

EXTRA AND INTRADURAL SPINAL HEMANGIOBLASTOMA

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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. Extradural 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/intradural 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 do 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 clínica e radiologicamente. Embora a impressão pré-operatória tenha sido de um schwannoma espinal, 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.

Descritores: Hemangioblastoma; Neoplasia de medula espinal; Doença de von Hippel-Lindau; Vértebras cervicais.

RESUMEN

Hemangioblastomas del sistema nervioso central (SNC) son tumores altamente vascularizados, de grado bajo, que pueden ser esporádicos o vinculados a la enfermedad de Von Hippel-Lindau. Hemangioblastomas extradurales no son 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 cuanto a considerar el diagnóstico correcto y para seleccionar el mejor abordaje quirúrgico. Un paciente, hombre blanco de 57 años de edad, presentaba mielopatía con radiculopatía C5 derecha. Las imágenes mostraban lesión extra/intradural, muy acrecentada, con forma de 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 acompañado clínica y radiológicamente. Aunque la impresión preoperatoria era de schwannoma espinal, el examen histopatológico reveló hemangioblastoma grado I según la Organización Mundial de la Salud. A pesar de su rareza, los actuales exámenes complementarios permiten considerar, preoperativamente, el diagnóstico de hemangioblastoma. Esto es esencial para hacer un mejor planeamiento quirúrgico, teniendo en cuenta los aspectos quirúrgicos peculiares de esta lesión.

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

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INTRODUCTION

Haemangioblastomas of the central nervous system are low-grade highly vascular tumors that may be sporadic or associated with Von Hippel-Lindau disease¹⁻³. The most common location is the cerebellum² 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 intramedullary³⁻⁶. 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^{3,4,7-9}. In 1975, Hurth et al. reported 17 extradural haemangioblastomas in a series of 138 spinal cases, of which only 3 cases were extra-intradural. 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^{1,3,10-15}.

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 and bladder disturbance. His additional medical history was significant only for hypertension. Physical examination demonstrated myelopathy and C5 radiculopathy characterized by tri paresis 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 hyperreflexy, 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 foramen 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. The surgical finding consisted of a firm and high-vascularized extradural lesion that made the intervertebral

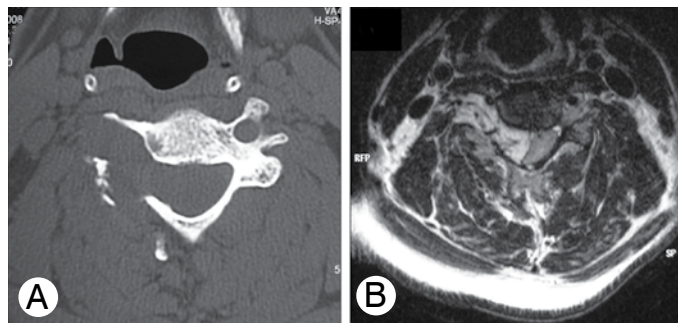


Figure 1. Spine CT (A) showing a marked widening of the right C4-C5 intervertebral foramen and erosion of facets. Axial T1-weighted MRI after gadolinium injection (B) showing a dumbbell shaped extra-intradural lesion occupying the intervertebral forame and displacing the spinal cord to the opposite site and posteriorly.

foramen and the C4 joint surface very thin and crossed the dura at the level of C4. The intradural lesion was soft, resembling a sponge, attached to the dura and supplied by anomalous vessels. Total resection of the intradural part of the 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.

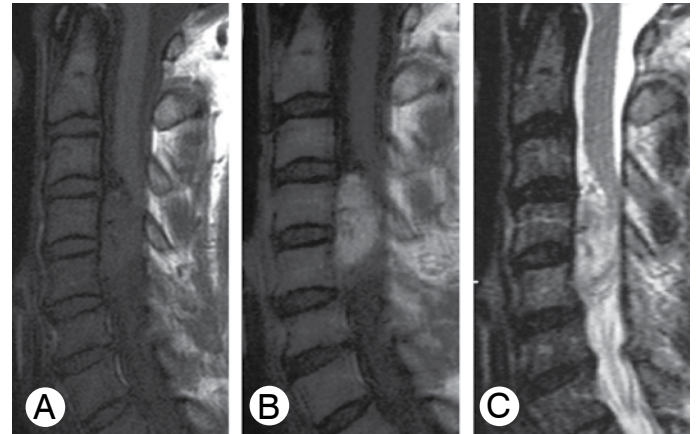


Figure 2. MRI sagittal images showing a tumoral lesion at C4-C5 level isointense to the spinal cord at T1-weighted (A), with marked enhancement after gadolinium injection (B) and with heterogeneous signal on T2 weighted (C). Low signals in T2-weighted are compatible with enlarged vessels. The lesion is located extradurally and displaces the spinal cord posteriorly.

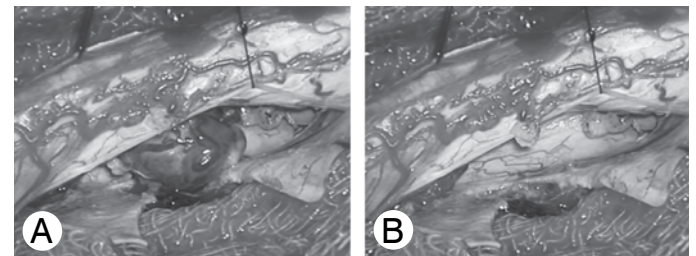


Figure 3. After C3-C4 laminectomy, (A) "T" shape durotomy directed to the point where the lesion crossed the dura is shown. The right dentate ligament was sectioned and pulled to the left side. This part of the tumor resembled a venous aneurysm and was supplied by anomalous vessels. (B) Total resection of the intradural lesion after coagulation of the dura was performed.

DISCUSSION

In these rare cases of extra-intradural haemangioblastomas, enlargement of an intervertebral foramen and a dumbbell shape wrongly suggest the preoperative hypothesis of nerve root schwannoma or neurofibroma¹. Nevertheless, schwannomas that grow with an extension to the intervertebral foramen are not common¹⁶; and there are some other different kinds of tumors that may present with this location. Besides these facts, extra-intradural hemanioblastomas are extremely rare^{1,9,10,13}. 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^{5,7,10,17-19}. Features suggesting

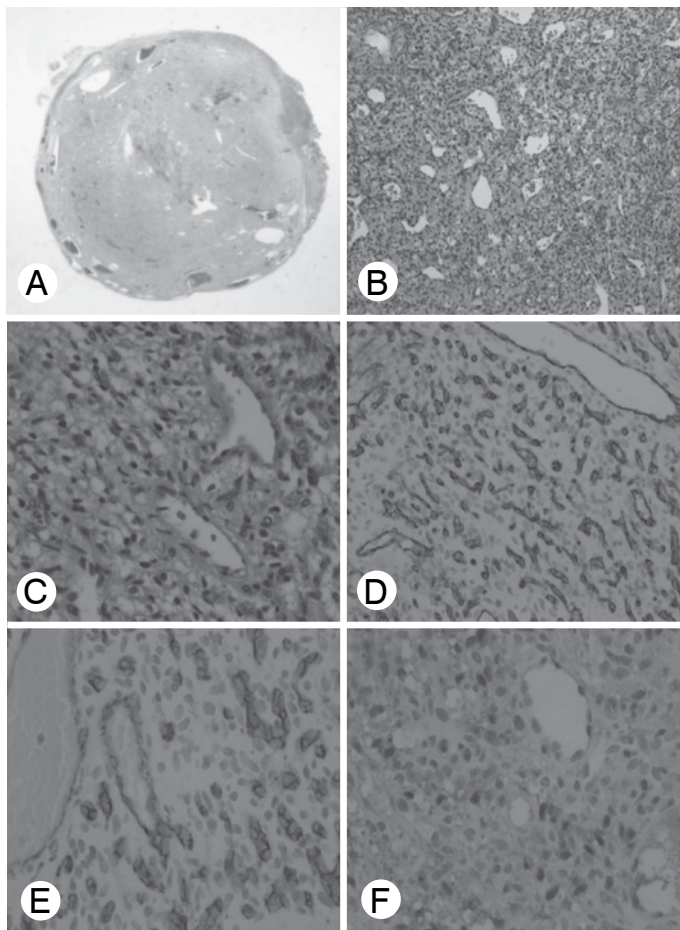


Figure 4 - Histopathological and immunohistochemical features confirming the diagnosis of haemangioblastoma – WHO grade I. (A) Well-delimited, non-encapsulated neoplasia (0,5x – H&E). (B) Irregular vascular channels (40X – H&E). (C) Neoplastic cells with clear cytoplasm and vacuolized aspect (100X – H&E). (D) and (E) Endothelial cells showing immunopositivity to CD 31 (100X) and CD34 (200X). (F) Focal immunopositivity of the neoplastic cells for the S100 antigen (200X).

vessels, such as flow-void images, are often visible, especially in large tumors⁵. Angiography confirms the diagnosis of a highly vascularized tumor⁴, thus eliminating the possibility of an arteriovenous malformation¹⁷. The key to preoperative diagnosis in these tumors is their marked enhancement and associated enlarged vessels²⁰. Surgery is the treatment of choice for symptomatic haemangioblastomas^{1,4,10}, but cervical dumbbell lesions raise the problems of radical resection, vertebral artery control, nerve root preservation, and spine stability¹.

An anterior approach alone through the vertebral body is not adequate, because it cannot reach the extra spinal and foraminal part of the tumor. Barrey et al.¹ defend a lateral approach because it allows control of the distal nerve root and the vertebral artery before resection of the tumor and permits reaching even the intradural part as was showed in their case report. In that case the vertebral artery was firstly tested and sacrificed during surgery, as well as the nerve root, which stimulation resulted no response.

A posterior approach with laminectomy and unilateral facetectomy has been proposed for dumbbell neuromas²¹ and was chosen for the present case. The good visualization of the intradural space made it possible to resect the whole intradural part without causing additional injury to the spinal cord. However, the hard visualization of the vertebral artery and no control of the blood supplying vessels before the tumor resection made it impossible to perform a radical resection. Another disadvantage of this approach is that a joint must always be sacrificed¹. The residual lesion next to the vertebral artery may cause gradual occlusion of this vessel and the possibility of its sacrifice in a likely reoperation.

If the posterior surgical approach is chosen, preoperative embolization or even vertebral artery occlusion may be considered after balloon occlusion test in cases of haemangioblastomas to decrease tumoral blood supply and facilitate surgical resection¹⁴. In lateral approaches, the vertebral artery will be visible before reaching the tumor, and the vascular control could be performed at the surgical act¹, although balloon occlusion testing before surgery is mandatory.

CONSIDERACIONES FINALES

Despite their rarity, current complementary exams allow considering the diagnosis of haemangioblastoma preoperatively. That is essential to a better surgical setup as these lesions have a great propensity for bleeding.

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