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A rare case of extradural hemangioblastoma in the thoracic spine in a patient with von Hippel-Lindau disease: a case report

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Introduction

Hemangioblastomas are benign tumors that occur sporadically or as part of von Hippel-Lindau (VHL) disease[3]. Hemangioblastomas account for 2-6% of all spinal cord tumors [2]. The purely extradural location is the rarest form of these tumors[3]. Contrast-enhanced Magnetic Resonance Imaging (MRI) is the gold standard for detecting and monitoring hemangioblastomas[1]. The treatment for symptomatic patients is microsurgical resection[1,3].

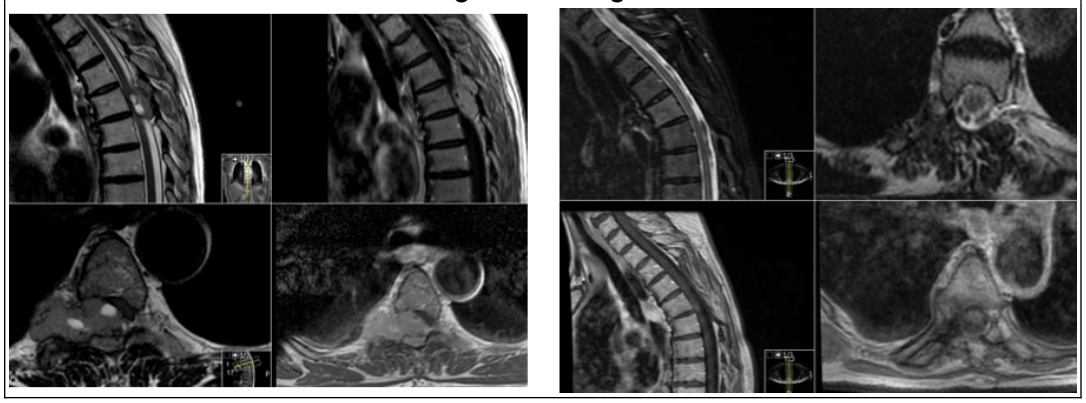
Methods

Case report of a patient with VHL with epidural tumor at the level of T5-T6.

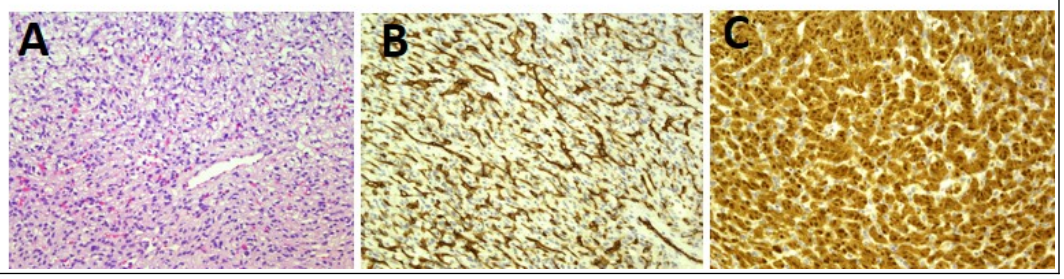
Results

Female, 58-year-old, operated on at age 36 by hemangioblastoma of the left cerebellar hemisphere, with left troclhear nerve palsy and gait ataxia. In March 2015, showed worsening of ataxia, spastic paraparesis, sphincter function impairment and a deficit of sensation at T5 level. Neurologically had a lower limb paraparesis with 4/5+ force, positive Babinski on the right and hyperreflexia of osteotendinous reflexes of the lower limbs. Imaging exams revealed epidural tumor centered on T5-T6, extending to the right rib and with medullar compression. A T4-T5 laminectomy, cost-transverse joint removal of right T5, and total epidural tumor removal were performed. The pathophysiological anatomy confirmed aspects compatible with hemangioblastoma. In Genetic consultation, DNA analysis obtained from paraffin cuts from hemangioblastoma identified the c.472C> G mutation in heterozygosity in exon 3 of the VHL gene (p.Leu158Val). Currently, he recovered motor and sensory deficit, maintaining only gait ataxia. Dorsal RM without evidence of relapse.

Fig 1. MRI images



A) Numerous thin-walled vessels, abundant vascular cells and stromal cells with the typical vacuolated morphology. B) Immunohistochemistry - evidence of numerous vessels. The stromal cells are negative for the cd34. C) Stromal cells express S100.



Conclusions

The review of literature demonstrated that extradural spinal hemangioblastoma associated with VHL is a rare diagnosis. For the first time in our practice, we observed a extradural hemangioblastoma in the thoracic spine in a patient with von Hippel-Lindau disease. And complete removal of the tumor was able to revert the neurological compromises and avoid local recurrences.

References

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