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Surgical Management of Triple Sporadic Lumbar Intradural Hemangioblastomas with Atypical Imaging Presentations: A Case Report in a Patient without VHL Syndrome

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Abstract

Introduction and Importance: Hemangioblastomas of the cauda equina are particularly exceptional. Therefore, the management has not yet been established. This study reports on a very rare case of three hemangioblastomas arising from the cauda equina with a heterogeneous appearance of each lesion on MRI. This appearance is atypical and requires some reflection on the type of management. We chose to operate on the two lower lesions in order to obtain histology and treat symptoms by being as minimally invasive as possible. The upper lesion is under radio-clinical monitoring. **Presentation of Case:** A 72-year-old patient without Von Hippel-Lindau (VHL) disease presented a 1-year history of low back pain and 6 months of bilateral S1 radicular symptoms. MRI revealed three lumbar intradural extramedullary tumors with different types of contrast enhancement. Preoperative angiography rules out vascular malformation nests. The other results of the work-up are negative. Following multidisciplinary discussion, only two lower lesions were removed in order to have anatomopathological pieces, be less invasive, and try to heal leg symptoms. Tumors are well limited and hypervascular. The post-operative course is completely satisfactory, with complete resolution of neurological symptoms and no perioperative complications. The third lesion, the highest, is being monitored and will be treated on the basis of radio-clinical findings in the future. **Clinical Discussion:** This article reports a very rare case of three hemangioblastomas with different contrast enhancements. It shows that three extramedullary intradural hemangioblastomas on cauda equina are possible. Two of these were operated on, and the third is under observation. **Conclusion:** Surgical management of only part of the lesions is entirely feasible in the first instance in order

to obtain histology, remain minimally invasive, and limit perioperative complications.

Keywords

Case Report, Hemangioblastoma, Cauda Equina, Intradural Lesion, Surgery

1. Introduction and Importance

Highlights

- This article reports that three sporadic hemangioblastomas can occur in a non-VHL patient.
- Surgery to remove symptomatic lesions may be proposed in order to obtain histology and alleviate symptoms.
- The remaining lesion(s) should be monitored radio-clinically. In the event of progression, further surgery or radiosurgery may be proposed.

Hemangioblastoma is a rare, benign, and vascularized tumor of the nervous system (Tucer et al., 2013). This lesion accounts for 2% - 6% of spinal cord tumors (Chen et al., 2019). Up to 85% are found in the cerebellum, and between 3% and 13% are detected in the spinal cord (Delisle et al., 2000). Hemangioblastomas intradural extramedullary are even rare. They may be single or multiple. They are often found in the context of Von Hippel-Lindau (VHL) disease (40%) (Sun et al., 2012). Surgical removal is often the main treatment for a single lesion (Martins et al., 2019). Generally, only symptomatic or enlarging hemangioblastomas are surgically resected (D'Oria et al., 2022). Some studies (da Costa et al., 2003; Wolbers, Ponssen, & Kamphorst, 1985) report on the management of one or two lumbar hemangioblastomas, but none refer to three lesions. This article describes the management of three lumbar intradural hemangioblastomas with an atypical contrast that may suggest other etiologies. These lesions appeared in a 72-year-old man without VHL disease. This case report has been reported in line with the SCARE Criteria (Sohrabi et al., 2023).

2. Presentation of Case

The patient is 72 years old. Informed consent was carried out from the patient to report this case. He has no medical history except an ablation of a benign colorectal polyp 1 year before lumbar lesions were discovered. He has no notion of VHL disease in his family. The spinal history began approximately 1 year ago, with a progressive onset of inflammatory low-back pain, predominantly in sitting positions. Walking reduced the intensity of his lower back pain. Neuropathic pain is located in the S1 territory, focused on both calves, without motor deficiency. Neuropathic pain is bearable but has been worsening for 4 months. Osteotendinous reflexes are all well-perceived and symmetrical. There are no signs of pyramidal irritation or sphincter disorders. In this context, his general practitioner ordered

a lumbar MRI (Figure 1). Remnological imaging revealed three lumbar intradural extramedullary masses in contrast, but in a heterogeneous way. The mass in front of S1 is the largest, measuring 18 × 12 mm with homogeneous contrast. The second is facing L5 on the left and measures 5 × 5 mm, and the third is in front of L2L3 on the right and measures 10 mm. The second and third lesions show different contrasts from the first, with only peripheral enhancement. The literature shows few studies in this context, with only three lesions of this type, which may suggest a number of causes, especially neuromas (D'Oria et al., 2022). First, it was decided to do a cerebral MRI and a medullar MRI in order to search for other lesions, but none were found. Furthermore, the thoraco-abdomino-pelvic CT scan shows no primitive tumor. An arteriography was performed and ruled out a vascular etiology. A PET was also carried out and was negative. The multidisciplinary meeting proposes a surgical biopsy or removal of the two lower lumbar lesions due to their different visual appearance. The purpose of the meeting decision is to obtain histology while being as minimally invasive as possible, to reduce neurological symptoms possibly related to the thickest lesion S1 and minimize breach complications. Depending on the histology and subsequent clinical course, a second, less invasive, distant surgery may be proposed. Thus, the L2L3 lesion is not explored by surgery.

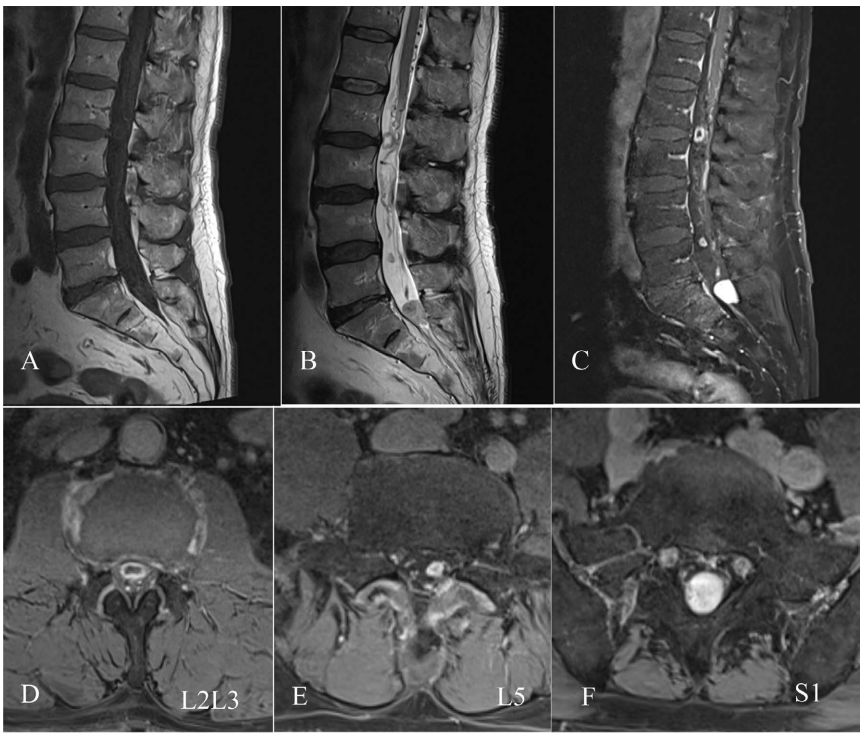

Figure 1. MRI showing three intradural tumors. Lesions appear as an iso-intense area on sagittal T1-weighted imaging (A). Two lower lesions are enhanced with gadolinium, with a low intensity on sagittal T2-weighted imaging (B). The largest tumor, S1, shows strong gadolinium homogeneous enhancement, while the two smallest tumors are not contrast-enhanced at their centers (C). An axial view of the 3 lesions is shown with gadolinium on slices (D), (E) and (F).

Surgery involves a complete laminectomy of L4, L5, and S1, followed by the opening of the dura mater. No intraoperative electrophysiological monitoring is performed. Macroscopically, both lesions show a well-limited nodular lesion that is richly vascularized and organized into several anastomosing capillary structures. Masses can be completely removed after loosening adhesions to the nerve roots and dissecting of the feeder vessel. Closing is achieved with dura mater overjet, without enlargement plasty, using Tachosil® and biological glue. The cells are epithelioid or spindle-shaped. The stroma is fibrous, hyaline, and non-inflammatory. No signs of malignancy are found. On immunohistochemistry, cells express PS100 in the absence of GFAP, EMA, and SSTR2A expression, PAX8. CD34 underlines the abundant vascularization of the lesion. Inhibin is focally expressed. The anatomopathological results of both lesions are consistent with hemangioblastoma.

The post-operative course is satisfactory, with complete resolution of neurological symptoms. A disabling inflammatory low-back pain appeared on the tenth day and was completely resolved by the fifteenth with anti-inflammatory medication. The 1-month check-up showed no abnormalities. The patient has regained full autonomy. At 6 months, the patient has no particular complaints. The MRI is satisfactory and the L2L3 lesion is stable on the imaging slices (**Figure 2**). Then, the MRI monitoring classically used in the department is every year for 3 years and then every 5 years.

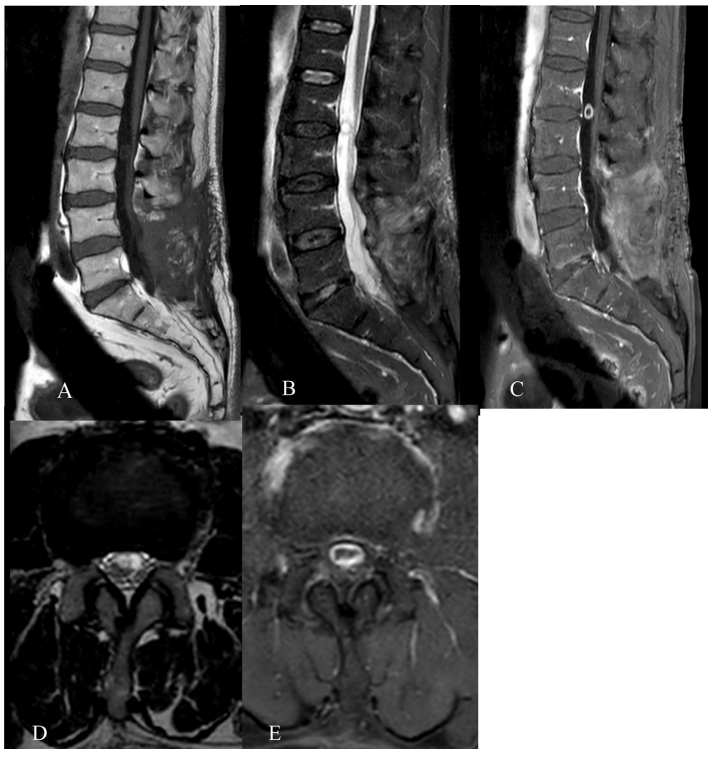

Figure 2. Post-operative MRI with sagittal T1-weighted (A), sagittal T2-weighted (B), T1-gadolinium (C) with axial view of L2L3 lesion axial T2-weighted (D) and T1-gadolinium (E).

3. Discussion

This case describes a patient without VHL with three lumbar intradural extramedullary hemangioblastomas. No studies have reported on three lumbar hemangioblastomas, so there are no guidelines for their management. This study also shows a radiological variant. Normally, hemangioblastomas are strongly gadolinium-enhanced and very homogeneous due to their hypervascularization (Evzikov et al., 2021). In this case, only the large S1 lesion meets this criterion. The other two showed atypia with a hyposignal center on sequences with injection. Obtaining histology is of prime importance in this work, as the heterogeneity of contrast cannot rule out other tumor types. Two of the three lesions are operated on, with this in mind in order to remain minimally invasive, treat symptoms, and limit perioperative complications. The third is under radio-clinical observation until it grows up or becomes symptomatic. This decision may change depending on the histology. Multiple locations in the spinal cauda equina are even rarer, so there is no consensus on management (Blaty et al., 2018). The main treatment in most studies, such as those by Fuji (Fujii et al., 2022) and Maire (Maire, Husag, & Probst, 1978), is surgical treatment but this management often concerns single lesion. Few articles refer to two lesions, and care remains varied (Han, Zhang, & Jia, 2024; Li et al., 2021). None involves three lesions. In other spinal lesions such as neuromas, multiple lesions are often all operated on at once, as the treatment is surgical resection. Management in this case must differ from this surgical standard, given that the lesions are atypical and could also be intracanal metastases, and treatment is therefore more onco-radiotherapeutic than surgical (Sohrabi et al., 2023). Therefore, this article suggests combining surgical treatment with radio-clinical monitoring. Other authors, such as Pan (Pan et al., 2018) or Chang (Chang et al., 2011), propose radiosurgery as an alternative solution, even though this innovative method has yet to show any real efficacy. This method seems to have gained prominence in recent years (Kim et al., 2009). Increasingly, this method is being used for single lesions whose histology is known. Results appear to be satisfactory, with few recurrences. This method is now used for lesions of increasing volume. Given that histology is already known in this case, radiosurgery could be tried on the third lesion if it becomes symptomatic or enlarges on follow-up imaging. This would avoid the need for further surgery in several years' time, given the considerable risks involved for an elderly patient. Otherwise, Biondi (Biondi et al., 2005) shows embolization can reduce post-operative complications, especially blood loss. However, this method is not compulsory, although it is used in several works (D'Oria et al., 2022). In fact, pre-operative embolization was not performed on this patient, and blood loss was only 100 mL. As in this work, cases reported in the literature often show very good improvement of symptoms with surgery (Blaty et al., 2018). Symptoms improved markedly in around 50% of cases and persisted in 25%. Neurological symptoms improved in 25% of patients who underwent surgery. Surgical resection of lumbar hemangioblastoma is not so safe and effective, and patients should be informed that their clinical condition may remain

unchanged or worsen in 50% of cases (Fujii et al., 2022; Marchesini, Ricci, & Pinna, 2021). The purpose of this article is to show that surgical treatment of symptomatic lesions, with the monitoring of others, is entirely feasible. These results will obviously need to be assessed in the medium and long term.

4. Conclusion

Lumbar hemangioblastomas are very rare. Therefore, management remains debated. Several care alternatives have been described for single lesions, whether symptomatic or not. Generally, surgery is the main therapy. This article suggests that the treatment of multiple hemangioblastomas, in particular over two lesions, can be based on a combination of several options. Surgery is the first line of treatment, enabling histology to be obtained and symptoms to be treated. It should be as minimally invasive as possible, avoiding the removal of lesions located in other areas. Surgery can be supplemented by radio-clinical follow-up of smaller lesions. With the arrival of radiosurgery in this pathology, irradiation could be envisaged on the lesions that have not been operated on, in order to avoid further invasive surgery.

Declarations

Informed consent: Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

All data are available in the patient's medical file.

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All authors approved the final version of the manuscript.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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