History
Teenage male with von Hippel-Lindau disease.

Diagnosis
Hemangioblastoma

Additional Clinical
Previous thoracic hemangioblastoma.

Discussion
von Hippel-Lindau is a multisystem genetic disease (autosomal dominant with high penetrance and variable expressivity) resulting in inactivation of tumor suppressor gene on chromosome 3 (p25.5). Manifestations include CNS hemangioblastomas, renal cysts, retinal hemangiomas, renal carcinoma, pheochromocytoma, epididymal cystadenomas, and pancreatic cysts.

About 1/3 of patients with spinal hemangioblastomas (sHbl) have von Hippel-Lindau disease; multiple hemangioblastomas are virtually pathognomonic of von Hippel-Lindau disease. The mean age of diagnosis of sHbl is 30 years. Symptoms of sHbl include sensory changes, motor deficits and pain. Hemangioblastomas are highly vascular tumors and may be associated with subarachnoid or intramedullary hemorrhage. Most sHbl occur in the thoracic or cervical spinal cord but may originate anywhere within the spinal canal or vertebral body. Of the intramedullary sHbl, 66% originate near the surface, 25% are completely intramedullary and 8% are exophytic. Intramedullary sHbl are associated with syrinx in 55% and cord swelling in 23%.

On MRI, sHbl are hypointense or isointense on T1, hyperintense on T2, densely enhancing and often associated with flow voids.

Findings
MR-Sagittal T1, T2 and postgadolinium T1 and axial T2 and postgadolinium T1 images of the cervical spine demonstrate two enhancing nodules (C3 and C5-C6) with intramedullary edema and cyst.

Reference
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