Eur Spine J
DOI 10.1007/s00586-016-4765-0

CASE REPORT

A case report of spondylectomy with circumference reconstruction for aggressive vertebral hemangioma covering the whole cervical spine (C4) with progressive spinal disorder

Masayuki Nakahara[1] · Kenki Nishida[3] · Shinji Kumamoto[3] · Yasukazu Hijikata[3] · Kei Harada[2]

Received: 16 May 2016 / Revised: 11 August 2016 / Accepted: 31 August 2016

Abstract
Purpose To describe the surgical experience of spondylectomy and spinal reconstruction for aggressive vertebral hemangioma (VH) induced at the C4 vertebra. No reports have described surgical strategy in cases covering an entire cervical vertebra presenting with progressive myelopathy.
Methods A 28-year-old man presented with rapidly progressing skilled motor dysfunction and gait disorder. The Japanese Orthopedic Association (JOA) score was 6. Radiography showed a honeycomb appearance for the entire circumference of the C4 vertebra. Spinal computed tomography and magnetic resonance imaging showed vertebral tumor with extraosseous extension causing spinal cord compression. Results of diagnostic imaging were strongly suggestive of VH. Transarterial embolization of the spinal body branch was performed first to decrease intraoperative bleeding, followed by cervical posterior fixation to stabilize the unstable segment and excision biopsy to obtain a definitive diagnosis. After definitive diagnosis of cavernous hemangioma, two-stage surgery (anterior and posterior) was performed to complete total spondylectomy and 360° spinal reconstruction.
Results Despite multiple operations, JOA scores were 8.5 after posterior fixation, 10.5 after anterior surgery, 11 after final surgery and 16 on postoperative day 90. The patient acquired excellent clinical results without complications and returned to society.
Conclusions The present three-stage surgery comprising fixation, biopsy, and final spondylectomy with circumferential fusion from anterior and posterior approaches may offer a useful choice for aggressive VH covering the entire cervical spine with rapidly progressive myelopathy.

Keywords Aggressive hemangioma · Cervical spine · Spondylectomy · Spinal reconstruction

Introduction

Vertebral hemangioma (VH) is a common benign tumor, representing 2–3 % of all spinal tumors. Autopsy and radiographic data have shown an incidence rate of 10–12 % [1–3]. The incidence rate increases to 26 % on computed tomography (CT), which enables detection of even very small VH. This pathology frequently occurs in the thoracic spine and is least common in the cervical spine [4]. Although most VHs are asymptomatic, 0.9–1.2 % of all VHs are symptomatic with some cases meeting the indications for surgical treatment to relieve pain [5, 6]. Furthermore, VH associated with neurological deficit is rare and is termed "aggressive VH" in cases with compression of neural elements. Some series of case reports have shown clinical surgical results for aggressive VH in the thoracic or lumbar spine [7–9]. However, few reports have described surgical treatment for aggressive VH in the cervical spine [10]. We report our experience and surgical strategy in a case presenting with severely progressed

✉ Masayuki Nakahara
n-masayuki@hotmail.com

[1] Department of Spinal Surgery, Fukuoka Wajiro Hospital, 2-75 Wajirogaoka, Higashi-ku, Fukuoka, Fukuoka 811-0213, Japan

[2] Department of Neuro Surgery, Fukuoka Wajiro Hospital, Fukuoka, Japan

[3] Department of Spinal Surgery, Shinkomonji Hospital, Fukuoka, Japan

Published online: 09 September 2016

myelopathy due to aggressive VH covering the whole cervical spine.

Case report

A 28-year-old man presented with a 2-week history of numbness and disturbance of skilled hand motions and gait due to significant, progressive myelopathy. He had no cervical pain, although his condition was a severe spinal disorder. Japanese Orthopedic Association (JOA) score for the cervical spine was 6 out of 17 points.

Radiography of the lateral cervical spine showed a honeycomb appearance of the entire circumference of C4 (Fig. 1a). CT showed bone tumor mixed with thickened and rarified trabeculae embedded in enlarged vascular foramina invading into the spinal canal and transverse foramen. Disruption of bone sclerosis within the right side of the vertebra and lamina was noticeable (Fig. 1b, c). The vertebral tumor at C4 with extraosseous extension causing spinal cord compression appeared hypointense on T1-weighted magnetic resonance imaging (MRI), and hyperintense on T2- and T1-weighted gadolinium-enhanced MRI. These findings suggested vascular filling of the tumor (Fig. 2). The degree of spinal cord compression was not particularly serious. The results of diagnostic imaging using CT and MRI were highly suggestive of VH. Coronal 3-dimensional transarterial angiography showed blood vessels branching from the right vertebral artery feeding the tumor. The location of abnormal blood vessel growth was approximately within the area of disruption due to bone sclerosis (Fig. 3). Enneking bone tumor stage based on tumor development and patient symptoms corresponded to stage IV (neurological deficit with epidural and/or soft tissue extension) [11].

The patient required emergency admission due to the progressive myelopathy. The neurological symptom of finger motion showed mild recovery after cervical brace fitting (JOA score, 7). We considered that the spinal segmental instability caused by the tumor contributed progressive myelopathy, in addition to spinal canal stenosis.

Concerning treatment strategy, transarterial embolization of the spinal body branch of the vertebral artery to decrease intraoperative bleeding was performed on day 13 after admission due to the imaging features strongly indicating aggressive VH. The following day, cervical posterior fixation with a pedicle screw system to acquire stabilization of the unstable segment and excisional biopsy of the C4 spinous process were performed. Operation time was 159 min and blood loss was 246 cc. The patient acquired further improvement of finger motion and ability to walk after internal fixation, with JOA score improving to 8.5. Histological study revealed proliferation of cavernous-type vessels in the marrow space with thickened trabecular bone (Fig. 4).

On day 20 after posterior biopsy and fixation, vertebrectomy and reconstructive surgery were performed from an anterior approach. An intravascular sheath was set within the radial artery to minimize the risk of serious vertebral artery bleeding using balloon dilatation catheter to shut off blood flow. Vertebrectomy at C4 was followed

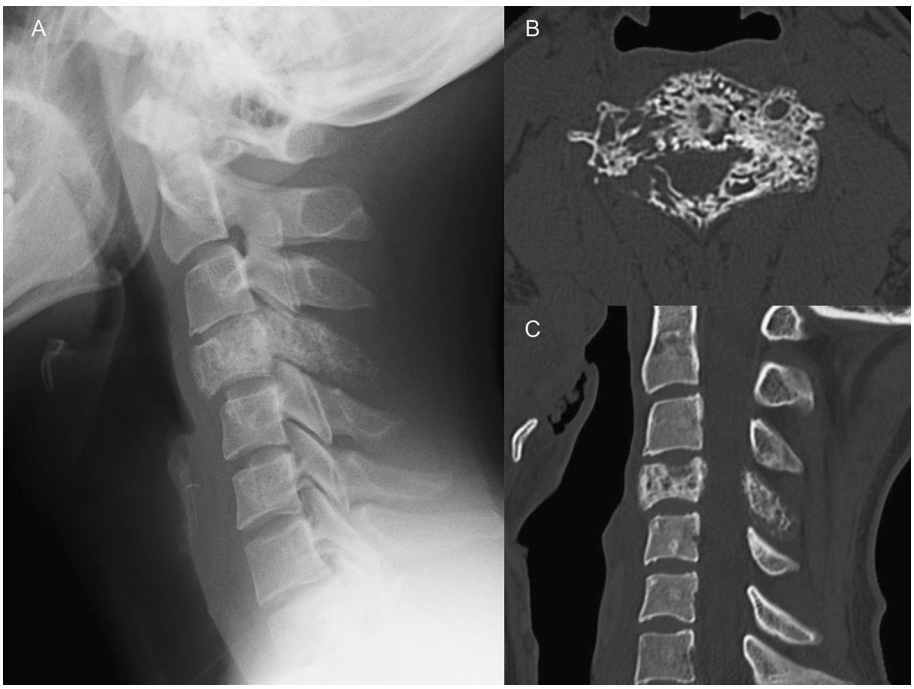

Fig. 1 **a** Initial lateral cervical radiograph shows honeycomb appearance of the whole spine at C4. **b, c** CT axial reconstruction reveals bone sclerosis mixed with thickened, rarified trabeculae embedded in enlarged vascular foramina invading into the spinal canal and transverse foramen. Disruption of bone sclerosis within right side of vertebra and lamina is noticeable

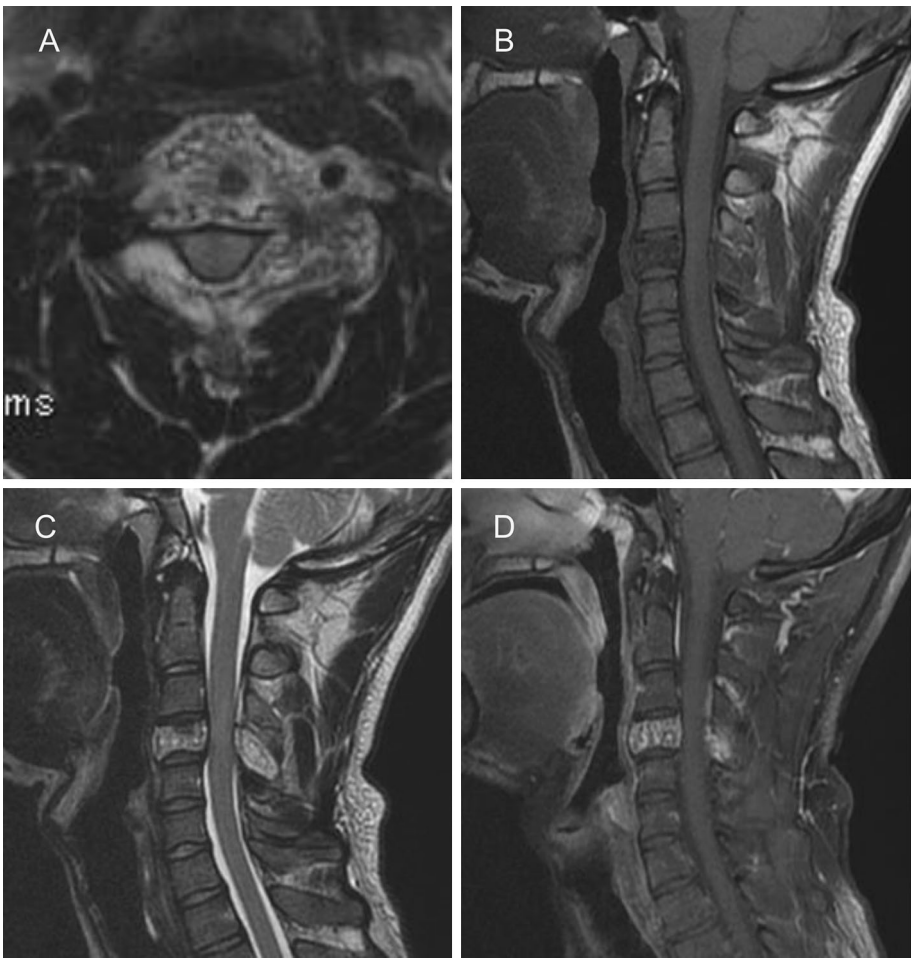

Fig. 2 **a** Axial T2-weighted magnetic resonance imaging (MRI) of the cervical spine demonstrates a vertebral tumor at C4 with extraosseous extension, compressing the spinal cord. **b** Sagittal T1-weighted MRI shows low-intensity changes in the C4 vertebra. **c** Sagittal T2-weighted MRI shows signal hyperintensity. **d** Sagittal T1-weighted gadolinium-enhanced MRI shows high intensity

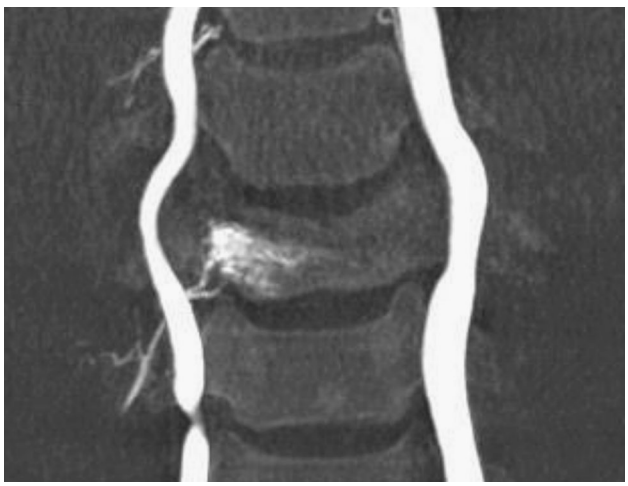

Fig. 3 Coronal 3-dimensional transarterial angiography reveals a feeder from the right vertebral artery and tumor stain in the vertebral body. The presence of abnormal blood vessel growth corresponds to the area of bone disruption with sclerosis

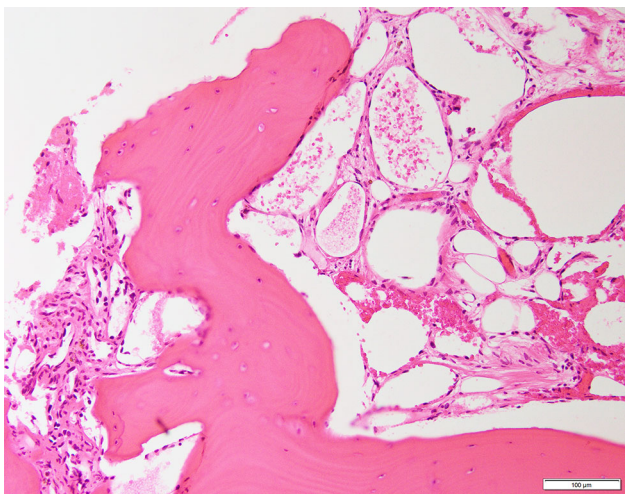

Fig. 4 Histological study revealed proliferation of cavernous-type vessels replacing the marrow space with thickened trabecular bone (hematoxylin and eosin staining, original magnification, ×200)

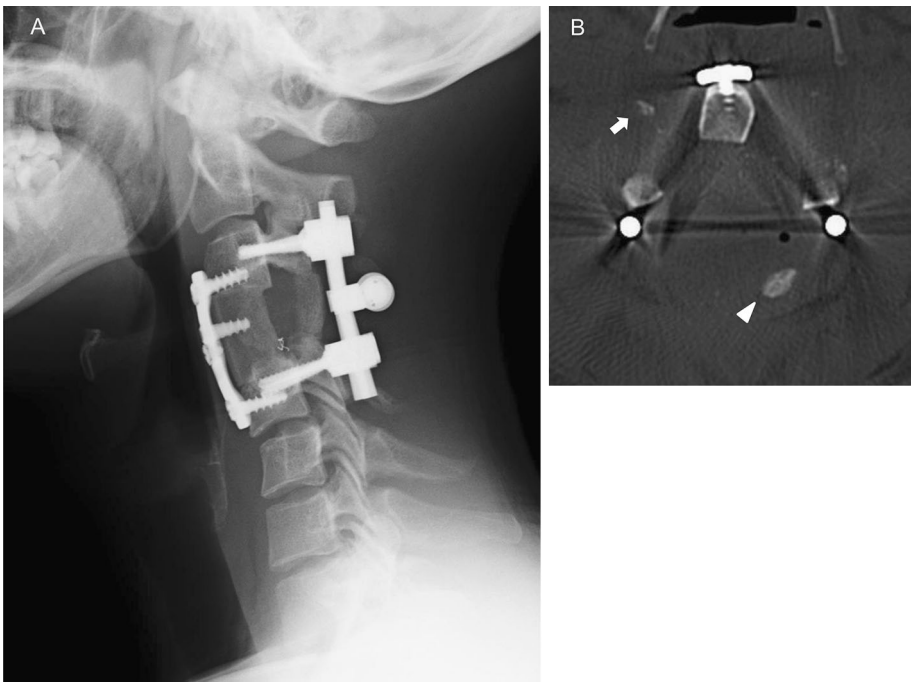

Fig. 5 **a** On a lateral radiograph, the postoperative cervical spine presents with C4 spondylectomy followed by grafting of iliac bone struts in the intervertebral and interarticular space with 360° circumference instrumentation. Embolic coil at the posterior margin of the anterior iliac bone graft. **b** Postoperative CT axial reconstruction shows bone struts after spondylectomy on axial view. *Arrow*, part of the residual tumor in the anterolateral margin; *arrowhead*, excised C3 spinous process in the posterior element

by setting an iliac bone strut and anterior plate system. Operation time was 214 min and blood loss was 180 cc. JOA score improved to 10.5 after anterior surgery.

Ten days after anterior surgery, total spondylectomy was attempted from a posterior approach. Iliac bone struts were set bilaterally in the inter-lateral mass spaces and promoted stability of the posterior element. However, a small part of VH remained at the anterolateral margin (Fig. 5). Operation time was 314 min and blood loss was 1180 cc. The patient required no allogeneic blood transfusions and the postoperative JOA score was 11.

Spinal cord deterioration was not seen after each surgery. Finally, the patient re-acquired the ability to walk independently and operate the keyboard of a personal computer, with JOA score improving to 16 by hospital day 90. Total spondylectomy was not completely achieved, and we will continue to keep a close watch on residual tumor.

Discussion

No detailed reports have described aggressive VH covering the entire cervical spine with progressive myelopathy.

Preoperative diagnosis including imaging and pathology is crucial to deciding on the surgical strategy for aggressive VH. An incorrect diagnosis can lead to an unsuitable surgical plan or the occurrence of serious complications. Clinical and imaging diagnosis of aggressive VH can be challenging. Generally, radiography shows the presence of a sclerotic vertebra with coarse vertical trabeculae (Corduroy sign) or a honeycomb appearance within the vertebra [5]. A polka-dot appearance within the vertebra body represents thickened vertical trabeculae on axial CT [12]. Increased signal intensity on both T1- and T2-weighted MRI represents the high fat content of the affected vertebra. The signal intensity indicates the characteristic of non-progressive hemangioma [2, 12]. The lower intensity on T1-weighted MRI indicates a hypervascular tumor with high growth potential [13]. Imaging diagnosis in the present case suggested a high growth VH.

In histopathological examination, VH is a benign tumor that can show pathological types such as cavernous (as in our case), capillary or mixed type. Pastushyn et al. described that 28 % of their VH cases were cavernous type, 50 % were capillary type and 22 % were mixed type [3]. Percutaneous CT biopsy is simple and less invasive, and of high value [14]. However, needle insertion for VH in the present case could have aggravated spinal deterioration by increasing spinal column instability.

Preoperative embolization is reportedly effective to reduce intraoperative bleeding [7]. Posterior spinal fixation with biopsy of the spinous process as the first surgery and anterior vertebrectomy as the second surgery did not lead to marked blood loss. The duration of 31 days after embolization may have reduced the hemostatic effect due to re-hypervascularization and thus have resulted in active bleeding at final posterior surgery.

Concerning the surgical methods, vertebroplasty with polymethylmethacrylate or direct injection of ethanol is an effective and quick method for VH. These materials cause intralesional thrombosis and shrinkage of the lesion as a result of devascularization [15, 16]. Vertebroplasty also

prevents vertebral collapse by strengthening destructed vertebral trabeculae and cortex. Guarnieri et al. reported good clinical results with no pathological fracture or recurrence after vertebroplasty [16]. However, the sample size of patients in stage III or IV compressing neural elements was too small. In addition, polymethylmethacrylate has a risk of leakage into the spinal canal, representing the most common complication [17, 18]. Whether vertebroplasty represents a curative treatment for VH in stage III (asymptomatic lesion with epidural and/or soft tissue extension) or IV thus remains uncertain. Ethanol injection can cause complications such as spinal deterioration, including Brown-Sequard syndrome, recurrence and vertebral collapse [14, 19]. Evidence for the effects on tumor volume reduction or maintenance of vertebral height to load strength is lacking. In stage III or IV, these methods carry a potential risk of spinal deterioration.

Acosta et al. described symptoms of pain or neurological deficit as resulting from spinal cord compression by direct extraosseous extension, vertebral body or neural arch expansion, or pathological fracture. They suggested that symptomatic VH in stage II (bone destruction with pain) is a candidate for vertebroplasty. Laminectomy is also an effective procedure to treat spinal cord compression caused by osseous lesion growth involving the vertebral body or posterior elements. Despite laminectomy offering good symptomatic relief, recurrence occurred in two cases [7]. After 5 years, they suggested total resection of all involved vertebral segments followed by 360° spinal fixation in cases with extraosseous extension [20]. Data were presented from ten consecutive cases of stage III or IV aggressive thoracic VH including recurrence. They noted that partial resection or vertebroplasty of more stage III lesions might lead to recurrence, citing three recurrences. Cases treated by total resection showed no recurrences at a mean follow-up of 2.4 years. Kato et al. also reported clinical results of total resection for five cases with aggressive VH in the thoracic spine [8]. Even though all cases showed extraosseous extension in stage IV, no patients had experienced recurrence at a mean follow-up of 135 months. From the above, total spondylectomy is recommended in our case.

In terms of the number of operation, we should have chosen a first-stage surgery consisting of combined anterior-posterior surgery aimed at total resection and spinal reconstruction. The patient needed surgery as an effective early intervention for severely progressive spinal disorder. In addition, the possibility of malignancy prevented us from making decisions on surgical strategy under the absence of pathological diagnosis. And we were uncertain as to whether a first-stage surgery aimed at total spondylectomy could be performed while controlling intraoperative bleeding. As a result, although the patient showed relatively high blood loss at final surgery, he acquired excellent clinical results without complications and successfully returned to work. The present treatment strategy represents one option in cases of stage IV aggressive VH needing early surgical intervention due to severely progressive myelopathy.

Conclusions

Aggressive vertebral hemangioma in stage III or IV covering the entire cervical spine is extremely rare and total spondylectomy is recommended to prevent recurrence. If spinal deterioration is progressive due to spinal segmental instability and imaging features are suggestive of aggressive vertebral hemangioma, cervical posterior fixation to acquire stabilization combined with excisional biopsy is validated. The three-stage surgery we applied, including fixation, biopsy, and spondylectomy with circumference fusion would represent one choice for cervical aggressive VH presenting with rapidly progressive myelopathy.

Compliance with ethical standards

Conflict of interest No conflicts of interests exist.

References

1. Fox MW, Onofrio BM (1993) The natural history and management of symptomatic and asymptomatic vertebral hemangiomas. J Neurosurg 78:36–45
2. Laredo JD, Reizine D, Bard M et al (1986) Vertebral hemangiomas: radiologic evaluation. Radiology 161:183–189
3. Pastushyn AI, Slin'ko EI, Mirzoyeva GM (1998) Vertebral hemangiomas: diagnosis, management, natural history and clinicopathological correlates in 86 patients. Surg Neurol 50:535–547
4. Slon V, Stein D, Cohen H et al (2015) Vertebral hemangiomas: their demographical characteristics, location along the spine and position within the vertebral body. Eur Spine J 10:2189–2195
5. Healy M, Herz DA, Pearl L (1983) Spinal hemangiomas. Neurosurgery 13:689–691
6. Nguyen JP, Djindjian M, Gaston A et al (1987) Vertebral hemangiomas presenting with neurologic symptoms. Surg Neurol 27:391–397
7. Acosta FL Jr, Dowd CF, Chin C et al (2006) Current treatment strategies and outcomes in the management of symptomatic vertebral hemangiomas. Neurosurgery 58:287–295
8. Kato S, Kawahara N, Murakami H et al (2010) Surgical management of aggressive vertebral hemangiomas causing spinal cord compression: long-term clinical follow-up of five cases. J Orthop Sci 15:350–356
9. Urrutia J, Postigo R, Larrondo R et al (2011) Clinical and imaging findings in patients with aggressive spinal hemangioma requiring surgical treatment. J Clin Neurosci 18:209–212
10. Jiang L, Liu XG, Yuan HS (2014) Diagnosis and treatment of vertebral hemangiomas with neurologic deficit: a report of 29 cases and literature review. Spine J 14:944–954

11. Enneking WF (1986) A system of staging musculoskeletal neoplasms. Clin Orthop Relat Res 204:9–24
12. Ross JS, Masaryk TJ, Modic MT et al (1987) Vertebral hemangiomas: MR imaging. Radiology 165:165–169
13. Laredo JD, Assouline E, Gelbert F et al (1990) Vertebral hemangiomas: fat content as a sign of aggressiveness. Radiology 177:467–472
14. Rimondi E, Staals EL, Errani C et al (2008) Percutaneous CT-guided biopsy of the spine: results of 430 biopsies. Eur Spine J 17:975–981
15. Doppman JL, Oldfield EH, Heiss JD (2000) Symptomatic vertebral hemangiomas: treatment by means of direct intralesional injection of ethanol. Radiology 214:341–348
16. Guarnieri G, Ambrosanio G, Vassallo P et al (2009) Vertebroplasty as treatment of aggressive and symptomatic vertebral hemangiomas: up to 4 years of follow-up. Neuroradiology 51:471–476
17. Evangelopoulos DS, Kontovazenitis P, Kokkinis K et al (2009) Cement leakage in a symptomatic vertebral hemangioma: a case report and review of the literature. Cases J 2:7148
18. Hao J, Hu Z (2012) Percutaneous cement vertebroplasty in the treatment of symptomatic vertebral hemangiomas. Pain Physician 15:43–49
19. Niemeyer T, McClellan J, Webb J et al (1999) Brown-Sequard syndrome after management of vertebral hemangioma with intralesional alcohol. A case report. Spine 24:1845–1847
20. Acosta FL Jr, Sanai N, Cloyd J et al (2011) Treatment of Enneking stage 3 aggressive vertebral hemangiomas with intralesional spondylectomy: report of 10 cases and review of the literature. J Spinal Disord Tech 24:268–275