ABSTRACT

Hemangioblastomas of the central nervous system (CNS) are low-grade highly vascularized tumors that may be sporadic or associated with Von Hippel-Lindau disease. Extratrumal hemangioblastomas are uncommon and those located extra and intradurally are even rarer. This study uses an illustrative case and literature review to discuss the difficulties to consider the correct diagnosis and to select the best surgical approach. A 57 years-old white male patient presented with myelopathy and right C5 radiculopathy. The images showed a lobulated, hourglass shaped, highly enhanced extra/intratradural lesion that occupied the spinal canal and widened the C4-C5 right intervertebral foramen. Total resection of the intradural lesion was achieved through a posterior approach, but the extradural part could only be partially removed. Complete improvement was observed after four months of follow-up and the residual tumor has been followed up clinically and radiologically. Even though the preoperative impression was of a spinal schwannoma, the histopathological examination revealed grade I hemangioblastoma as per WHO. Despite their rarity, current complementary exams allow considering the diagnosis of hemangioblastoma preoperatively. That is essential to a better surgical planning in view of the particular surgical features of this lesion.

Keywords: Hemangioblastoma; Spinal cord neoplasms; von Hippel-Lindau disease; Cervical vertebrae.

RESUMO

Hemangioblastomas do sistema nervoso central são lesões de baixo grau de malignidade, altamente vascularizadas, que podem se apresentar esporadicamente ou associadas com a doença de Von Hippel-Lindau. Hemangioblastomas extradurais são incomuns e os extra e intradurais são ainda mais raros. Este estudo usa um caso ilustrativo e revisão da literatura para discutir as dificuldades de considerar o diagnóstico correto e selecionar a melhor abordagem cirúrgica. Um paciente de sexo masculino, branco, com 57 anos de idade apresentou-se com mielopatia e radiculopatia de C5 à direita. As imagens mostraram lesão extra/intradural lobulada, em forma de ampulheta, com alta impregnação após contraste, que ocupava o canal vertebral e estreitava o forame intervertebral de C4-C5 à direita. A ressecção total da lesão intradural foi alcançada através de abordagem posterior, mas a porção extradural só pôde ser parcialmente removida. Melhora total dos sintomas foi observada após quatro meses e o tumor residual tem sido seguido clínicamente e radiologicamente. Embora a impressão pré-operatória tenha sido de um schwannoma espinhal, o exame histopatológico revelou hemangioblastoma grau I, segundo a OMS. Apesar de sua raridade, exames complementares atuais permitem o correto diagnóstico pré-operatório. Isto é essencial para melhor programação cirúrgica, tendo em vista as características particulares desta lesão.

Descrições: Hemangioblastoma; Neoplasia de medula espinhal; Doença de Von Hippel-Lindau; Vértebras cervicais.

RESUMEN

Hemangioblastomas del sistema nervioso central (SNC) son tumores de bajo grado de malignidad, altamente vascularizados, que pueden presentarse esporádicamente o asociados a la enfermedad de Von Hippel-Lindau. Hemangioblastomas extradurales son poco comunes, y aquellos localizados extra e intraduralmente son aún más raros. Este estudio usa un caso ilustrativo y la revisión de la literatura para analizar las dificultades que se presentan al considerar el diagnóstico correcto y elegir la mejor abordaje quirúrgico. Un paciente, hombre blanco de 57 años de edad, presentó mielopatía con radiculopatía de C5 derecha. Las imágenes mostraban lesión extra/intradural, de forma ampolla y lobulada, la cual ocupaba el conducto espinal y ensanchaba el agujero intervertebral derecho C4-C5. La resección de la lesión intradural fue conseguida mediante un abordaje posterior, pero la parte extradural solamente pudo ser removida parcialmente. La mejoría completa fue observada después de cuatro meses de seguimiento y el tumor residual ha sido acompañada clínicamente y radiológicamente. Aunque la impresión preoperatoria era de schwannoma espinhal, el examen histopatológico reveló hemangioblastoma grado I según la Organización Mundial de la Salud. A pesar de su rara, los actuales exámenes complementarios permiten considerar, preoperatorivamente, el diagnóstico de hemangioblastoma. Esto es esencial para hacer un mejor planeamiento quirúrgico, teniendo en cuenta los aspectos quirúrgicos peculiares de esta lesión.

Descripciones: Hemangioblastoma; Neoplasias de la médula espinal; Enfermedad de von Hippel-Lindau; Vértebras cervicales.
INTRODUCTION

Haemangioblastomas of the central nervous system are low-grade highly vascular tumors that may be sporadic or associated with Von Hippel-Lindau disease. The most common location is the cerebellum and only 3 to 13% of all haemangioblastomas occur in the spine where they account for 1.6 to 5.8% of all spinal cord tumors. In the spine they are usually intramedullary. Extramedullary haemangioblastomas are uncommon. These may be intradural, extradural, or extra-intradural with a dumbbell extension, arising from the filum terminale or a proximal nerve root. In 1975, Hurlt et al. reported 17 extradural haemangioblastomas in a series of 138 spinal cases, of which only 3 cases were extra-intrudural. Only 8 cases of these tumors were found described in the literature since computed tomography (CT) scanning and magnetic resonance imaging (MRI) have become available.

The aim of this study is to discuss the difficulties to determine the correct preoperative diagnosis of these lesions and its importance on deciding the best treatment strategy based on a case report and literature review.

ILLUSTRATIVE CASE

A 57 years-old white male presented with a 6 months history of neck pain and 1 month of increasing weakness of the superior limbs, followed by lower left limb paresis, bowel and bladder disturbance. His additional medical history was significant only for hypertension. Physical examination demonstrated myelopathy and C5 radiculopathy characterized by triparesis that was worse in the upper limbs with preserved strength in the lower right limb, increased tone and Babinski signal in his lower extremities as well as deep hyporeflexy, though right bicipital hyporeflexy. On spine CT scans (Figure 1A) a marked widening of the right C4-C5 intervertebral foramen was noted, as well as facets erosion and no hyperostosis. The MRI (Figure 1B and 2A,B,C) showed a lobulated, dumbbell shaped, extra-intradural lesion that was markedly enhanced after gadolinium injection. The lesion occupied the spinal canal and the C4-C5 intervertebral foramens compressing and displacing the spinal cord medially and posteriorly. The extra-spinal component came into contact with the vertebral artery, which was slightly displaced medially and anteriorly. Flow-void signals were retrospectively observed inside and around the tumoral mass, suggesting a high-vascularized tumor. Schwannoma and malignant tumors were considered as preoperative diagnosis.

The patient was placed in a right lateral position with the head fixed in the Mayfield and aligned with the vertebral spine. (Figure 3A,B) A straight medial occipito-cervical incision was performed followed by a C3-C4 laminectomy and right C4-C5 foraminotomy and "T" shape durotomy directed to the lesion entry point at the intradural space. The entry point was coagulated and cut to permit the total resection of the intradural lesion after coagulation of the dura was performed. A venous aneurysm and was supplied by anomalous vessels. (B) Total resection of the intradural lesion could be achieved, but the extradural part was removed partially. The lateral dural opening was occluded with a muscle patch and sutured to the dura. The histopathological examination revealed an haemangioblastoma - WHO grade I (Figure 4A,B,C,D,F). The patient improved completely and was already asymptomatic 4 month after surgery. Postoperative MRI showed remaining lesion extradurally, that has been followed clinically and radiologically for 1 year already. Investigation for Von Hippel-Lindau disease was performed and resulted negative. The brain and the rest of the spinal canal were scanned and no additional tumoral lesion was found.

DISCUSSION

In these rare cases of extra-intrudural haemangioblastomas, enlargement of an intervertebral foramen and a dumbbell shape wrongly suggest the preoperative hypothesis of nerve root schwannoma or neurofibroma. Nevertheless, schwannomas that grow with an extension to the intervertebral foramen are not common, and there are some other different kinds of tumors that may present with this location. Besides these facts, extra-intrudural hemangioblastomas are extremely rare. However, with the diagnostic tools nowadays available, even the diagnosis of such an uncommon lesion is possible. On MRI sequences, solid portions of haemangioblastomas generally have high-intensity signal on T2-weighted images, intermediate or low-intensity signal on T1-weighted sequences, and marked enhancement with gadolinium. Features suggesting...
REFERENCES


