Aggressive vertebral hemangioma of the thoracic spine without typical radiological appearance

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Abstract

Purpose Vertebral hemangioma (VH) is virtually vascular malformation, which is usually asymptomatic. Only 3.7 % of VH may become active and symptomatic, and 1 % may invade the spinal canal and/or paravertebral space. Treatment protocols for active or aggressive VHs are still in

controversy. Reported treatments include radiotherapy, vertebroplasty, direct alcohol injection, embolization, surgery and a combination of these modalities.

Methods A 41-year-old lady was presented with 18 month history of intermittent back pain. CT revealed T5 osteolytic lesion with epidural and paravertebral extension. The first CT guided biopsy yielded little information.

Results Histopathological diagnosis of the second biopsy was VH. Vertebroplasty, posterior decompression and fixation were performed followed by postoperative radiotherapy. Her symptoms were resolved immediately after the operation. At 12 months follow-up, no recurrence was detected by CT with contrast enhancement.

Conclusion Surgical decompression, vertebroplasty and fixation are safe and effective for aggressive VH. More attention is needed in determining the algorithm for the diagnosis and treatment of aggressive VH.

Keywords Vertebral hemangioma · Diagnosis · Radiotherapy · Vertebroplasty · Surgery

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Case presentation

The case is a 41-year-old female who is a physician herself. The patient began to suffer from intermittent back pain for 18 months before visiting us. In the latest 6 months, she also presented with numbness of the left thoracic wall with intermittent radiating pain that aggravated in the last 2 months. She had no numbness or weakness in her extremities though.

Before being referred to our centre, she was found to have a vertebral lesion at T5 level using computed tomography (CT) and magnetic resonance imaging (MRI) scan at another hospital. She also under went CT guided

percutaneous needle biopsy that revealed only necrosis tissue and bone tissue. A positron emission computed tomography CT (PET-CT) scan was also performed with no other lesion being detected elsewhere.

Diagnostic imaging section

On examination, there was tenderness along her middle thoracic spine. Her upper and lower limbs were neurologically intact. The eagle's sign could be observed on anteroposterior radiography (Fig. 1). CT scan revealed an osteolytic lesion in the vertebral body, left pedicles and lamina of T5. The cortex of T5 vertebral body was also partially disrupted. MR scan showed the tumor occupying the entire T5 vertebral body and invaded the spinal canal and paravertebral region. It was heterogeneously hypointense on T1-weighted image and hyperintense on T2-weighted image. Significant increase in signal intensity was shown by MR scan after gadolinium contrast enhancement. Compression of the dural sac by the epidural extension of the tumor was evident on axial MR imaging. Only T5 lesion was detected by PET-CT, and the Standardized Uptake Value (SUV) was 1.3.

Given the aggressive radiological appearance, metastatic disease or primary malignant tumor was first to be considered. Although the negative result of PET-CT ruled out metastasis leaving malignant tumor the most likely

diagnosis, it was not supported by the obscure result from the CT guided biopsy specimen either. The second biopsy was prompted in search for diagnosis by our radiology department.

Instead of aiming at the bony lesion as in the previous biopsy (Fig. 1), the second biopsy targeted on the paravertebral mass (Fig. 2) and subsequently came up with a distinct histopathological diagnosis as vertebral hemangioma (VH).

Historical review

According to large autopsy and radiography studies [9], vertebral hemangioma is a relatively common tumor, with an estimated incidence of 10–12 % in the general population. Although normally being referred to as a benign tumor, VH is virtually vascular malformation by nature. It can be further categorized into three histological types: capillary, cavernous, and mixed VH [8].

VHs most frequently occur to the thoracic spine. They are usually asymptomatic and often found incidentally by plain radiographs, CT and/or MRI [1, 2]. Only 3.7 % of VH may become active and symptomatic, usually causing pain alone. For a small subset of these patients, however, VH may become aggressive and invade the spinal canal and/or paravertebral space [9]. Although rarely causing cord compression and neurological symptoms, should it

Fig. 1 a, b Anteroposterior radiography showed the eagle sign (*thin arrow*). c–e CT scan (axial, sagittal and coronal) revealed an osteolytic lesion in T5 vertebral body, left pedicles and lamina. The cortex of T5 vertebral body was partially disrupted. f, g. The lesion was heterogeneously hypointense on T1-weighted image and

hyperintense on T2-weighted image. h. The lesion was hyperintense on fat suppression image. i. Axial MR image showed the vertebral tumor extended into the spinal canal and paravertebral region (*thick arrow*). j. CT scan obtained during the first biopsy carried out in another hospital showed the tip by a transpedicular approach

occur, it is predominantly to the thoracic spine. Before 1961, there were 70 reported VH cases with spinal cord compression in the literature, including 10 cervical cases [13, 15].

In typical cases, hemangiomas are usually homogeneous, and can be diagnosed using CT and MRI. Roentographically, the lesion is characterized by the vertical striation and/or honeycomb pattern shown in the involved vertebral bodies. CT scans can also demonstrate the classical “honeycomb” sign (Fig. 3a) or vertically oriented vertebral lucencies separated by thickened trabecular bone, as shown by the contrast MR image and/or fat suppression MR as hypervascular lesion (Fig. 1h). In latent VHs, MRI showed increased signal intensity on both T1 and T2

weighted images with alternated hypointense areas, giving a salt and pepper appearance in typical VH cases (Fig. 3b). The hyperintense areas represent fat and degenerated marrow, while the hypointense areas represent flow voids.

It has been illustrated that aggressive VH usually contains less fat tissue, hence produces a low signal on T1 weighted image and a high signal on T2 (Fig. 1f, g). Sometimes the change can be atypical, causing difficulties in diagnosis. On other occasions, the vertebral body can be destructed without typical thickening of the trabecular bone (Fig. 1). Even the cortex can occasionally be breached resulting in epidural and/or paravertebral soft tissue extension of the tumor. Under these circumstances, it is difficult to differentiate aggressive VH from malignant vascular tumors, e.g., epithelioid hemangioendothelioma and angiosarcoma, metastasis and myeloma.

Based on the patients’ lesion and symptoms, VH could be classified into three categories: type I, latent (Enneking S1), mild bony destruction with no symptom; type II, active (S2), bony destruction with pain; type III aggressive (S3), asymptomatic lesion with epidural and/or soft tissue extension; type IV aggressive (S3), neurological deficit with epidural and/or soft tissue extension [6].

Biopsy. Percutaneous CT guided biopsy is indicated in the suspected malignant cases. Under CT guidance, the biopsy needle was usually inserted through pedicle or via the lateral side of the affected vertebra (Fig. 1j). In our opinion, bony biopsy is safer, while soft tissue tumor mass biopsy (Fig. 2) is more likely to obtain pathological diagnosis [14]. Whenever the percutaneous biopsy was deemed to be dangerous, the patient was suggested for treatment directly.

For this particular case, without the biopsy pathology, wide or marginal excision, such as total En bloc spondylectomy, might be planned for its aggressive radiological appearance [11]. Second biopsy should still be carried out in experience hands for suspected malignant spine lesions.

Fig. 2 The second biopsy carried out in our centre shows the tip of the trocar placed in the paravertebral soft tissue

Fig. 3 Another vertebral hemangiomas case who had neurological deficit with typical radiological findings. **a** Axial CT scan showed the classical “honeycomb” sign. The laminar lesion extruded into the canal with bony compression. **b** The lesion gave a salt and pepper appearance in axial MR image. The lesion had both epidural (*thin arrow*) and paravertebral (*thick arrow*) extension

Rationale for treatment and evidence-based literature

In general, treatment for vertebral hemangiomas should entirely base on patients' symptoms, for even the aggressive VH lesions are benign in nature and rarely progress rapidly.

1. For *type I* VHs, only observation is needed [9].

2. In contrast to type I VHs, treatment protocols for active or aggressive VHs are still in controversy. For *type II* VHs, radiotherapy was used to be the first of choice. Its vascular necrosis and/or anti-inflammatory effect acts to obliterate hemangiomas and control pain. The effective dosage is 30–40 Gy over a 3- to 4-week period and the associated complications, such as radionecrosis, radiation-induced myelitis and secondary malignancy are rare. Nevertheless, the fear for its secondary malignancy had prompted the development of new treatment, i.e., vertebroplasty. More recently, however, Heyd et al. [10] points out that secondary malignancy have been over-emphasized, and vertebroplasty has its own fatal complications such as leakage and pulmonary embolism.

In most VH cases, the vertebral trabecules tend to be thicker and stronger than the normal vertebral trabecules, hence no vertebral augmentation is needed. Vertebroplasty could cure VH by obstructing the vessels of tumor (internal embolization).

3. As for *type III* VHs, the importance of early diagnosis and treatment to prevent neurological deficit has been emphasized by Doppman and his colleagues [7]. However, it is argued by Boriani and colleagues who insist that VH is pathologically benign and treatment should be symptoms orientated [4]. To them, this type of indolent VH lesions is not really aggressive even with extracompartment extension hence preventive treatments are not mandatory.

For us, we prefer observation, but spinal radiosurgery is also a reasonable choice. We inform the patient and his/her family, and they make the treatment decision after comprehensive consultation.

4. Treatment protocol for *type IV* VHs is even more diversiform. Reported treatments include radiotherapy [10], vertebroplasty, direct alcohol injection [7], embolization of the feeding arteries, surgery, and a combination of these modalities. Successful results have been reported for each of these treatments. But the corresponding technical requirement, learning curve, complications and costs vary greatly. Besides, it is difficult to assess these treatments individually as most reports were retrospective case reports with mixed cases with local pain alone and most patients had combined treatments.

4.1 *Radiotherapy* alone is only appropriate for patients with slight and slow progressive neurological deficit because of the concern that the effect of radiotherapy is not as prompt as desired for patients with progressive

neurological deficits caused by cord compression. Spinal radiosurgical treatment could lead to better neurological outcome by focusing on the epidural lesion [16]. Surgery is indicated if radiotherapy fails in 3 months' time.

Heyd and colleagues reported that neurological symptoms completely resolved in 79 % of their cases after radiotherapy [10]. But amongst the 24 cases with neurological symptoms in their retrospective multicenter study, 20 also had surgical intervention.

4.2 *Vertebroplasty* is not suggested for type IV lesions, for it might exacerbate cord compression. Instead, it is safe to carry out an intraoperative vertebroplasty after laminectomy under direct visualization.

In some aggressive cases where both trabecules and the cortex are destructed, vertebroplasty can strengthen the vertebral bodies and prevent vertebral collapse. After radiotherapy, the radiological reossification rate was only 26 % [10].

4.3 *Direct alcohol injection* into the tumor is highly effective in achieving hemostasis. It causes intralesional thrombosis and destruction of the endothelium of which the hemangioma composed. Devascularisation is followed by shrinkage of the lesion, which decompresses the cord and nerve roots. CT angiography revealed that these hypervascular components are often inhomogeneous [7], and the injection targeted the most hypervascular subsection. High successful rate was reported by Heiss, Losner and Doppman [7]. More recently in India, direct injection has been carried out during decompressive surgery in VH cases.

The complications include transient neurological deterioration (including Brown–Sequard syndrome), pathologic fractures and recurrence. Doppman et al. [7] suggested that a moderate dose of ethanol (<15 ml) obliterated the hemangioma without affecting spinal stability. In Bas' series, 6 out of 24 patients were not ideal for alcohol injection because the contrast medium was not retained in the lesions [5].

4.4 *Embolization* of feeding vessel has been used widely as a presurgical adjuvant treatment for reducing intraoperative hemorrhage. Kawahara and colleagues had indicated that the interruption of bilateral segmental arteries at three levels does not damage spinal cord function in either dog or human [12]. But it may not be possible in the absence of discernible feeding vessel. With the development of intraoperative vertebroplasty and/or direct alcohol injection, preoperative embolization for VH might not be needed any more.

4.5 *Operation* is indicated in cases with rapid progressive and/or severe myelopathy. Sometimes, urgent surgical decompression is indicated. However, diagnostic failure can potentially lead to inappropriate surgical plan and serious complications, especially life-threatening hemorrhage. Before 1960, reported surgical neurological recovery

rate was on average 73 % (range 43–85 %), and the mortality rate was 11.7 % [13]. In recent studies, however, neurological recovery rate is nearly 100 %, whereas the mortality is rarely mentioned [9].

The goals of surgery include bony decompression and excision of soft tissue components of the tumor that compress the neural elements. Different surgical techniques have been reported, namely, laminectomy [9], corpectomy, spondylectomy [3], even total en-bloc spondylectomy (TES) [11, 12].

4.5.1 *Laminectomy* could be combined with intraoperative vertebroplasty or alcohol injection, which could shrink the hemangiomas and lead to intralesional thrombosis. In most type IV cases, it is the epidural soft tumor mass in

stead of bony lesion which causes neurological deficit. Appropriate decompression could be achieved without anterior decompression. After laminectomy, alcohol or cement leakage could be easily controlled under direct visualization. With these techniques, preoperative embolization might no longer be needed.

Although postoperative radiotherapy plays an important role in reducing reoccurrence in cases of partial tumor excision [9], it may not be needed in cases with ideal intraoperative vertebroplasty or alcohol injection.

Our standard procedure is laminectomy and vertebroplasty with the excision of epidural soft tissue components. Postoperative radiotherapy is indicated, only if residual lesion is confirmed by contrast enhanced CT.

Fig. 4 **a** Intra-operative vertebroplasty. After vertebroplasty and pedicle screw fixation, the dura was exposed. **b** Intra-operative picture after laminectomy. After *left* facetomy, the soft tumor mass was clearly exposed and then resected. **c** Lateral radiography before closure

Fig. 5 Our suggested protocol (PUTH) for the diagnosis and management of aggressive vertebral hemangioma with neurological deficit. It is based on literature review and our previous experience. § our first suggested treatment choice. *classical CT findings are honeycomb and vertically oriented vertebral lucencies separated by thickened trabecular bone; classical MRI findings are a salt and pepper appearance and significantly enhanced by gadolinium

4.5.2 *Spondylectomy* is too radical even for aggressive VH, considering its benign nature. It may be specifically indicated for ventrally located lesions with bony compression. Given the VHs are usually highly vascularised, it can cause substantially great intraoperative blood loss. Even with the improvement of surgical technique, it remains an extremely technically demanding procedure. With preoperative embolization, the average blood loss was 2.1 L (range 0.8–5 L) in the 10 spondylectomy cases reported by Acosta et al. [3]. The estimated blood loss was 4.0 L in the TES case reported by Inoue et al. [11].

Procedure and outcome

We proposed two options to the patient. The first was radiotherapy alone, with the risk of pathological fracture during the therapy for her severe bony destruction. The second was combined laminectomy and vertebroplasty with excision of the epidural soft tumor mass followed by postoperative radiotherapy. After consulting with radiotherapist and her family, the patient chose surgery. During the surgery, standard vertebroplasty and pedicle screw fixation were performed followed by laminectomy (including T5 and part of T4 and T6) and exposure of the dura. After left facetomy, the soft tumor mass was clearly exposed laterally and resected with bipolar coagulation (Figs. 4, 5). The estimated blood loss was 1,000 ml. Her symptoms were resolved immediately after the operation. Radiotherapy with a dose of 30 Gy was carried out 4 weeks after surgery. At 12 months follow-up, no recurrence was detected by CT with contrast.

Conflict of interest None.

References

- Alexander J, Meir A, Vrodos N et al (2010) Vertebral hemangioma an important differential in the evaluation of locally aggressive spinal lesions. *Spine* 35(18):E920–E923
- Acosta FL Jr, Sanai N, Chi JH et al (2008) Comprehensive management of symptomatic and aggressive vertebral hemangiomas. *Neurosurg Clin N Am* 19(1):17–29
- 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
- Bandiera S, Gasbarrini A, De Iure F et al (2002) Symptomatic vertebral hemangioma: the treatment of 23 cases and a review of the literature. *Chir Organi Mov* 87:1–15
- Bas T, Aparisi F, Bas JL (2001) Efficacy and safety of ethanol injections in 18 cases of vertebral hemangioma: a mean follow-up of 2 years. *Spine* 26(14):1577–1582
- Boriani S, Weinstein JN, Biagini R (1997) Primary bone tumors of the spine. Terminology and surgical staging. *Spine* 22:1036–1044
- Doppman JL, Oldfield EH, Heiss JD (2000) Symptomatic vertebral hemangiomas: treatment by means of direct intralesional injection of ethanol. *Radiology* 214(2):341–348
- Fletcher CDM, Unni KK, Mertens F (2002) World Health Organisation classification of tumours Pathology and genetics of tumours of soft tissue and bone. IARC Press, Lyon, pp 310–314
- Fox MW, Onofrio BM (1993) The natural history and management of symptomatic and asymptomatic vertebral hemangiomas. *J Neurosurg* 78:36–45
- Heyd R, Seegenschmiedt MH, Rades D et al (2010) Radiotherapy for symptomatic vertebral hemangiomas: results of a multicenter study and literature review. *Int J Radiat Oncol Biol Phys* 77:217–225
- Inoue T, Miyamoto K, Kodama H et al (2007) Total spondylectomy of a symptomatic hemangioma of the lumbar spine. *J Clin Neurosci* 14(8):806–809
- Kawahara N, Tomita K, Murakami H et al (2011) Total en bloc spondylectomy of the lower lumbar spine: a surgical techniques of combined posterior-anterior approach. *Spine* 36:74–82
- Krueger EG, Sobel GL, Weinstein C (1961) Vertebral hemangioma with compression of spinal cord. *J Neurosurg* 18:331–338
- Liu XG, Liu ZJ, Dang GT et al (2004) CT-guided percutaneous biopsy of the spine (352 cases review). *Chin J Spine Spinal Cord* 14:82–85
- Robbins LR, Fountain EM (1958) Hemangioma of cervical vertebrae with spinal-cord compression. *N Engl J Med* 258:685–687
- Sohn MJ, Lee DJ, Jeon SR et al (2009) Spinal radiosurgical treatment for thoracic epidural cavernous hemangioma presenting as radiculomyelopathy: technical case report. *Neurosurgery* 64:E1202–E1203