Acute Paraparesis due to Aggressive Vertebral Hemangioma. Report of Two Cases and Literature Review

Matías Pereira Duarte, Gastón Camino Wilhuber, Gonzalo Kido, Julio Bassani, Matías Petracchi, Carlos Solá, Marcelo Gruenberg

Adult Spinal Pathology Sector, Orthopedics and Traumatology Service, Hospital Italiano de Buenos Aires (Buenos Aires, Argentina)

ABSTRACT

Introduction: Aggressive hemangiomas make up 1% of all vertebral hemangiomas. They can produce pain, fractures, deformity and slowly progressive neurological compromise. Different treatment approaches have been described, but optimal management remains unclear. We report two cases of acute paraparesis secondary to aggressive thoracic hemangioma, we describe their treatment and evolution. We carry out a narrative review of the literature on vertebral hemangiomas. **Conclusion:** Through early decompression and arthrodesis followed by local adjuvant radiotherapy, we obtained complete neurological recovery and disease control in a medium-term follow-up.

Keywords: Aggressive hemangioma; benign tumor; spine; radiotherapy; paraparesis.

Level of Evidence: IV

Paraparesia aguda por hemangioma vertebral agresivo. Reporte de dos casos y revisión bibliográfica

RESUMEN

Introducción: Los hemangiomas agresivos constituyen el 1% del total de los hemangiomas vertebrales. Pueden producir dolor, fracturas, deformidad y compromiso neurológico, generalmente de larga evolución. Se han descrito diferentes opciones terapéuticas, pero el manejo óptimo sigue sin estar claro. Comunicamos dos casos de paraparesia aguda secundaria a un hemangioma torácico agresivo, describimos su tratamiento y evolución. Realizamos una revisión narrativa de la bibliografía. **Conclusión:** Mediante la descompresión y la artrodesis tempranas seguidas de radioterapia, se logró la recuperación neurológica completa y el control de la enfermedad en un seguimiento a mediano plazo.

Palabras clave: Hemangioma agresivo; tumor benigno; columna vertebral; radioterapia; paraparesia.

Nivel de Evidencia: IV

INTRODUCTION

Vertebral hemangiomas (VH) are the most frequently diagnosed benign tumors in the axial skeleton, they have an incidence of 10-12% between the fourth and fifth decades of life.[1,2] Histologically, they are made up of vascular spaces lined with endothelium separated by bone stromal septa.[3,4] In general, they are asymptomatic and quiescent, most are discovered incidentally when performing an MRI indicated for other reasons. They mainly affect the vertebral body and, rarely, spread to the posterior components.[5]

On imaging studies, "typical" VHs exhibit a characteristic pattern, lithic foci with trabeculae reminiscent of a honeycomb with large and long vertical streaks on plain radiography. In the computed tomography, irregularly distributed punctate calcifications are observed, product of the coalescence of the vertical trabeculae described. Magnetic resonance imaging (MRI) highlights a hyperintense image in both T1 and T2 sequences (due to the predominance of fat content), as well as in fluid-sensitive sequences, STIR, due to vascular components.[5-7] However,

Received on July 19th, 2020. Accepted after evaluation on September 2nd, 2020 • MATÍAS PEREIRA DUARTE, MD • matiaspereiraduarte@gmail.com http://orcid.org/0000-0001-5652-2631

How to cite this article: Pereira Duarte M, Camino Willhuber G, Kido G, Bassani J, Petracchi M, Solá C, Gruenberg M. Acute Paraparesis due to Aggressive Vertebral Hemangioma. Report of Two Cases and Literature Review. *Rev Asoc Argent Ortop Traumatol* 2021; 86 (3): 398-406. https://doi.org/10.15417/issn.1852-7434.2021.86.3.1161

VHs may not show these distinctive characteristics when there is a lower fat content and more vascular content. These are called "atypical" VHs.[5]

The "aggressive" VHs are the third category of these lesions that—despite being benign from the histological point of view—have a locally aggressive behavior, corresponding to grade 3 of the Enneking classification.[8] They are rare, representing 1% of all VHs,[9] and can cause symptoms, such as pain in 54% of cases, pathological fractures, spinal deformity and neurological compromise in up to 40% of cases.[10-13] They are most frequently located in the thoracic spine and the male: female ratio is 1: 1.5.[14]

The aim of this article is to present two cases of acute paraparesis due to aggressive thoracic hemangiomas and to conduct a narrative review of the literature on the subject.

CLINICAL CASE 1

A 22-year-old male patient who consulted for progressive loss of strength in the lower limbs associated with back pain secondary to a fall from his own height, seven days before. On physical examination, he presented 2/5 muscle strength for muscle groups innervated by L2, L3, L4, L5, and S1; hypoesthesia below the umbilical area; and hyper-reactive patellar and Achilles reflexes. The score on the Nurick scale[15] was 5 and on the modified Japanese Orthopedic Association scale[16] (mJOA) was 9/18. A Frankel C incomplete spinal cord syndrome with severe paraparesis was diagnosed.

The radiographic analysis showed an alteration of the bone tissue in the vertebral body of T9 (Figure 1). A computed tomography scan was performed, in which the classic punctate pattern of hemangiomas was not observed, which made the initial diagnosis difficult. The MRI showed an isointense image in the T1 and T2 sequences with extravertebral extension and involvement of the medullary canal, while, in the STIR sequence, the lesion was evidently hyperintense (Figure 2).

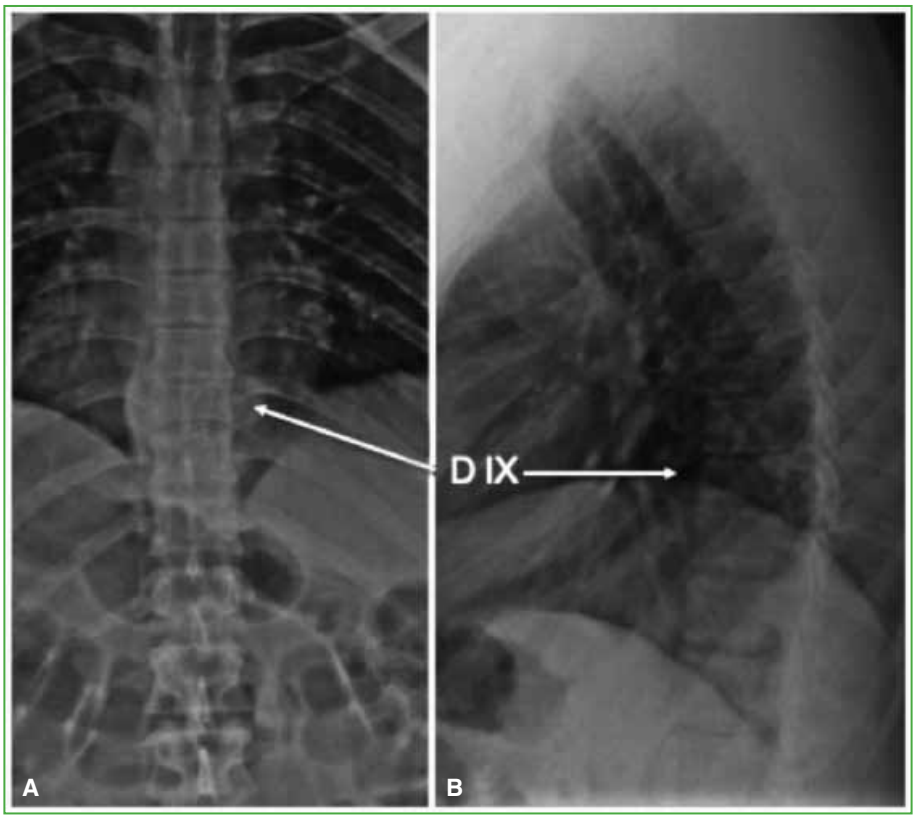

Figure 1. Case 1. A 22-year-old man. Anteroposterior (**A**) and lateral (**B**) radiographs of the thoracic spine. An alteration in the bone tissue at the T9 level and a slight collapse of the vertebral body are observed.

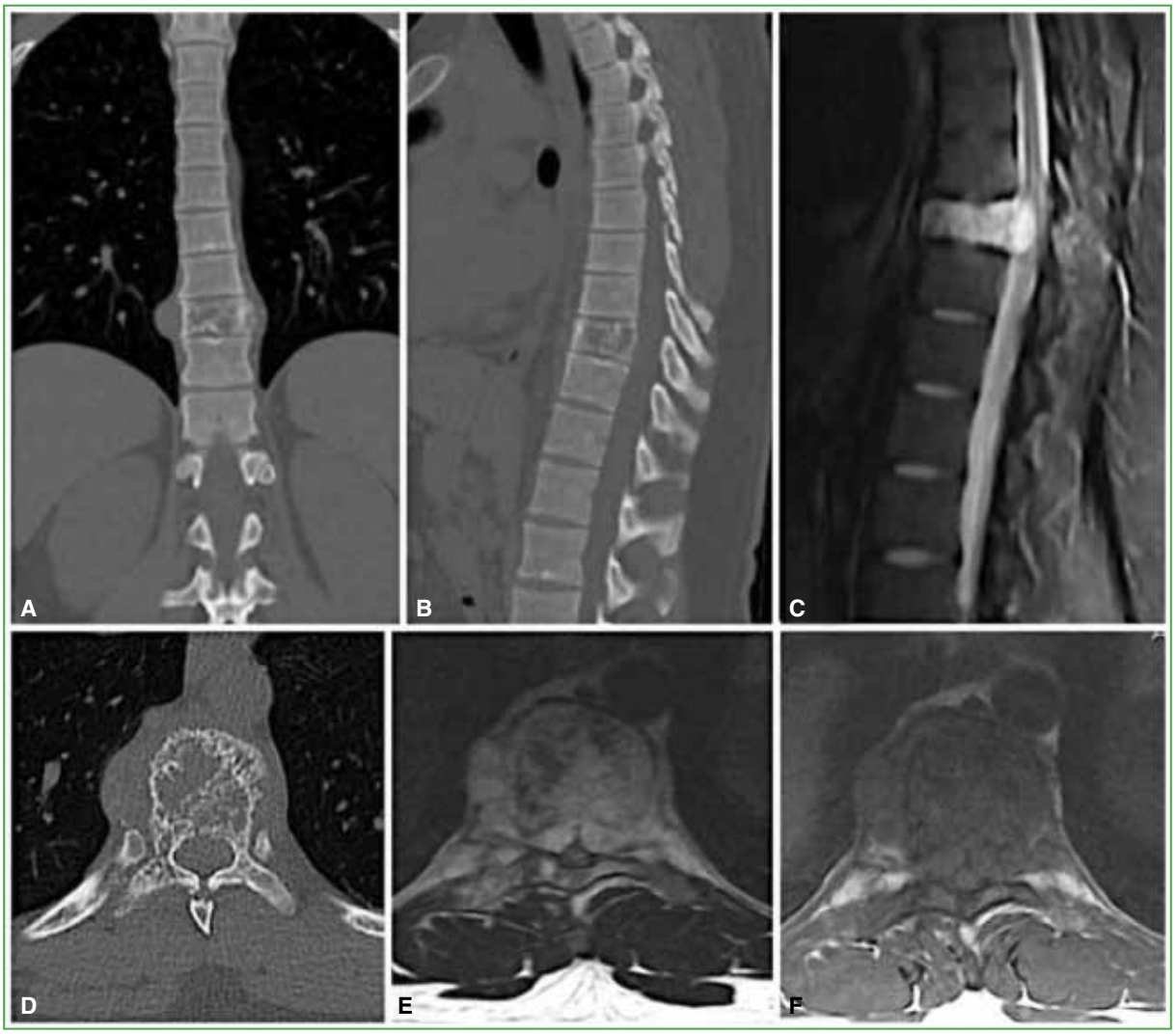

Figure 2. Case 1. A 22-year-old man. Computed tomography of the thoraco-lumbar spine. **A.** Coronal plane, **B.** Sagittal plane, **C.** Axial plane. Heterogeneous lytic lesions with a permeative pattern are observed, altering the bone tissue of the vertebral body of T9 with compromise of the cortices and extending to the paravertebral and epidural spaces. The compromise of both pedicles is evidenced, mostly on the right side, with involvement of the pars and ipsilateral facets. Magnetic resonance imaging of the dorsal spine. **C.** Sagittal plane, STIR sequence. The hyperintensity of the lesion that affects the entire vertebral body is highlighted, generating a pathological fracture, with compromise of the posterior wall and invasion of the medullary canal. **E.** T1-weighted sequence, **F.** T2-weighted sequence. Isointense images show a greater right paravertebral extension.

Due to the acute and progressive neurological condition, a decompression was performed by means of a T9 laminectomy and an emergency arthrodesis of T7-T11 via the posterior approach (Figure 3). Samples were sent for histopathological analysis of the lesion, which informed the diagnosis of VH (Figure 4).

Five weeks after surgery, the patient began radiation treatment with a total dose of 4000 cGy divided into sessions of 150-200 cGy, every 24-48 h, according to tolerance, with the aim of preventing recurrences.

At 36 months after the intervention, after intensive progressive rehabilitation, the patient presented neurological improvement, with scores of 17/18 on the mJOA, Nurick 3 and Frankel E scale, with no signs of tumor recurrence.

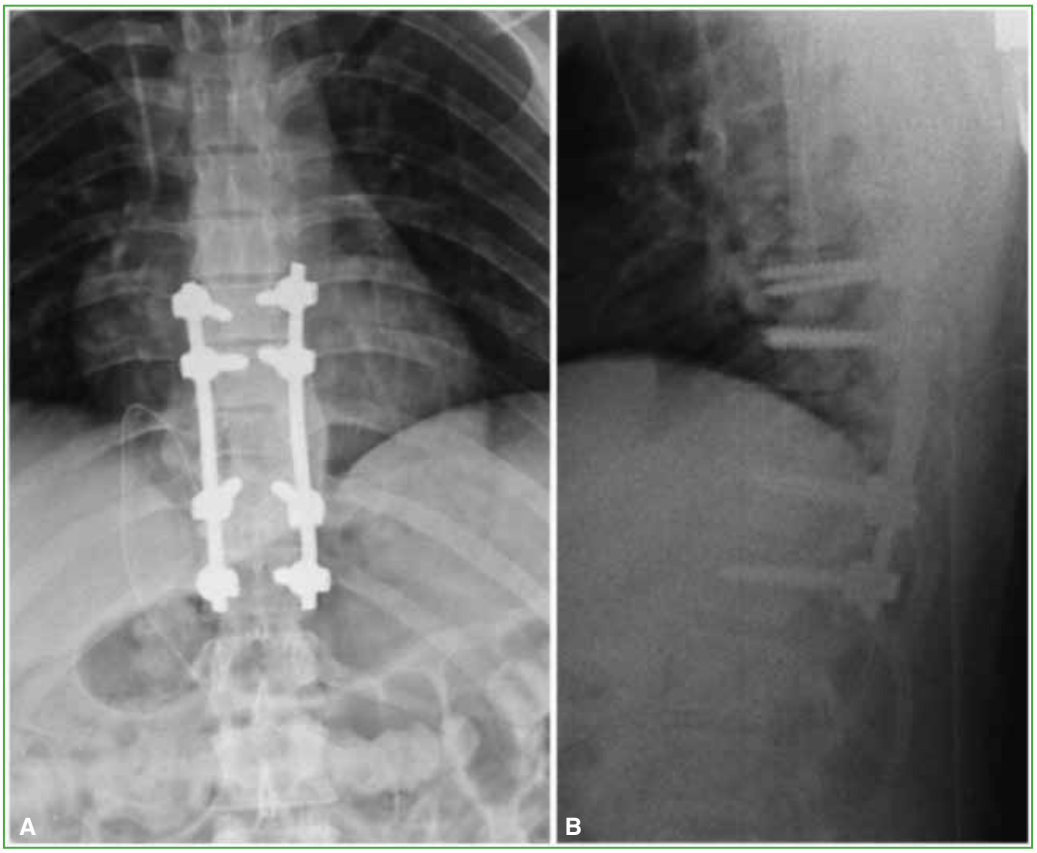

Figure 3. Case 1. A 22-year-old man. Postoperative anteroposterior (**A**) and lateral (**B**) radiographs of the thoracic spine. Pedicle screw fixation from T7 to T11 is visualized.

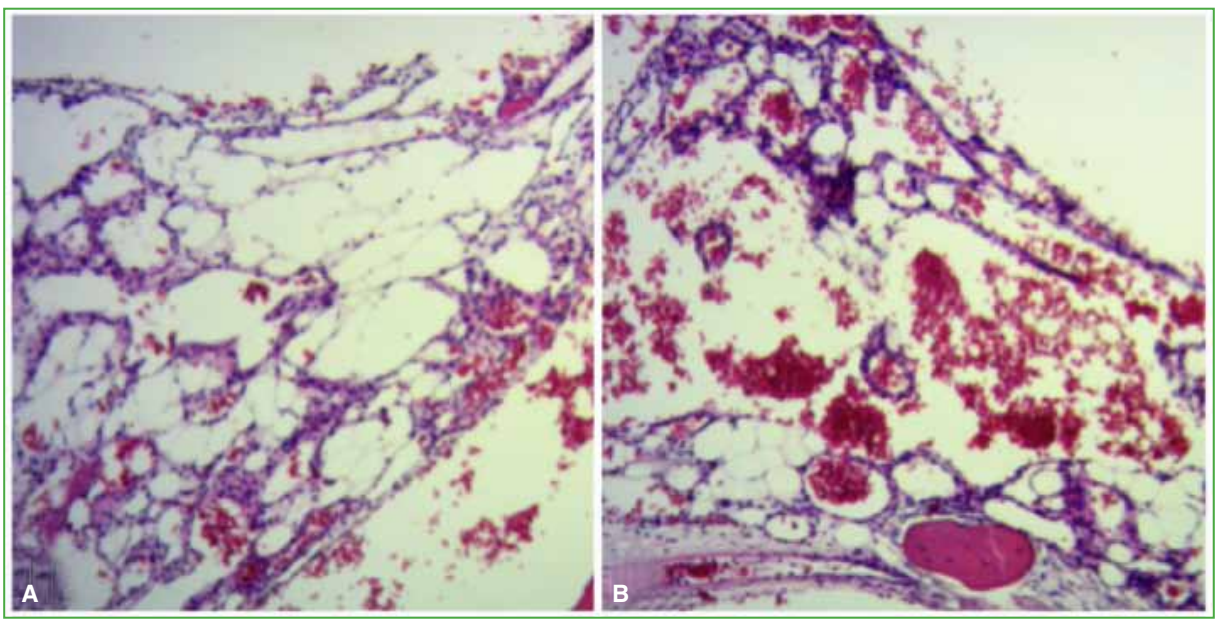

Figure 4. Case 1. A 22-year-old man. Histological analysis under x20 (**A**) and x40 (**B**) microscopy and hematoxylin-eosin staining showed the lining of the spaces created within the collagenous matrix stroma by thin layers of flat endothelial cells that form capillary-sized cavernous blood vessels, consistent with the diagnosis of hemangioma.

CLINICAL CASE 2

A 23-year-old female patient who consulted at the emergency room for progressive back pain and paraparesis of 24 h of evolution, with no relevant traumatic history. As a relevant antecedent, the patient reported previous episodes of back pain, with imaging studies and vertebral biopsy puncture compatible with VH, treated conservatively. On physical examination, she presented 3/5 decreased muscle strength from L2 to distal, bilateral patellar hyperreflexia, and positive Babinski. Neurological scores were 4/5 for Nurik, 14/18 for mJOA, and Frankel C. An emergency MRI revealed complete involvement of the vertebral body of T11, with altered bone structure, extension through the right pedicle towards the posterior apophyseal ring and invasion of the medullary canal (Figure 5). Given the previous diagnosis of VH, a selective embolization of the nutrient artery of the lesion was performed, followed by decompression of T10-T12 and arthrodesis of T9-L1 via the posterior approach (Figure 6). Four weeks after surgery, she began radiation therapy at a dose of 4500 cGy. After 60 months of follow-up, the patient was pain-free, and her scores were Frankel E, Nurik 1, and 17/18 on the mJOA scale. No signs of recurrence of the lesion were observed.

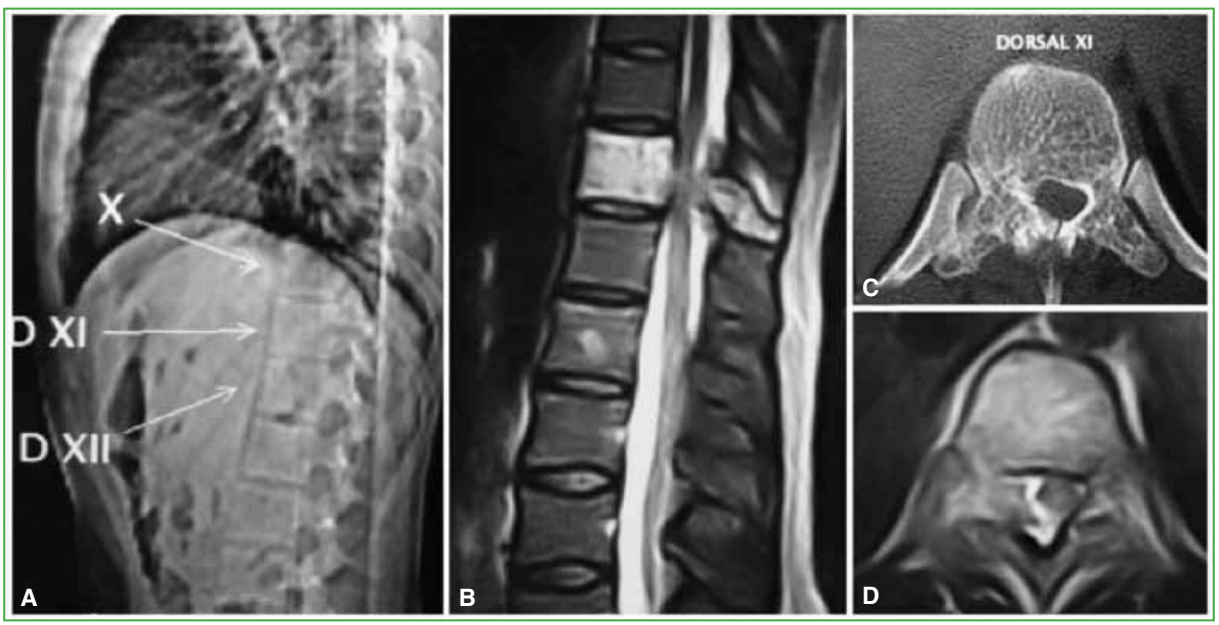

Figure 5. Case 2. A 23-year-old woman. **A.** Lateral radiograph of the thoracic spine with marking of the level involved without relevant findings. MRI. **B.** Sagittal plane, T2-weighted sequence. Hyperintensity of the vertebral body is observed, with extension towards the right pedicle and invasion of the medullary canal, displacing the marrow to the left (**D**). **C.** Computed tomography, axial plane. The alteration of the bone structure in the vertebral body is observed, with insufflation of the right components of the apophyseal ring.

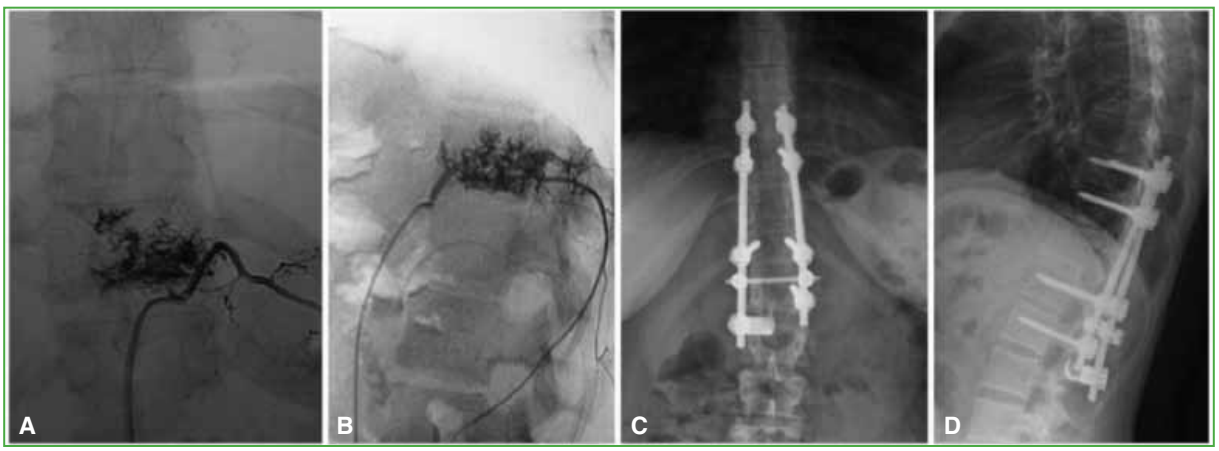

Figure 6. Case 2. A 23-year-old woman. Anteroposterior (**A**) and lateral (**B**) fluoroscopy images of the dorsal spine lesion at the T11 level during the selective arterial embolization procedure of the nutrient artery. Anteroposterior (**C**) and lateral (**D**) radiographs of the thoracic spine at 5 years of follow-up. Decompression and dorsolumbar arthrodesis from T9 to L1 are visualized.

DISCUSSION

We have described two cases of aggressive dorsal hemangioma with severe acute paraparesis that required urgent surgical intervention. Aggressive VH represent less than 1% of the total, can cause pain, pathological fractures and neurological compromise. Most aggressive hemangiomas show a typical polka-dot sign[17] or honeycomb pattern on images; however, in up to a third of them, the patterns may be atypical.[18] Acosta et al.[19] describe six peculiarities in imaging studies, which are associated with "symptomatic" or "aggressive" VH: 1) thoracic location (particularly T3-9); 2) involvement of the entire vertebral body; 3) involvement of the neural arch (particularly pedicles); 4) an irregular appearance without a honeycomb shape, 5) an expanded, insufflated and poorly defined cortex and 6) involvement of the adjacent paravertebral soft tissue. Diagnosing "atypical" and "aggressive" VHs by images can be difficult, regardless of the imaging modality used, as they can mimic malignant bone tumors or primary metastases.[5] Our two patients presented all these atypical characteristics, which made diagnosis by imaging difficult. Therefore, it was necessary to resort to the pathological anatomy of the lesion for an accurate diagnosis.

Optimal treatment strategies and adjuvant therapy options remain controversial, as large randomized studies are difficult to conduct due to the low prevalence of this condition.[1,19,20] Different options have been described, such as intralesional ethanol injection,[21] arterial embolization,[22] vertebroplasty[23] or kyphoplasty,[24] total *en bloc* spondylectomy[19] or partial spondylectomy plus stabilization,[25,26] radiotherapy[27,28] or a combination of them. Kaoudi et al.[29] have recently described a robot-assisted radiofrequency thermal ablation in an aggressive hemangioma of the sacrum, with good results at one year of follow-up.

Preoperative intra-arterial embolization offers the advantages of controlling bleeding and reducing perioperative morbidity.[30] In some cases, slowing or growth arrest of the lesion has been observed when performed in isolation.[3,31] In the first case presented, embolization was not considered due to the lack of a specific diagnosis and the urgency of the condition. However, in the second case, there was prior histological confirmation, so it was possible to perform an emergency embolization followed by decompression surgery and arthrodesis.

The use of adjuvant radiation therapy after intralesional excision surgeries with the aim of reducing the recurrence rate is controversial. While some authors have reported good long-term results in series of patients treated in this way,[1,11,32] Qiu et al.[20] reported having carried out radiotherapy in only three of 10 patients, and they also did not detect recurrences at the average 11-month follow-up, so the real benefit of this treatment is questioned.

Both of our patients received radiotherapy at the fourth and fifth week after surgery. This delay reduces the rate of wound complications, such as dehiscence and infection. No postoperative complications were detected and both patients had a progressive and complete neurological recovery without signs of tumor recurrence at the end of follow-up.

We described two cases of a rare pathology with an infrequent presentation. There are few reports of acute spinal syndromes due to aggressive hemangiomas, most of which are cases of slow progression over time. In a retrospective analysis, Qiu et al.[20] reported 10 cases of aggressive hemangiomas over a 13-year period, five of them with progressive neurological deterioration and one Frankel D at the time of treatment. Our patients presented with a Frankel C developed in a short period. We obtained good results after emergency decompression and stabilization surgery followed by radiation treatment. It is worth noting that the success of this type of surgery depends on the speed of detection, diagnosis and treatment, since the delay can cause a less marked or absent neurological improvement.

A postoperative recurrence rate of 0% to 30% has been published,[20] regardless of treatment with or without adjuvant radiation therapy. It is worth highlighting the particular aggressiveness of this injury in pregnant women, in whom the recurrence rate is greater than 20%.[19,33] This is the product of the influence of gestational progesterone on the tumor, which would also be related to the higher incidence in women.[14]

According to our search, these are the first two cases published in the Latin American literature on acute neurological compromise in young patients caused by aggressive VH. We believe that it is important to be aware of this diagnosis, its typical and atypical images, as well as its forms of treatment in order to provide our patients with the best results. At present, there is no consensus on the optimal treatment and each case must be particularly evaluated taking into account all the factors listed above.

CONCLUSION

We presented two cases of severe acute paraparesis secondary to aggressive thoracic VH. By means of early decompression and arthrodesis followed by radiation therapy, a complete neurological recovery and disease control were achieved in a medium-term follow-up.

Conflict of interests: The authors declare they do not have any conflict of interests.

G. Camino Willhuber ORCID ID: http://orcid.org/0000-0002-5684-7679
G. Kido ORCID ID: http://orcid.org/0000-0001-5521-7918
J. Bassani ORCID ID: http://orcid.org/0000-0002-3340-8606
M. Petracchi ORCID ID: http://orcid.org/0000-0001-7350-1816
C. Solá ORCID ID: http://orcid.org/0000-0001-9606-2107
M. Gruenberg ORCID ID: http://orcid.org/0000-0001-7681-7058

REFERENCES

1. Fox MW, Onofrio BM. The natural history and management of symptomatic and asymptomatic vertebral hemangiomas. *J Neurosurg* 1993;(7881):36-45. https://doi.org/10.3171/jns.1993.78.1.0036
2. Huvos AG. Hemangioma, lymphangioma, angiomatosis/lymphangiomatosis, glomus tumor. En: Huvos AG (ed.). *Bone tumors: diagnosis, treatment, and prognosis*. 2nd ed. Philadelphia: Saunders; 1991:553-78.
3. Rudnick J, Stern M. Symptomatic thoracic vertebral hemangioma: a case report and literature review. *Arch Phys Med Rehabil* 2004;85(9):1544-47. https://doi.org/10.1016/j.apmr.2003.08.099
4. Cross JJ, Antoun NM, Laing RJ, Xuereb J. Imaging of compressive vertebral haemangiomas. *Eur Radiol* 2000;10(6):997-1002. https://doi.org/10.1007/s003300051051
5. Gaudino S, Martucci M, Colantonio R, Lozupone E, Visconti E, Leone A, et al. A systematic approach to vertebral hemangioma. *Skeletal Radiol* 2015;44(1):25-36. https://doi.org/10.1007/s00256-014-2035-y
6. Orrison W. *Neuroimaging*. Philadelphia: W. B. Saunders Co; 2000:1335-6.
7. Osborn A. *Diagnostic neuroradiology*. St. Louis, MO: Mosby; 1994:877-9.

8. Enneking WF. Musculoskeletal tumor staging: 1988 update. *Cancer Treat Res* 1989;44:39-49. https://doi.org/10.1007/978-1-4613-1757-9_3
9. Healy M, Rerz DA, Pearl L. Spinal hemangiomas. *Neurosurgery* 1985;13(6):689-91. https://doi.org/10.1227/00006123-198312000-00013
10. Templin CR, Stambough JB, Stambough JL. Acute spinal cord compression caused by vertebral hemangiomas. *Spine J* 2004;4(5):595-600. https://doi.org/10.1016/j.spinee.2003.08.034
11. Castel E, Lazennec JY, Chiras J, Enkaoua E, Saillant G. Acute spinal cord compression due to intraspinal bleeding from a vertebral hemangioma: two case-reports. *Eur Spine J* 1999;8(3):244-8. https://doi.org/:10.1007/s005860050167
12. Ahn H, Jhaveri S, Yee A, Finkelstein J. Lumbar vertebral hemangioma causing cauda equina syndrome: a case report. *Spine (Phila Pa 1976)* 2005;30(21):662-4. https://doi.org/10.1097/01.brs.0000184560.78192.f6
13. Nguyen JP, Djinjian M, Pavlovitch JM, Badiane S. Vertebral hemangioma with neurologic signs: therapeutic results. *Neurochirurgie* 1989;35(5):299-303, 305-8. PMID: 2630927
14. Campanacci M. Hemangioma. En: Campanacci M (ed.). *Bone and soft tissue tumors*. Padova: Piccin Nuova Libraria &Wien: Springer; 1999:599-618.
15. Nurick S. The pathogenesis of the spinal cord disorder associated with cervical spondylosis. *Brain* 1972;95(1):87-100. https://doi.org/10.1093/brain/95.1.87
16. Tetreault L, Kopjar B, Nouri A, Arnold P, Barbagallo G, Bartels R, et al. The modified Japanese Orthopaedic Association scale: establishing criteria for mild, moderate and severe impairment in patients with degenerative cervical myelopathy. *Eur Spine J* 2017;26(1):78-84. https://doi.org/10.1007/s00586-016-4660-8
17. Persaud T. The polka-dot sign. *Radiology* 2008;246(3):980-1. https://doi.org/10.1148/radiol.2463050903
18. Wang B, Zhang L, Yang S, Han S, Jiang L, Wei F, et al. Atypical radiographic features of aggressive vertebral hemangiomas. *J Bone Joint Surg Am* 2019;101(11):979-86. https://doi.org/10.2106/JBJS.18.00746
19. Acosta FL, Sanai N, Cloyd J, Deviren V, Chou D, Ames CP. Treatment of Enneking stage 3 aggressive vertebral hemangiomas with intralesional spondylectomy: report of 10 cases and review of the literature. *J Spinal Disord Tech* 2011;24(4):268-75. https://doi.org/10.1097/BSD.0b013e3181efe0a4
20. Qiu B, Joo P, Ajabnoor R, Boyce B, Mesfin A. Surgical management of aggressive hemangiomas of the spine. *J Clin Neurosci* 2020;78:67-72. https://doi.org/10.1016/j.jocn.2020.06.012
21. Heiss JD, Doppman JL, Olfield EH. Brief report: relief of spinal cord compression from vertebral hemangioma by intralesional injection of absolute etanol. *N Engl J Med* 1994;331(8):508-11. https://doi.org/10.1056/NEJM199408253310804
22. Hekster REM, Luyendijk W, Tan TI. Spinal cord compression caused by vertebral hemangiomas relieved by percutaneous catheter embolisation. *Neuroradiology* 1972;3:160-4. https://doi.org/10.1007/BF00341502
23. Hao J, Hu Z. Percutaneous cement vertebroplasty in the treatment of symptomatic vertebral hemangiomas. *Pain Physician* 2012;15(1):43-9. PMID: 22270737
24. Pinto M, Añon M, D'Innocenzo AC, Gomez AA, Zanabria Sosa C, Gonzalez FJ. Cifoplastia con balón para el tratamiento de los hemangiomas vertebrales. Descripción de un caso y revisión bibliográfica. *Rev Asoc Arg Ortop Traumatol* 2009;74(1):66-9. Disponible en: https://www.aaot.org.ar/revista/2009/n1/art11.pdf
25. Garzon-Muvdi T, Iyer R, Goodwin CR, Abu-Bonsrah N, Orru E, Gailloud P, et al. Percutaneous embolization and spondylectomy of an aggressive L2 hemangioma. *Spine J* 2016;16(3):167-8. https://doi.org/10.1016/j.spinee.2015.10.001
26. Jaramillo Carling E. Compresión medular causada por hemangioma vertebral. Presentación de un caso y análisis de la literatura médica. *Rev Colomb Ortop Traumatol* 2005;19(1):86-9. Available at: http://www.sccot.org.co/pdf/RevistaDigital/19-01-2005/14Compresion.pdf
27. Sewell MD, Dbeis R, Bliss P, Watkinson T, Hutton M. Radiotherapy for acute, high-grade spinal cord compression caused by vertebral hemangioma. *Spine J* 2016;16(3):195-6. https://doi.org/10.1016/j.spinee.2015.10.018
28. Rades D, Bajrovic A, Alberti W, Rudat V. Is there a dose-effect relationship for the treatment of symptomatic vertebral hemangioma? *Int J Radiat Oncol Biol Phys* 2003;55(1):178-81. https://doi.org/10.1016/s0360-3016(02)03734-3
29. Kaoudi A, Capel C, Chenin L, Peltier J, Lefranc M. Robot-assisted radiofrequency ablation of a sacral s1–s2 aggressive hemangioma. *World Neurosurg* 2018;116:226-9. https://doi.org/10.1016/j.wneu.2018.05.060
30. Manning HJ. Symptomatic hemangioma of the spine. *Radiology* 1951;56(1):58-65. https://doi.org/10.1148/56.1.58

31. Raco A, Ciappetta P, Artico M, Salvati M, Guidetti G, Guglielmi G. Vertebral hemangiomas with cord compression: the role of embolization in five cases. *Surg Neurol* 1990;34(3):164-8. https://doi.org/10.1016/0090-3019(90)90067-y
32. Grau SJ, Holtmannspoetter M, Seelos K, Tonn JC, Siefert A. Giant multilevel thoracic hemangioma with spinal cord compression in a patient with Klippel-Weber-Trenaunay syndrome: case report. *Spine (Phila Pa 1976)* 2009;34(14):E498-E500. https://doi.org/10.1097/BRS.0b013e3181a4e4b8
33. Wang GX, Mu YD, Che JY, Zhang GF, Jiang G, Gao CP. Compressive myelopathy and compression fracture of aggressive vertebral hemangioma after parturition: A case report and review of literature. *Medicine (Baltimore)* 2019;98(50):e18285. https://doi.org/10.1097/MD.0000000000018285