Multiple Spinal Hemangioblastomas Complicated with Postoperative Remote Cerebellar Hemorrhage: Review of the Literature of Two Rare Entities

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Key words
- Postoperative cerebellar hemorrhage
- Spinal hemangioblastoma
- Von Hippel-Lindau disease

Abbreviations and Acronyms
CSF: Cerebrospinal fluid
VHL: Von Hippel-Lindau

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BACKGROUND: **Multiple spinal hemangioblastomas constitute a rare pathology; most patients have von Hippel-Lindau disease. In the literature, spinal surgery has been complicated with remote intracranial hemorrhage in a very few cases.**

METHODS: **A 51-year-old woman, with no pertinent familial past medical history, presented with low back pain. Medullary magnetic resonance imaging revealed 3 spinal extramedullary tumors. The tumor at the L4 level appeared as a dumbbell-shaped neurinoma-like mass with enlarged vessels nearby. Medullary angiography excluded a dural fistula. Surgery was performed in 2 stages for all lesions. The day after lumbar surgery, the patient became comatose because of a remote cerebellar hemorrhage associated with hydrocephalus. She underwent external ventricular drainage and posterior fossa craniectomy.**

RESULTS: **The patient recovered fully at 35 days except for a slight disorientation and diplopia. Histology of all lesions revealed hemangioblastomas. Discussion of this case concerns radiologic features of spinal hemangioblastomas, diagnosis of von Hippel-Lindau disease, and pathophysiology of hemorrhagic complications following spinal surgery.**

CONCLUSIONS: **This case report includes 2 rare entities, of which every spinal surgeon should be aware.**

INTRODUCTION

Spinal nerve hemangioblastomas are uncommon benign neoplasms that occur mostly in patients with von Hippel-Lindau (VHL) disease (1). We report the case of a woman, without a pertinent familial medical history, who presented with multiple spinal hemangioblastomas. Spinal surgery of lumbar hemangioblastoma was complicated with remote cerebellar hemorrhage. We discuss the radiologic features of multiple spinal hemangioblastomas and the pathophysiology of this postoperative complication.

CASE REPORT

A 51-year-old woman, with a medical history of retinal lesion, presented with low back pain and sciatica. Absence of ankle jerk reflex and thermoanesthesia on the left side were noted. The family history did not reveal any sign of VHL disease.

Spinal cord magnetic resonance imaging revealed 3 extramedullary lesions, which resembled schwannomas (Figure 1): round T2 and L3 tumors and a dumbbell-shaped L4 mass, all hyperintense on T2-weighted images and hypointense on T1-weighted images, with massive enhancement after gadolinium administration. Enlarged vessels were observed within the lumbar dural sac, and medullary angiography was performed (Figure 2). Tumoral blush within the dumbbell-shaped mass was observed with an arterial feeder from the L4 lumbar artery. No dural fistula was diagnosed.

Surgery performed on the T2 lesion was uneventful. A few months later, a second surgery was deemed necessary for L3 and L4 tumors because the patient presented with gradually worsening low back pain and bilateral lower limb irradiation. The extracapsular en bloc removal of the L3 intradural mass was uneventful. The L4 dumbbell-shaped lesion required partial facetectomy for total resection. Abundant arterial bleeding occurred during intracapsular dissection. The arterial feeder of the tumor was coagulated and sectioned. A drain was inserted in the epidural space without negative pressure.

After awakening, the patient presented with headaches. The following day, she became comatose with a Glasgow Coma Scale score of 6. A cerebral computed tomography scan revealed a cerebellar bilateral hemispheric hemorrhage associated with acute hydrocephalus (Figure 3). Emergency external ventricular drainage and posterior fossa craniectomy were performed. The patient progressively improved and was discharged from the intensive care unit on the 35th postoperative day, with slight disorientation and diplopia. The patient had completely recovered by 6 months after surgery. Histologic examination of all lesions revealed usual features of hemangioblastoma.

DISCUSSION

Hemangioblastomas are benign tumors mainly involving the cerebellum as part of

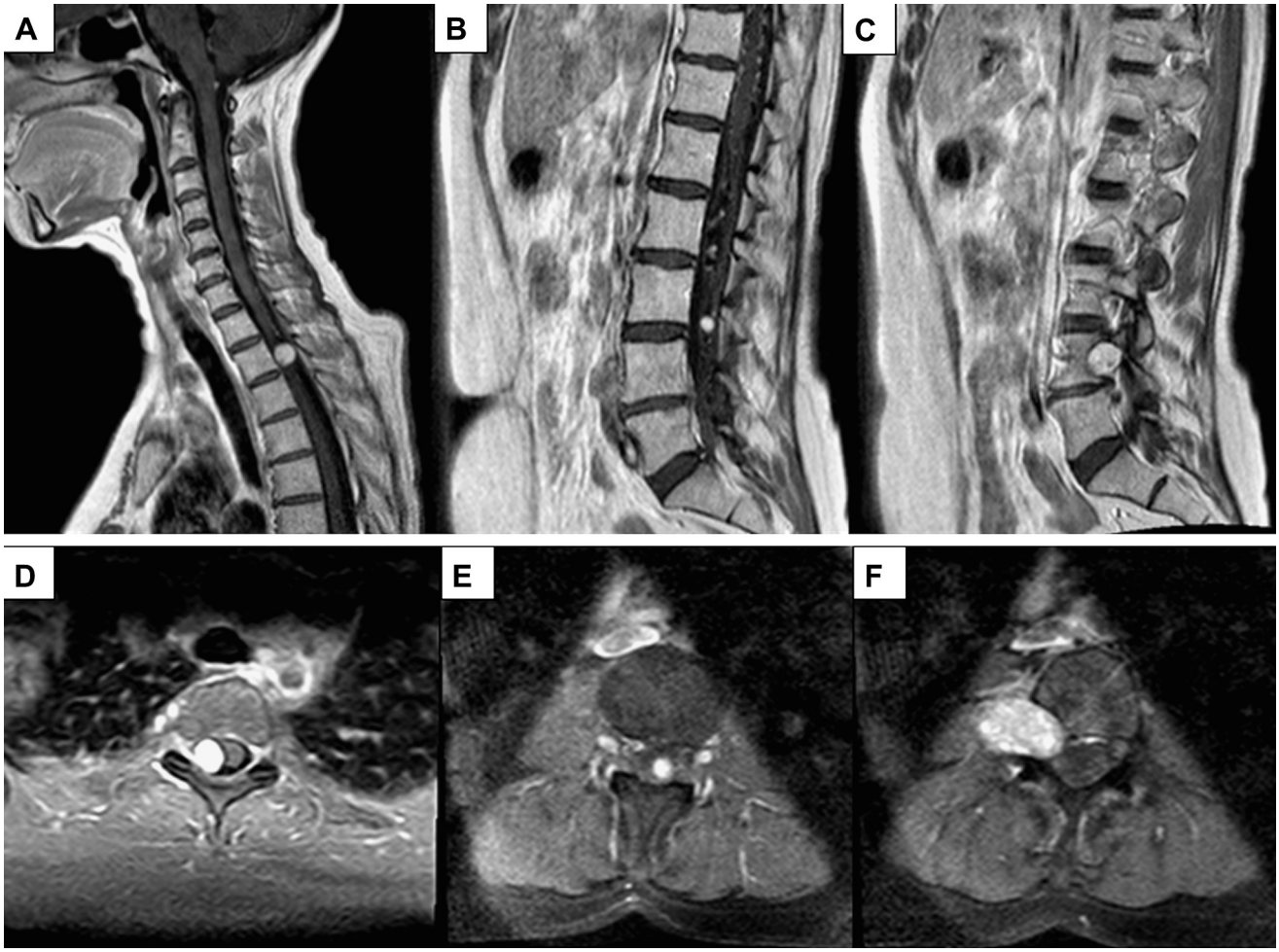

Figure 1. (**A**–**F**) Medullary magnetic resonance imaging. Sagittal and axial planes show spinal intradural extramedullary tumors at T2 (**A** and **D**) and L3 (**B** and **E**) and an intradural-extradural dumbbell-shaped mass at L4 (**C** and **F**).

VHL disease or occurring sporadically (1). There have been <20 cases of spinal nerve hemangioblastomas described in the literature (2, 12). We have reported here an exceptional case of multiple extramedullary hemangioblastomas. When a dumbbell-shaped spinal mass with contrast enhancement is encountered, the first differential diagnosis to be considered is schwannoma. However, specific radiologic features of hemangioblastomas must be sought, such as heterogeneous hyperintensity on T2-weighted magnetic resonance imaging revealing the high vascularity of the mass (5). Some dilated vessels can be observed within the canal. Angiography is helpful to assess arterial feeders and venous drainage of such tumors. Some authors have proposed embolization of the tumoral arterial pedicle before surgery (3). Multiple spinal hemangioblastomas are more prevalent in VHL disease than in sporadic cases.

Conway et al. (3) described 5 cases of spinal hemangioblastomas among 40 cases without identifying VHL disease. Our case is troublesome because no sign of VHL disease was detected, and determining whether our patient could be considered to have VHL disease was difficult. The single suspicious lesion was the retinal one, but ophthalmologic examination was unrevealing. Age at diagnosis for the present case seems to be in favor of sporadic disease. The literature mentions that ages at first surgery are younger for VHL disease than for sporadic cases, in the third decade of life versus the fourth decade (10, 11). In the absence of a family history of VHL disease, the presence of 2 central nervous system hemangioblastomas is enough for the diagnosis. A limitation of our report would be the absence of genetic screening. However, Hes et al. (7) recommended gene mutation analysis only for patients <50 years old. Woodward et al. (13) emphasized that for patients >50 years old with negative investigations, the risk of familial VHL disease remains small.

We encountered a rare cerebellar hemorrhagic complication after spinal surgery. In the literature, remote cerebellar