Aggressive hemangioma of the thoracic spine

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Radiology Case. 2011 Oct; 5(10):7-13 :: DOI: 10.3941/jrcr.v5i10.828

ABSTRACT

Vertebral hemangiomas are common lesions and usually considered benign. A rare subset of them, however, are characterized by extra-osseous extension, bone expansion, disturbance of blood flow, and occasionally compression fractures and thereby referred to as aggressive hemangiomas. We present a case of a 67-year-old woman with progressive paraplegia and an infiltrative mass of T4 vertebra causing mass effect on the spinal cord. Multiple conventional imaging modalities were utilized to suggest the diagnosis of aggressive hemangioma. Final pathologic diagnosis after decompressive surgery confirmed the diagnosis of an osseous hemangioma.

CASE REPORT

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A 67-year-old African American woman with a past medical history of diabetes, hypertension, hyperparathyroidism, and COPD presented with insidious development of left lower extremity weakness and spasticity over the previous 5 months which worsened rapidly during the two weeks prior to admission. On presentation, neurologic exam demonstrated decreased strength with spasticity of the bilateral lower extremities and decreased sensation below the level of T6. Clonus as well as extensor plantar reflexes were also noted bilaterally. There was no ataxia, dysmetria or dysdiadochokinesia.

Pre- and post-contrast magnetic resonance imaging (MRI) of the thoracic spine, ordered as initial study based on her clinical signs and progressive worsening of symptoms, demonstrated a T2 hyperintense, T1 hypointense enhancing expansile osseous lesion with trabecular thickening involving the entire T4 vertebral body extending into the left posterior elements. Additionally, the mass extended into the bony spinal canal and left posterolateral paraspinal musculature causing compression of the thoracic spinal cord at this level (Fig 1 ad). Subtle increased T2/short T1 inversion recovery (STIR) signal was demonstrated in the spinal cord from approximately T3 to T5 vertebral levels (Fig 1 e-f). Aggressive hemangioma versus metastatic lesion was suggested based on these findings and computed tomography (CT) of the thoracic spine was recommended for further evaluation.

Noncontrasted CT demonstrated thickened, vertically oriented trabeculae involving the entire vertebral body and left posterior elements with expansion and irregularity of the osseous cortex in addition to an epidural soft tissue mass. There was characteristic "polka dot" appearance on axial images and "corduroy cloth/jail bar" appearance on sagittal and coronal images (Fig. 2).

Spinal angiography, recommended by the neurosurgery consulting service, revealed minimal corresponding tumor blush, supplied mainly by the right supreme intercostal artery, at the T4 level (Fig. 3). Embolization was not performed secondary to only faint tumor blush.

Surgical option was preferred with a posterior left hemilaminectomy and partial corpectomy at T4 with resulting resection of a single-segment, extradural spinal tumor. Posterior segmental stabilization was obtained from T2 through T6 with pedicle screw fixation at T2, T3, T5 and T6.

Diagnosis of an osseous hemangioma was confirmed by histologic analysis (Fig 4).

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DISCUSSION

First described by Perman, in 1926, followed by Bailey and Bucy in 1930, vertebral hemangiomas are common benign lesions of the spinal column with an estimated incidence of 10-12% based on large autopsy series and reviews of spine radiographs. Although typically incidental findings, they are symptomatic in 0.9 to 1.2% of adults [1, 2]. This figure is much lower in the pediatric population with only five cases reported in the literature, including a recently reported case by Cheung et al. [2]. These small but significant subset of symptomatic lesions are known as aggressive hemangiomas and are characterized by bone expansion, extraosseous extension of tumor, disturbance of local blood flow, and rarely compression fractures [1]. Approximately 45% of aggressive hemangiomas are associated with neurologic deficits, the others only characterized by pain [3].

Vertebral hemangiomas are classically characterized by sparing and thickening of vertically striated trabeculae which preserve the functional capability of the vertebral body to withstand an axial load. This appearance has been described on radiographs and CT as "polka-dot", "honeycomb", "corduroy cloth", "jail bar", and "salt and pepper", with decrease in the overall density of the vertebral body due to the presence of fatty marrow [5].

Intralesional fat of hemangiomas causes increased signal intensity on T1 weighted MR images. On T2-weighted images, the signal intensity increases due to high water content [6]. T2-hyperintensity is often greater than that of fat, thereby differentiating hemangiomas from focal fat deposition [7]. Aggressive hemangiomas typically contain less fat and more vascular stroma thereby producing a low MR signal on T1 weighted images [1,4,8]. This appearance may resemble a metastatic lesion, however, metastatic lesions usually have low signal on T1 weighted images and high signal on T2 weighted images [7]. In addition, the morphology, including the presence of coarsened trabeculae can be used for differentiation. For indeterminate cases, CT aids in problem solving, since the modality is more sensitive to the characteristic osseous remodeling [9].

Paget's disease of the spine can have a similar appearance but is usually distinguished by expansion of the vertebral body with peripheral cortical thickening demonstrated on CT, corresponding to the characteristic "picture frame" vertebral body on radiographs [10]. Lymphoma may also have a similar appearance but is usually distinguished by homogeneous enhancement on contrasted MRI [11].

Aggressive vertebral hemangiomas most often occur between T3 and T9 vertebral segments [12]. They generally occupy the entire vertebral body, extend into the neural arch, expand the osseous margins, and contain a soft tissue component [12]. Cord compression and subsequent myelopathy may result from either encroachment of extradural soft tissue, pathologic fracture, or hemorrhage [13]. Vertebral hemangiomas may become symptomatic during pregnancy due to increased intraabdominal pressure and vascular redistribution of flow in the vertebral venous plexus when the uterus enlarges [14].

The histologic pattern of osseous hemangiomas is characterized by the proliferation of anomalous thin-walled blood vessels and sinuses lined by endothelium between the thickened, vertically oriented trabeculae of bone. The dilated vascular channels are set in a stroma of fat. The ratio of fat to vascular tissue wrapped between the pillars of bone determines the signal intensity on MR images [4].

Treatment options for symptomatic or aggressive hemangiomas without cord compression include vertebroplasty [14,15], embolization, and sclerotherapy [16]. In a recent case series of four patients, Urrutia et al. demonstrated that surgical resection is a relatively safe method for treatment of compressive vertebral hemangiomas [17]. Radiation therapy may be used in those cases of subtotal resection [8, 17].

TEACHING POINT

Vertebral hemangiomas are most often benign, incidentally imaged lesions with a good prognosis; however, a rare subset of them can be aggressive with devastating neurologic sequela. It is important for the clinician to keep aggressive hemangioma as a differential diagnosis in any patient presenting with myelopathic symptoms and obtain early imaging, especially MRI, in order to guide proper treatment. As aggressive hemangiomas often mimic other aggressive spinal lesions, additional imaging modalities such as CT, angiography, and radiography are usually needed to make a definitive diagnosis.

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FIGURES



Figure 1: 67-year-old woman with aggressive hemangioma of the thoracic spine and progressive paraplegia. GE Signa Excite 1.5 Tesla magnet. Precontrast axial (TR: 366, TE: 14) (a) and sagittal (TR: 500, TE:14) (b) T1 weighted images depicting an initially hypointense soft tissue mass which enhances greatly after intravenous injection of 15cc OptiMARK (gadoversetamide) as seen on postcontrast axial (TR: 467, TE: 14) (c) and fat-suppressed sagittal (TR: 400, TE: 14) (d) T1 weighted images. The mass involves the T4 vertebral body (asterisks) with extension into the spinal canal (arrows), postererior elements (curved arrows), and left paraspinous soft tissues (arrow heads). Sagittal T2 weighted (TR: 4866, TE: 113) (e) and Short T1 Inversion Recovery (STIR) (TR: 4900, TE: 54) (f) images demonstrating high signal within the spinal cord at the level of T4 consistent with spinal cord edema (open arrowheads).



Figure 2: 67-year-old woman with aggressive hemangioma of the thoracic spine and progressive paraplegia. Noncontrast CT images depicting an expansile lesion of T4 vertebral body (arrows) with trabecular thickening and intervening decreased density giving the characteristic "polka dot" appearance on axial image (a) and "corduroy cloth/jail bar" appearance on coronal (b) and sagittal (c) images compatible with an osseous hemangioma. Note the aggressive nature with soft tissue component extending and involving the spinal canal (arrowheads) and posterior elements (curved arrows). (Scanner: GE LightSpeed Pro16; Protocol: Source axial image slice thickness: 0.63mm, kVp 120, mA 631)



Figure 3: 67-year-old woman with aggressive hemangioma of the thoracic spine and progressive paraplegia. Selected digital subtraction angiography (DSA) images at the T3-T4 vertebral level demonstrating abnormal pooling of contrast (white arrows) in the T4 vertebral body on early phase (a) with accumulation of contrast on more delayed phases (b) consistent with a hemangioma. The blood supply was mainly from the right supreme intercostal artery (black arrows).



Figure 4: 67-year-old woman with aggressive hemangioma of the thoracic spine and progressive paraplegia. Histology is from surgical biopsy of the mass. Haematoxylin & Eosin stain, 40x magnification. Thin-walled blood vessels of various size (asterisks) filled with blood and serous fluid extend through the bony trabeculae (arrows) of the vertebral body, replacing the normal marrow; consistent with osseous hemangioma.

Etiology	Debated. Likely multifactorial.		
Incidence	10-12% of adult population		
Gender ratio	Slightly more common in women		
Age predilection	Middle aged adults		
Risk factors	Rarely associated with chromosomal abnormalities		
Treatment	Embolization, Sclerotherapy, Radiation therapy, Surgery, and Vertebroplasty		
Prognosis	Variable, depending on degree of spinal cord involvement.		
Findings on imaging	• Radiography: Thick coarsened vertical trabeculae in a "corduroy" or "honeycomb" pattern.		
	• CT:		
	Hypodense lesion centered in vertebral body.		
	Sparse, thickened trabeculae surrounded by hypodense fat.		
	"Polka-dot" appearance on axial images.		
	Avid contrast enhancement.		
	• MRI:		
	T1WI: iso- to hypo-intense with avid contrast enhancement.		
	T2WI: Hyperintense.		
	May have extraosseous soft tissue extension and/or pathologic fracture.		

Table 1: Summary table for aggressive vertebral hemangioma

	СТ	MR T1 and T2 Weighted	Gadolinium enhanced MRI
Hemangioma	 Hypodense lesion centered in vertebral body. Sometimes expansile. Sparse, thickened trabeculae surrounded by hypodense fat. "Polka-dot" appearance on axial images. "Corduroy" appearance on sagittal and coronal plane Avid enhancement. 	 T1WI: Iso- to hypo-intense T2WI: Hyperintense. May have extraosseous soft tissue extension and/or pathologic fracture. 	Avid enhancement.
Osseous metastases	 Can be lytic or blastic, usually differentiated from hemangioma by destruction of the trabeculae. Variable enhancement 	 T1WI: Iso- to hypo-intense T2WI: Usually hyper-intense. May have extraosseous soft tissue extension and/or pathologic fracture. 	Variable enhancement.
Lymphoma	 Homogeneous, slightly dense mass. Permeative bone destruction. Homogeneous enhancement 	T1WI: HypointenseT2WI: Hypo- to hyper-intense.	Diffuse, uniform enhancement.
Paget disease	 Expansile mass. Disorganized trabeculae. Cortical thickening Significant marrow enhancement in active phase. 	 T1WI: Hypointense cortex. Hypointense marrow in active phase, hyperintense in mixed phase. T2WI: Hypointense cortex. Hyperintense marrow in active and mixed phases. 	Enhancement in active phase.

Table 2: Differential diagnosis for aggressive vertebral hemangioma

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ABBREVIATIONS COPD - chronic obstructive pulmonary disease

CT - computed tomography DSA - digital subtraction angiography MRI - magnetic resonance imaging

STIR - short T1 inversion recovery

KEYWORDS

Aggressive hemangioma; Vertebral body; Cord compression