

Radiology Case Reports

Volume 9, Issue 4, 2014

Epithelioid hemangioma of the spine: Two cases

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We report two cases of epithelioid hemangioma (EH) manifested in the thoracic spine with associated clinical, radiographic, and pathological findings. Epithelioid hemangioma is a benign vascular tumor that can involve any bone (including the spine in a subset of patients). Although recognized as a benign tumor by the WHO, it can display locally aggressive features. Within the spine, these features may lead to pain, instability, and/or neurologic dysfunction. The radiographic appearance is most typically that of a lytic, well-defined lesion on plain film or CT. The MRI appearance is typically hypointense on T1WI, hyperintense on T2WI, and avidly enhancing, often with an extraosseous soft-tissue component.

Case report 1

A 49-year-old Caucasian male patient presented to the emergency department complaining of back pain, bilateral lower-extremity weakness, numbness, and poor balance that had progressively worsened over 3 months. This recently necessitated use of crutches in order to ambulate.

Imaging findings

Initial radiographic survey of the thoracic spine was unremarkable, with the exception of mild degenerative changes (Fig. 1). Subsequent MRI revealed a solid, relatively well-circumscribed mass in the left posterolateral aspect of the T6-T7 epidural space (Fig. 2). The mass was hypointense on T1WI and hyperintense on T2WI, and demonstrated slightly reduced diffusion with a moderately elevated apparent diffusion coefficient. There was extraosseous extension through the left neural foramen. An intraosseous component of the mass was present in the left transverse process of T6 and the superior facet of T7. The mass demonstrated homogeneous avid enhancement as well as reactive enhancement of the adjacent dura.

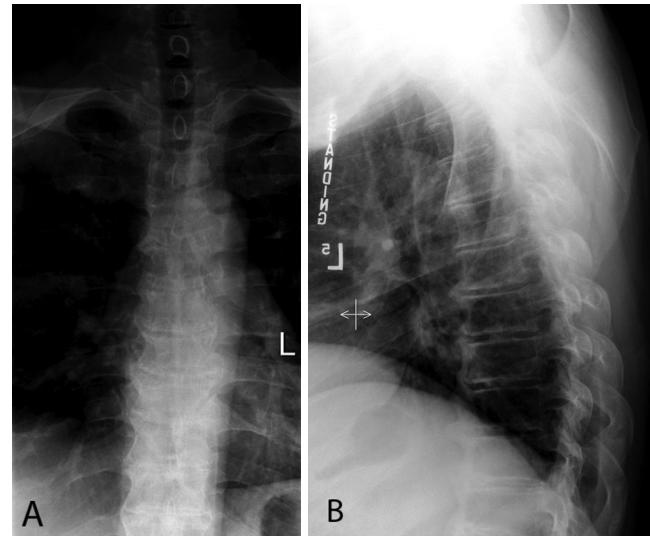


Figure 1. 49-year-old male with thoracic spine mass. AP and lateral radiographs are unremarkable, with the exception of mild degenerative changes.

Management

The possibility of lymphoma was entertained, and subtotal extradural tumor excision with T5-7 laminectomy was performed. The operative note described a well-demarcated mass under the lamina of T6 extending superiorly. The surgical procedure was uncomplicated, and postoperatively the patient had significantly improved neurological function. Given subtotal resection, the patient subsequently completed radiation therapy to the surgical bed of 50 Gy without complication.

Citation: O'Shea BM, Kim J. Epithelioid hemangioma of the spine: Two cases. *Radiology Case Reports*. (Online) 2014;9(4):984.

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Competing Interests: The authors have declared that no competing interests exist.

DOI: 10.2484/rccr.v9i4.984

Epithelioid hemangioma of the spine: Two cases

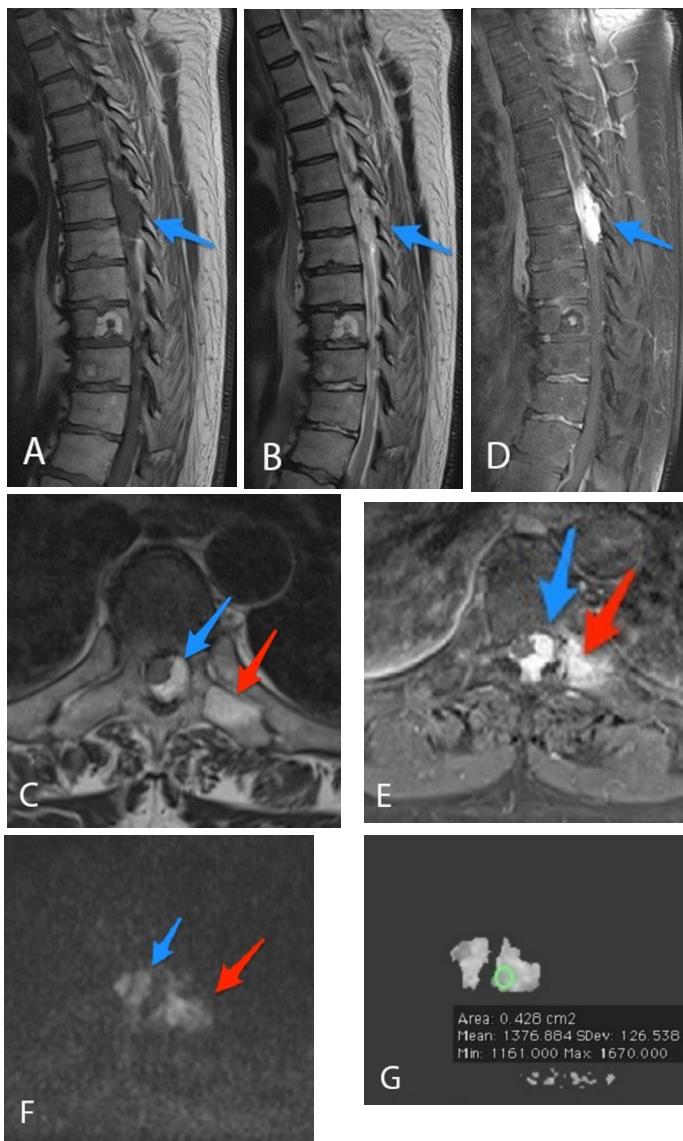


Figure 2. 49-year-old male with thoracic spine mass. Sagittal T1, T2, axial T2, sagittal, and axial fat-saturated postcontrast, B-1000 weighted DWI images, and ADC map of the thoracic spine demonstrate a T1-hypointense, T2-hyperintense, avidly enhancing mass with mildly restricted diffusion extending through the T6-T7 posterolateral epidural space into the left neural foramina. There is involvement of the left transverse process of T6 and superior facet of T7. Blue arrows denote the epidural component. Red arrows denote the intraosseous component.

Pathology

Histopathology demonstrated lobules of numerous small, capillary-sized vessels lined by plump, epithelioid endothelial cells with abundant eosinophilic cytoplasm. Rare mitoses were seen. No significant atypia was identified. Immunohistochemical staining for CD31 and CD34 was positive

in the tumor cells and highlighted the vascular channels. Pankeratin was negative. Final pathology diagnosis was epithelioid hemangioma.

Followup

Followup MRI imaging (Fig. 3) demonstrated residual intra- and extraosseous masslike enhancement, which was stable from an immediate postoperative exam. The patient was able to ambulate without a cane and returned to work.

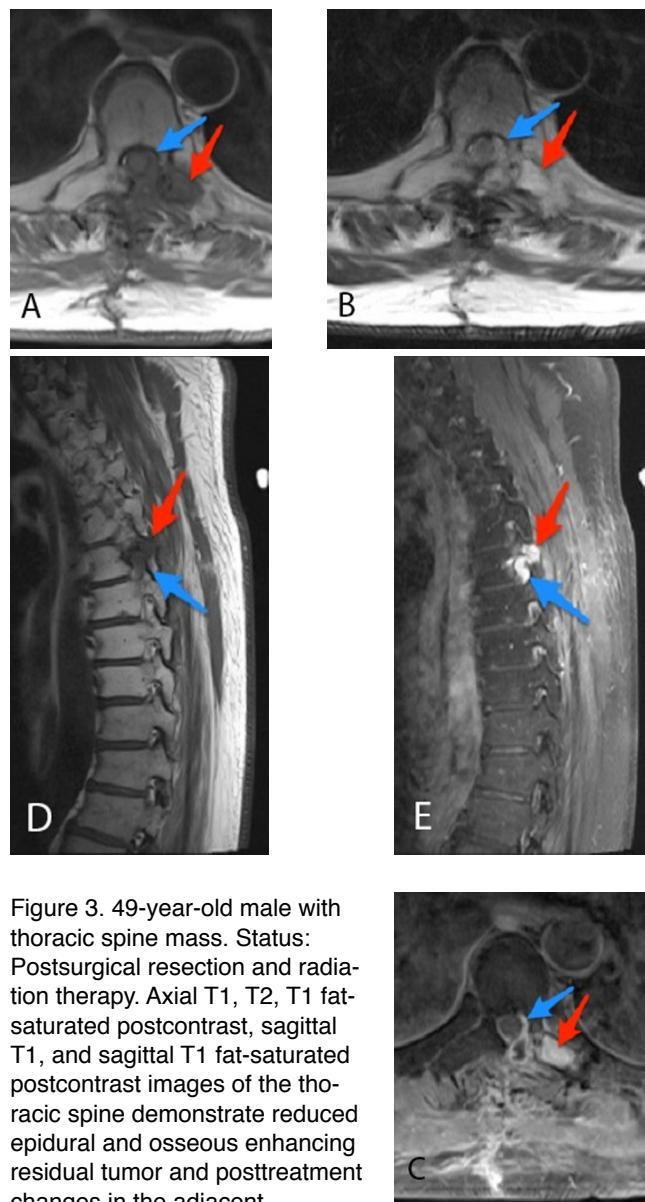


Figure 3. 49-year-old male with thoracic spine mass. Status: Postsurgical resection and radiation therapy. Axial T1, T2, T1 fat-saturated postcontrast, sagittal T1, and sagittal T1 fat-saturated postcontrast images of the thoracic spine demonstrate reduced epidural and osseous enhancing residual tumor and posttreatment changes in the adjacent tissue. Blue arrow denotes epidural mass extending through the left neural foramina. Red arrow denotes the intraosseous tumor.

Epithelioid hemangioma of the spine: Two cases

Case report 2

A 50-year-old Caucasian male patient presented with chest tightness for the past 2 months that worsened with deep breathing. The patient had no complaints of pain or neurologic dysfunction.

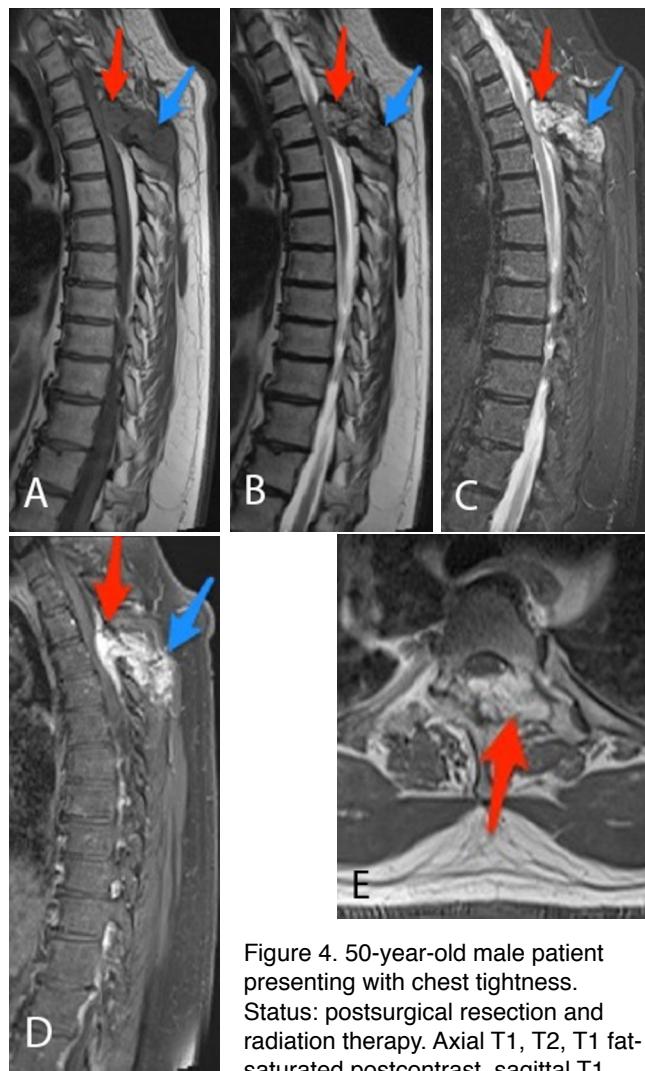


Figure 4. 50-year-old male patient presenting with chest tightness. Status: postsurgical resection and radiation therapy. Axial T1, T2, T1 fat-saturated postcontrast, sagittal T1, and sagittal T1 fat-saturated postcontrast images of the thoracic spine demonstrate reduced epidural and osseous enhancing residual tumor and post-treatment changes in the adjacent tissue. Blue arrow denotes epidural mass extending through the left neural foramina. Red arrow denotes the intraosseous tumor.

Imaging findings

Initial MRI demonstrated an expansile osseous mass with a well-demarcated lobular margin involving the left transverse process, pedicle, and spinous process of T3 (Fig. 4). An extraosseous portion of the mass extended into the dorsal epidural space and caused compression of the spinal cord. The tumor was isointense to muscle on T1WI, heterogeneously hyperin-

tense with multiple low-signal-intensity septations on T2WI, and had avid postcontrast enhancement. On contrast-enhanced CT, the tumor demonstrated generally homogeneous enhancement of the epidural portion with focal areas of hypo-attenuation (Fig. 5). On noncontrast CT, the tumor demonstrated an indistinct zone of transition with a weakly sclerotic margin and multiple internal hyperdense septations. Differential considerations of plasmacytoma, metastasis, and atypical hemangioma were entertained. Subsequent CT-guided biopsy produced a non-diagnostic sample containing only blood elements (Fig. 6).

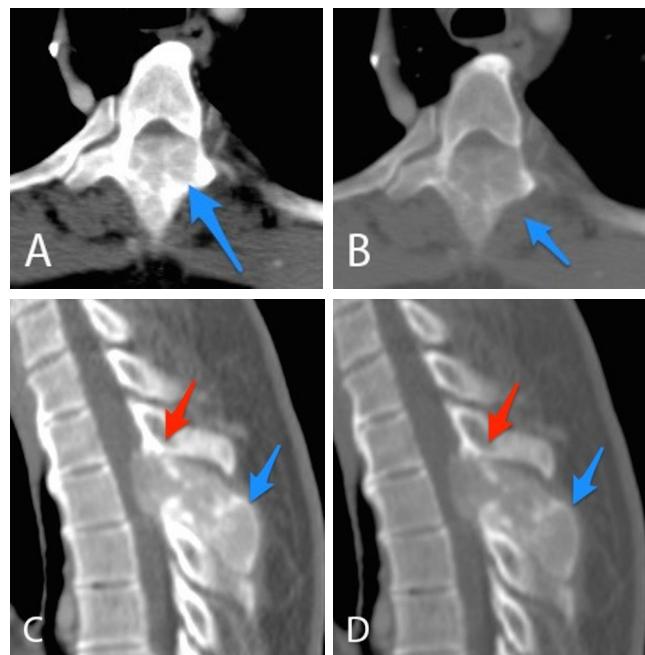


Figure 5. 50-year-old male patient presenting with chest tightness. Axial and sagittal postcontrast CT of the thoracic spine in soft tissue and bone windows demonstrate the expansile mass arising from the posterior elements of T3, with extraosseous component involving the dorsal epidural space. The mass has avid enhancement at the epidural portion and subtle globular foci of low-density and high-density septations. Blue arrows denote the intraosseous mass. Red arrows denote the epidural component.

Management

The patient underwent T3 laminectomy and debulking surgery. Intraoperatively, the spinous process of T3 was markedly expanded by the tumor, with multiple feeding vessels infiltrating the bone. The entire tumor could not be removed. The patient subsequently underwent radiation therapy with 50 Gy to the resection bed. Postoperative imaging demonstrated posttreatment bed enhancement without discrete mass or new destructive changes (Fig. 7).

Epithelioid hemangioma of the spine: Two cases

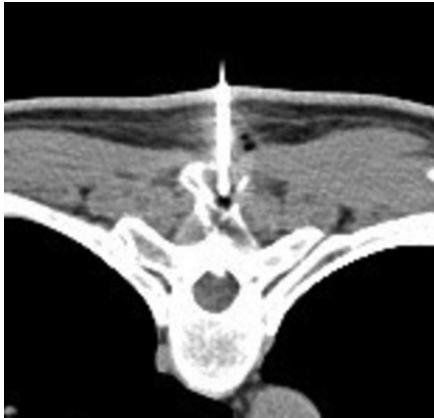


Figure 6. 50-year-old male patient presenting with thoracic spine mass. Axial noncontrast CT of the thoracic spine for CT-guided biopsy again demonstrates that the T3 mass has irregular indistinct zone of transition with a weakly sclerotic margin and multiple internal septae. The expansile portion of the spinous process was targeted.

Pathology

Surgical pathology demonstrated a variably cellular tumor composed of capillary-to-slightly-larger-sized vessels lined by a single layer of plump, epithelioid endothelial cells with abundant eosinophilic cytoplasm in a loose, paucicellular collagenized background. Rare mitoses were seen, and there was only mild cellular atypia. Immunohistochemical staining for CD34 and FLI-1 showed positivity supporting endothelial differentiation of the cells. Very infrequent endothelial cells showed positivity for Pankeratin. The final pathologic diagnosis was epithelioid hemangioma.

Discussion

Etiology and demographics

Epithelioid hemangioma (EH) is a mesenchymal tumor of vascular origin. First recognized as a distinct entity in 1983, EH has been previously described as angiolympoid hyperplasia with eosinophilia, or histiocytoid hemangioma (1, 2). Although the World Health Organization recognizes EH as a distinct neoplasm, it can be difficult to differentiate histologically from endothelioid hemangioendothelioma (1, 2). EH arises most frequently in the skin and subcutaneous tissues of the head, neck, and distal extremities. Osseous EH has been reported most commonly in the tubular bones of the extremities (1). The presence of osseous EH involving the spine is less common, with a reported frequency of 11-20% (1, 3, 4).

The age of presentation for EH of the bone is wide (7-75 years), with a mean of 35-39 years (2, 4). A slight male predominance has been reported at 1.4:1 in the osseous form of the disease (2). EH is reported to be found in multiple bones on presentation in 18-22% of cases (2, 4). Multiplicity was mostly found in reports of EH involving interphalangeal bones (4). No clear genetic association has been

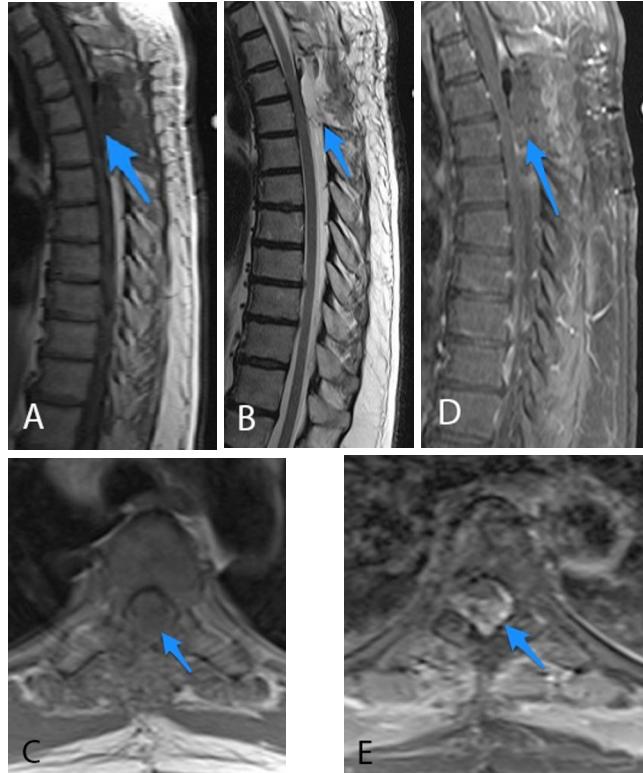


Figure 7. 50-year-old male presenting with thoracic spine mass. Status: post thoracic resection and radiation therapy. Sagittal T1, T2, axial T1, and sagittal and axial T1 fat-saturated postcontrast images of the thoracic spine demonstrate residual epidural enhancing tumor with post-treatment changes in the adjacent tissues. Blue arrow denotes the residual mass.

reported with EH, in contrast to the reported genetic translocation and fusion anomalies associated with epithelioid hemangioendothelioma (5, 6).

Clinical & imaging findings

As in our two cases, EH of the spine may have aggressive local features and may present with pain or neurological impairment secondary to instability. An extraosseous component may be present, which can significantly contribute to neurological compromise, given the rigid anatomical boundaries of the spinal canal. Osseous destruction by EH can cause instability or pathologic fractures, also leading to neurologic impairment. This can lead to an acute or chronic presentation (1).

In contrast to the well-known capillary or cavernous hemangioma, the radiographic appearance of epithelioid hemangioma does not have a typically near-pathognomonic appearance (1-4). Most skeletal EH lesions are lucent, with well-defined margins on plain films and CT. This is also true of vertebral body lesions. Soft-tissue extension beyond the osseous lesions, as in our cases, has also been reported by multiple sources (1, 3). Bone-scan uptake may be increased or normal (1, 4). MRI signal

Epithelioid hemangioma of the spine: Two cases

characteristics are typically hypo- to isointense on T1WI, hyperintense on T2WI, with avid postcontrast enhancement (4).

Treatment & prognosis

Optimal treatment management of EH within the spine is still an area of research. A combination of en bloc resection, curettage, pre/postoperative embolization, radiation, and observation has been used with success (1). Local recurrence was reported in 11% of patients, in a series of 36 patients (2). Imaging surveillance is often recommended for this reason. In addition, lymph-node involvement in the setting of osseous disease has also been reported in rare instances (2, 6).

In our series of two cases, surgical management established histopathological diagnosis and rendered symptomatic improvement in pain and neurologic impairments. Postoperative radiation therapy was advised because of residual extraosseous tumor identified either intraoperatively or on radiologic followup.

Differential diagnoses

EH overlaps with multiple other histopathological variants of hemangiomas of the spine, including cavernous, capillary, arteriovenous, venous, and spindle-cell (1). Of these, capillary and cavernous variants represent the vast majority of osseous hemangiomas (2). The various histopathologic variants of hemangiomas may present as an “aggressive hemangioma,” with destruction of the adjacent bone cortex and soft-tissue extension. Within the spectrum of epithelioid-type vascular tumors, epithelioid hemangioendothelioma and epithelioid angiosarcoma have overlapping imaging characteristics with EH. However, these tumors are considered malignant, the former being low-moderate and the latter high-grade malignancies (2). Metastasis and plasmacytoma are additional tumors with locally destructive appearances and are within the imaging differential. Thus, the radiologic differential of EH is broad and includes other benign vascular tumors of the skeletal system as well as malignant tumors.

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