History
Teenager with vonHippel-Lindau disease with new painful scoliosis.

Diagnosis
Hemangioblastoma

Discussion
CNS hemangioblastoma is one of the most common manifestations of VHL disease. The cerebellum and spinal cord are the most common sites with the medulla and supratentorial region being uncommon. Only 5%–30% of all cerebellar hemangioblastomas are attributed to VHL disease, whereas 80% of spinal cord hemangioblastomas occur with the disease. Although most of these tumors (75%) are intramedullary, they may involve the dura or even be extradural. Within the spinal cord, the thoracic region is most commonly involved followed closely by the cervical cord. Most cord hemangioblastomas are solitary and slow growing. Hemangioblastomas manifest with diffuse cord expansion and variable signal intensity on T1-weighted images, with the most common appearance being isointense or hyperintense relative to the normal spinal cord. On T2-weighted images, these lesions characteristically have high signal intensity with intermixed focal flow voids. Surrounding edema and a cap sign may also be seen. Tumoral cyst and/or syringohydromyelia is very common. Focal spinal pain is the most common symptom of spinal cord hemangioblastoma, however signs of obstructive hydrocephalus and spinal cord compression may also be encountered. Treatment of CNS hemangioblastomas involves surgical resection of symptomatic tumors, with preoperative arterial embolization for extensive spinal cord tumors.

Findings
MR-Sagittal T2 and postgadolinium T1 images of the upper spine show enhancing intramedullary nodules at C3 and C5-CC6 with associated cysts and edema.

Reference
Disclaimer
This teaching site is partially funded by an educational grant from GE Healthcare and Advanced Radiology Services, PC. The material on this site is independently controlled by Advanced Radiology Services, PC, and GE Healthcare and Spectrum Health have no influence over the content of this site.

Content Download Agreement
The cases and images on this website are owned by Spectrum Health. Permission is granted (for nonprofit educational purposes) to download and print materials to distribute for the purpose of facilitating the education of health professionals. The authors retain all rights to the material and users are requested to acknowledge the source of the material.

Site Disclaimer
This site is developed to reach healthcare professionals and medical students. Nothing this site should be considered medical advice.

Only your own doctor can help you make decisions about your medical care. If you have a specific medical question or are seeking medical care, please contact your physician.

The information in this website is provided for general medical education purposes only and is not meant to substitute for the independent medical judgment of a physician relative to diagnostic and treatment options of a specific medical condition.

The viewpoints expressed in these cases are those of the authors. They do not represent an endorsement. In no event will Advanced Radiology Associates, PC, Spectrum Health Hospitals (Helen Devos Children's Hospital) or GE Healthcare be liable for any decision made or action taken in reliance upon the information provided through this website.