*

บทคัดย่อ **เนื้องอกไขสันหลังชนิด hemangioblastoma**

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รายงานผู้ป่วยเนื้องอกไขสันหลังระดับทรวงอกชนิด hemangioblastoma การตรวจช่องไขสันหลังโดยสารทึบรังสี (myelogram) พบไขสันหลังระดับทรวงอกใหญ่ขึ้นมาก และการตรวจไขสันหลังโดยคลื่นแม่เหล็ก (MRI) พบลักษณะของก้อนในไขสันหลังที่ระดับกระดูกสันหลังทรวงอกที่ 8–9 และจุดไม่มีสัญญาณ (signal void) เล็กๆ หลายจุด พร้อมกับสภาวะ hydrosyringomyelia ในไขสันหลัง

INTRODUCTION

Most of the spinal intramedullary lesions are neoplasms and 90–95% of all spinal cord tumors are gliomas; ependymoma 60% and astrocytoma 35%[1] Spinal hemangioblastomas are rare (3.3% of intramedullary tumor) and the median age at onset of symptoms is 30 years[2]

Myelography, MR imaging and microscopic pictures of the thoracic intramedullary tumor are presented.

CASE REPORT

A 19 year–old woman presented with progressive paraparesis for 8 months. The physical examination showed weakness of lower extremities as muscle power grade III/V, loss of pinprick sensation below T10 level and impaired proprioception.

Myelography (Fig. 1) showed an intramedullary lesion at mid and lower thoracic spinal cord as an enlargement of the spinal cord causing marked narrowing of the subarachnoid space and few

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contrast passing cephaladly. MR imaging (Fig 2) revealed a 1.5x2.0x1.8 cm. intramedullary mass at T8–9 level which showed low signal intensity on T1W and T2W images and was markedly intense enhancement on post Gadolinium enhanced study. Numerous foci of signal loss within the lesion on all pulse sequence scans and at posterior meninges of T9 to T11 level well seen on T2W image could be high–velocity signal loss. Fluid signal intensity occupying within the spinal cord, T4–T11 level, represented hydrosyringomyelia.

The patient underwent surgery and the well circumscribed hypervascular mass at T8–9 level and an extensive hydrosyringomyelia were found. Total removal of the mass was completed and the pathological diagnosis of the mass is hemangioblastoma (Fig 3).

DISCUSSION

The spinal hemangioblastomas are intra–medullary 75%, intradural extramedullary 21.3%, intramedullary and extramedullary 10.7% and, very rare, extradural 8%[2,4] The hemangioblastomas involving the spinal cord frequently are single (78.7%) and the thoracic spinal cord is the most often involved (51.2%) followed by the cervical cord (41.2%)[2,3] 67% of the spinal intramedullary hemangioblastomas are associated with cysts as hydrosyringomyelias[2,7] Meningeal varicosities are also found in the spinal hemangioblastomas 48% of cases and are usually situated on the dorsal surface of the spinal cord.

Approximately 30% of the patients with spinal cord hemangioblastomas are associated

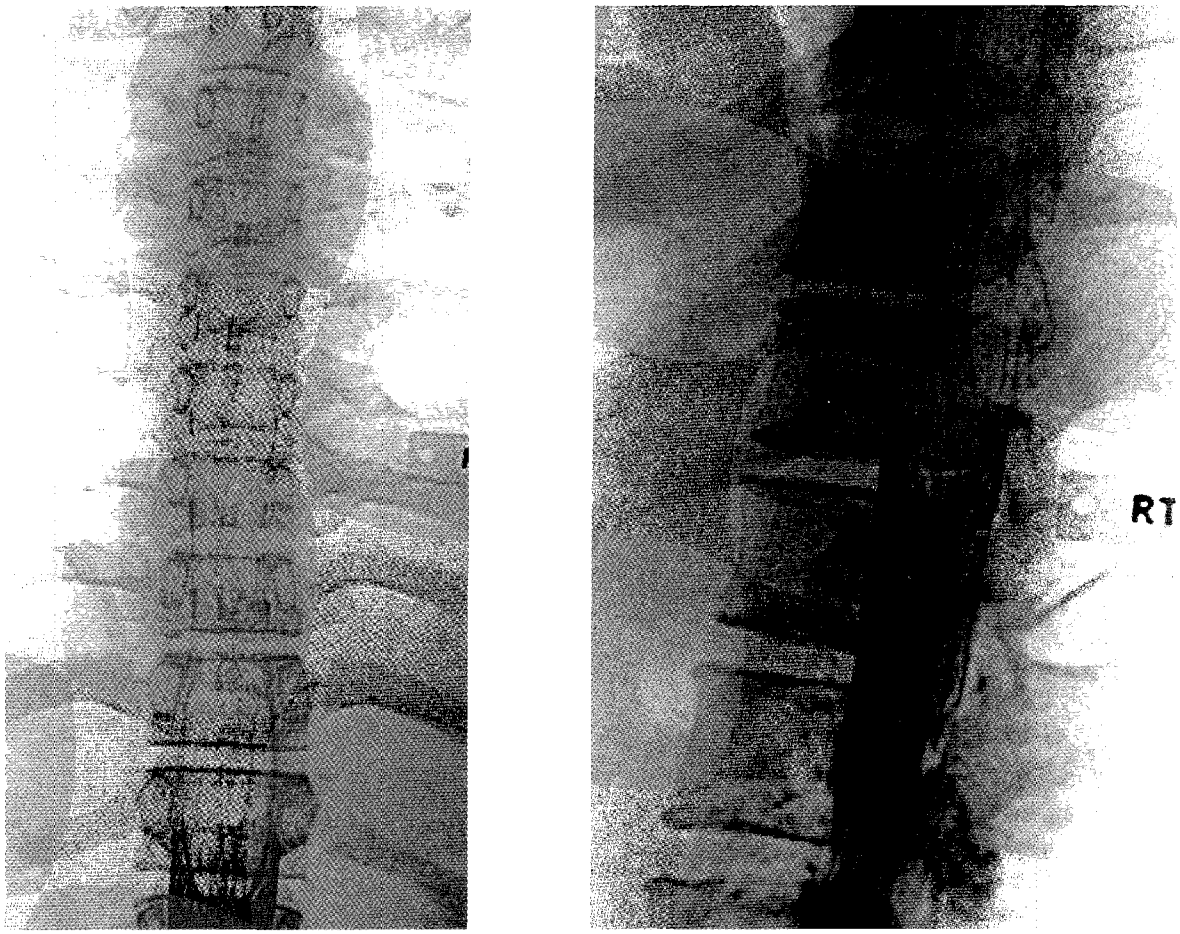
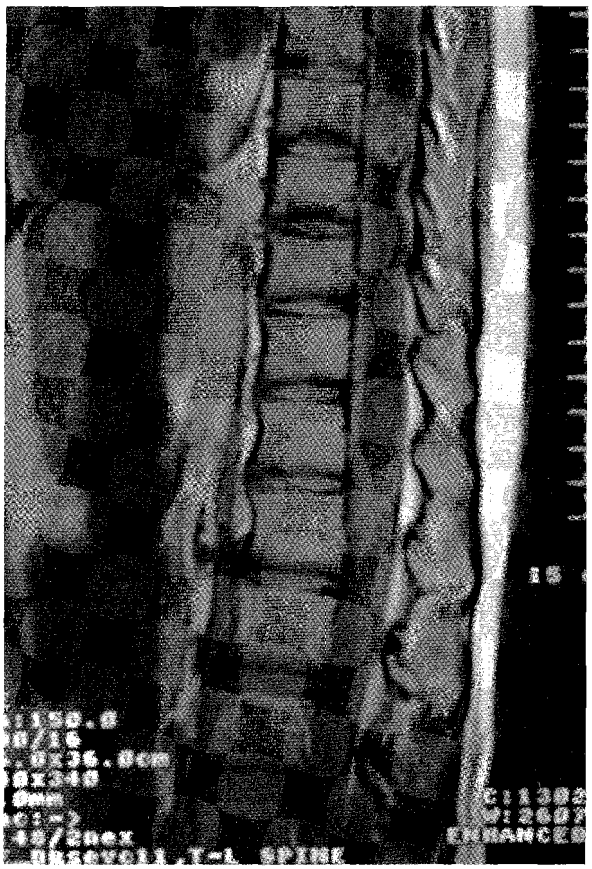


Fig 1; Myelogram shows an intramedullary lesion at mid and lower thoracic spinal cord.



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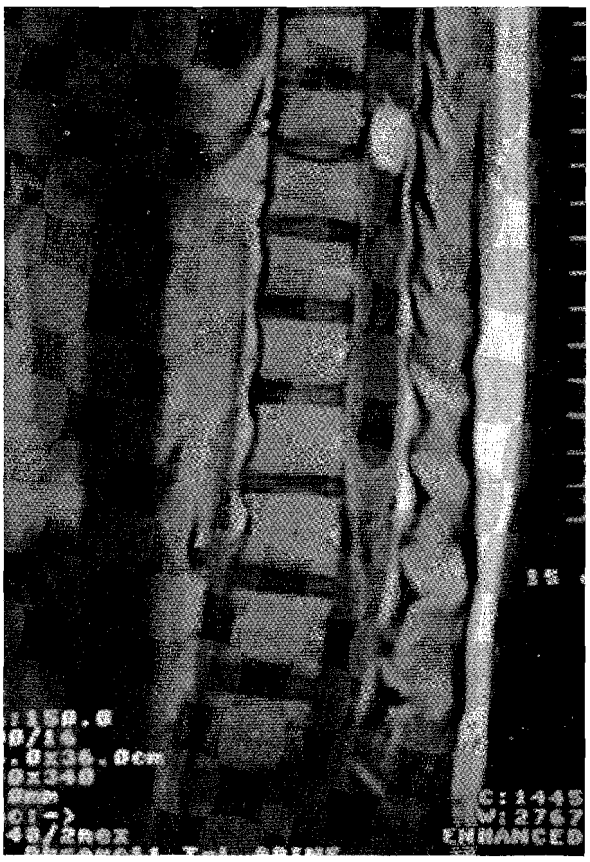

C

Fig 2; MRI reveals an intramedullary mass at T8–9 level which shows low signal on both T1W (A) and T2W (B) images and is markedly enhanced on post Gadolinium enhanced study (C). Multiple small signal void regions within the lesion and at posterior meninges of T9–T11 level (the latters well seen on T2WI) due to high–velocity signal loss. An extensive hydrosyringomyelia is seen.

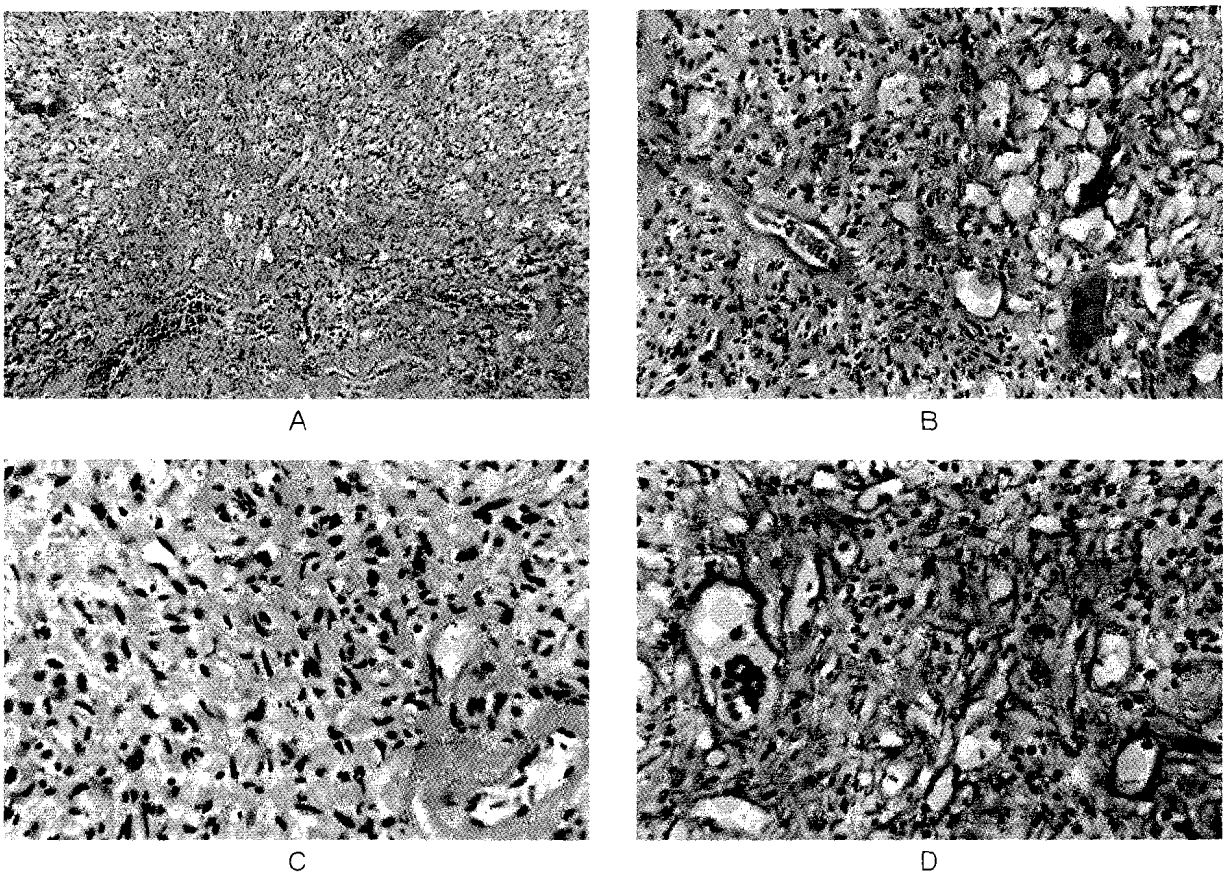

Fig 3; Microscopic pictures of the specimen. A; Solid region of the tumor containing capillary network among stromal cells. (H&E, original magnification x 25) B; Variation in cell density, compact region with blood vessels and cystic spaces. (H&E, original magnification x 75) C; Tumor tissue with stromal cells having nuclear pleomorhism and hyperchromasia, but mitoses are rare. D; Reticulin fibers were intense among the cellular tissue and around the blood vessels. (Reticulin, original magnificationx128)

with Von Hippel–Lindau syndrome. The syndrome, an autosomal dominant disorder with almost 100% penetration, is comprised of tumors at various sites in the central nervous system and abdominal viscera, including cerebellar hemangioblastomas (36–60%), retinal angiomatosis (greater than 50 %), renal cell carcinomas (25–38%), pheochromocytoma (greater than 10%) and spinal hemangioblastomas (less than 5 %)[2,3,5,6] When retinal or cerebellar hemangioblastomas coexist with spinal hemangioblastomas, they usually become symptomatic before the symptoms from the latter occur[5,6]

Grossly, the typical spinal cord hemangioblastoma has a highly vascular nodule with an extensive cyst that diffusely enlarges the cord. Prominent leptomenigeal vessels are usually present. Microscopically, spinal hemangioblastomas are characterized by dense vascular tissue that consists of thin–walled, closely packed blood vessels interspersed with large pale stromal cells[8].

Plain films of the spine are abnormal in12 out of 32 cases (37%)[2] and may demonstrate widening of the spinal canal. Myelograph frequently shows expansion of the spinal cord and serpiginous filling defects posterior to the cord representing meningeal varicosities. Spinal angiography reveals enlarged feeding arteries, densely staining tumor

nodule and rapid shunting into a distended venous plexus. MRI often demonstrates diffusion and expansion with high signal intensity T2WI and prominent foci of high-velocity signal loss. Cyst formation or syrinx is seen in 50% to 70% of cases[9] The tumor nodule in spinal cord hemangioblastoma enhances strongly following contrast administration.

Differential diagnoses are other intra-medullary tumors. The hemangioblastoma comprises a hypervascular tumor nodule as a markedly enhancing nodule on MR image, prominent leptomeningeal vessels as numerous foci of high-velocity signal loss on MR images, as well as, cyst formation or syrinx.

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