

Intraosseous hemangioma of the rib mimicking an aggressive chest wall tumor

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ABSTRACT

Bone hemangiomas are extremely rare in the ribs, with only a handful of cases reported in the literature. A case of a rib hemangioma is presented in which the pre-resection diagnosis was an aggressive chest wall tumor. The plain film, CT, MRI and bone scan features of the lesion were reviewed with the pathological correlation. On imaging, the lesion was expansile and lytic, and it also had fine bony trabeculae. The lesion also demonstrated growth beyond a disrupted bony cortex, suggesting malignancy. This case report adds to the literature on this rare condition and discusses the issues in the diagnosis of chest wall tumors.

Key words: • hemangioma • ribs • computed tomography, x-ray • magnetic resonance imaging

Rib hemangiomas are rare (1–3); this diagnosis is often forgotten in the differential diagnosis of a chest wall tumor. Half of primary chest wall tumors are malignant; hence, accurate diagnosis is imperative (2). Hemangiomas occurring in the ribs are extremely rare, with only a handful reported in the literature (1, 2, 4). This case report adds to the literature on this rare condition and discusses the issues in the diagnosis of chest wall tumors.

Case report

A 20-year-old male had a chest x-ray showing an incidental asymptomatic large expansile lesion of the right 5th rib (Fig. 1).

Computed tomography (CT) showed a well-defined, 5-cm expansile lytic rib lesion with soft tissue extension (Fig. 2). Near the costovertebral junction, there was cortical disruption in the rib. Fine bony trabeculae were seen in the soft tissue extension as well as in the rest of the expanded 5th rib. The soft tissue component also had heterogeneous density. There was no periosteal reaction. The lesion was solitary, and there was no pleural effusion.

On magnetic resonance imaging (MRI), the lesion had a heterogeneous intermediate signal on the T1-weighted images, a heterogeneous high signal on fat suppressed T2-weighted images and enhancement with gadolinium (Fig. 3). At the time of the initial report, there was a finding that was not appreciated as significant. There were small, scattered regions within the expansile component of the lesion that had the signal characteristics of fat (Fig. 4). This finding was noted on reviewing the MR images post-resection, after the diagnosis had been confirmed.

The bone scan showed that the lesion was solitary. There was increased activity on the blood pool images and only mildly increased activity on the delayed images (Fig. 5).

The radiological diagnosis was an aggressive, primary bone neoplasm, such as Ewing sarcoma. The lesion was amenable to surgical resection, and a pre-resection biopsy was not performed.

On inspection, the resected specimen showed a smooth, hemorrhagic nodule covered by the periosteum and protruding from the concave part of the rib (Fig. 6a). Post-decalcification, the cut surface was spongy and dusky, with the marrow space extensively replaced by soft, hemorrhagic tissue (Fig. 6b). Low-power microscopy revealed that the bone marrow had been replaced by numerous, thin-walled blood vessels with dilated channels (Fig. 6c). There was no cytologic atypia. The diagnosis was a benign hemangioma.

Discussion

Bone hemangiomas are benign hamartomatous vascular tumors. They account for less than 1% of all bone neoplasms (1).

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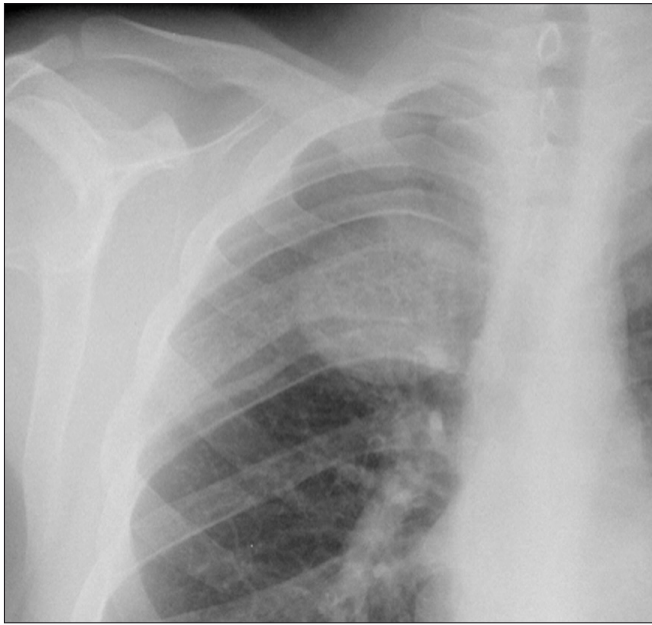


Figure 1. A chest radiograph of a 20-year-old man showed an incidental, asymptomatic expansile lesion of the right 5th rib with an associated soft tissue component.

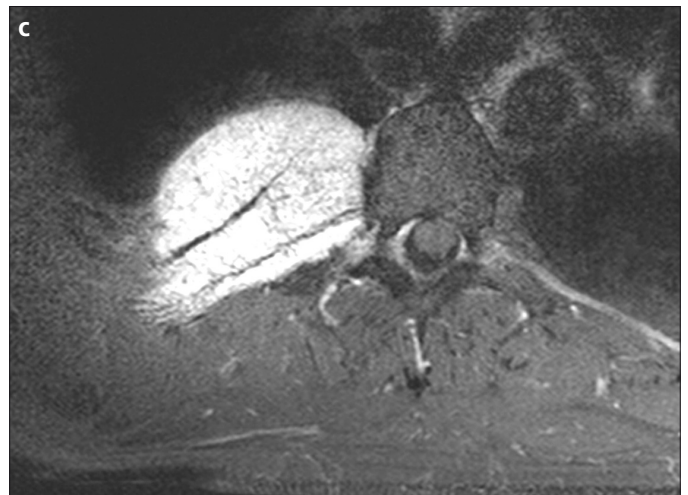
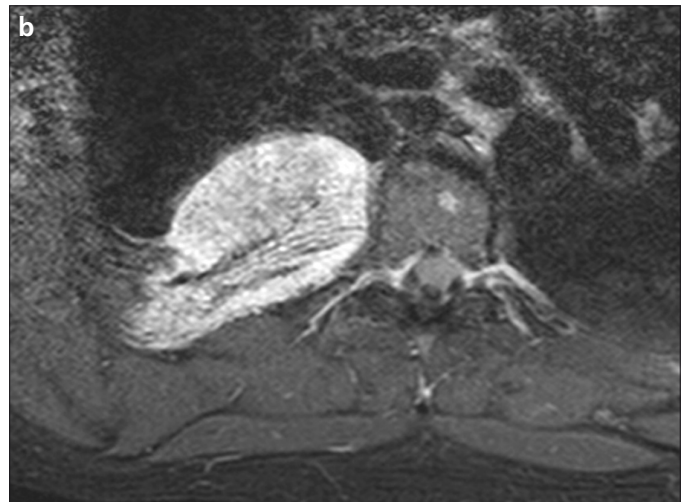
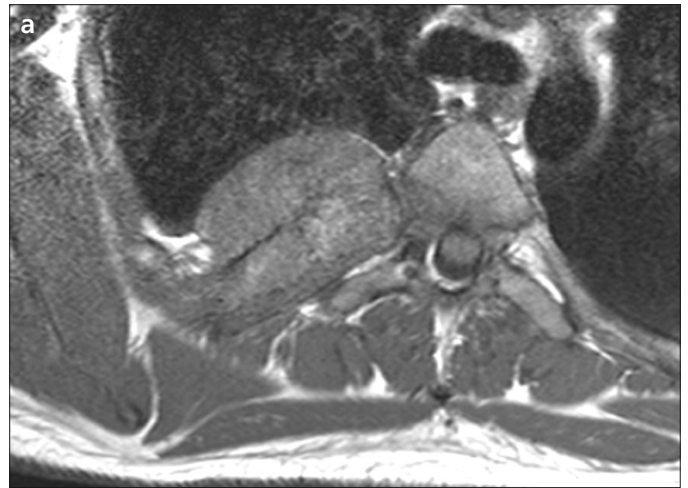


Figure 2. a, b. Bone window (a) and soft tissue window (b) CT images of the posterior right 5th rib showed a bony lesion with a cortical disruption and a well-defined expansile soft tissue component. Fine bony trabeculae and heterogeneous density could be appreciated in the expansile component. There was no periosteal reaction.

Figure 3. a–c. MR images of the posterior 5th rib showed the extraosseous component of the lesion. The expanded medullary cavity of the rib had a heterogeneous intermediate signal on T1-weighted images (a), a heterogeneous high signal on fat suppressed T2-weighted images (b), and an increased enhancement with gadolinium (c).

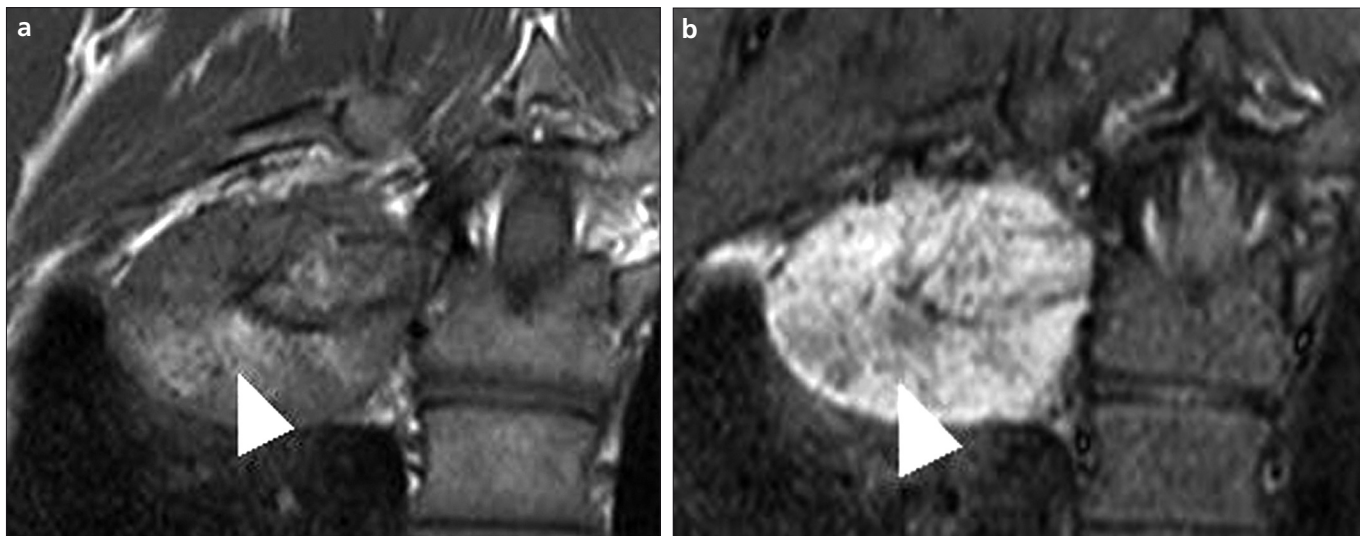


Figure 4. a, b. Coronal T1-weighted MR image (a) shows that within the predominantly low T1 signal expansile component, there are ill-defined central regions of high signal (arrowhead) that become low signal on fat-suppressed T2-weighted images (b). This finding suggests the presence of fat, which is often found in hemangiomas.



Figure 5. The delayed images of the bone scan showed mildly increased activity in the solitary lesion in the posterior half of the right 5th rib.

Bone hemangiomas occurring in the ribs are extremely rare. A review of 14 previously reported cases of rib hemangiomas noted that the majority were solitary lesions (5). There was only one case report in which a rib hemangioma was found with multiple other hemangiomas in a patient who had hemangiomatosis (4). Most cases were diagnosed in older patients. Three patients were between 11 and 25 years of age; all other patients were between 45 and 74 years of age. Half of the 14 cases were reported as asymptomatic.

On histology, rib hemangiomas are usually of the cavernous type, with large, dilated vessels lined with a single layer of epithelial cells surrounded by a fibrous stromal layer (6, 7). Non-vascular tissues (e.g., fat, smooth muscle, bone trabeculae, fibrous tissue and clotted blood products) may exist in the matrix between the vascular spaces.

Rib hemangiomas in imaging data are characteristically expansile and well circumscribed, with a thin, intact bony cortex, fine bony trabeculae and no associated pleural effusion (8). The

arrangement of fine bone trabeculae in a rib hemangioma can give a “honeycomb” appearance to the medullary cavity of the rib (3, 7). Expansile growth by a rib hemangioma beyond the disrupted bony cortex, a finding that suggests an aggressive lesion, has been previously described in only a few cases (3, 5, 9). Although rib hemangiomas may mimic aggressive lesions in imaging data, they are usually still asymptomatic (3, 4, 9).

By MRI, rib hemangiomas have low to intermediate signal on T1-weighted images, high signal on T2-weighted images and avid enhancement with gadolinium (9). The presence of fat can be demonstrated with MRI by using fat suppression protocols (9).

On bone scans, rib hemangiomas usually show increased activity on blood pool images and mildly increased activity on delayed images (9).

Because hemangiomas rarely occur in the ribs, they are often misdiagnosed. The differential diagnosis of a rib lesion includes primary tumors and metastatic lesions. Examples of primary malignant bone tumors include myeloma, chondrosarcoma, osteosarcoma and Ewing sarcoma (5). Benign primary rib lesions include osteochondroma, enchondroma, fibrous dysplasia, eosinophilic granuloma and aneurysmal bone cysts.

Some rib lesions, such as fibrous dysplasia, aneurysmal bone cysts and osteochondroma, may have characteristic imaging findings that allow a specific diagnosis (6). When a pleural effusion

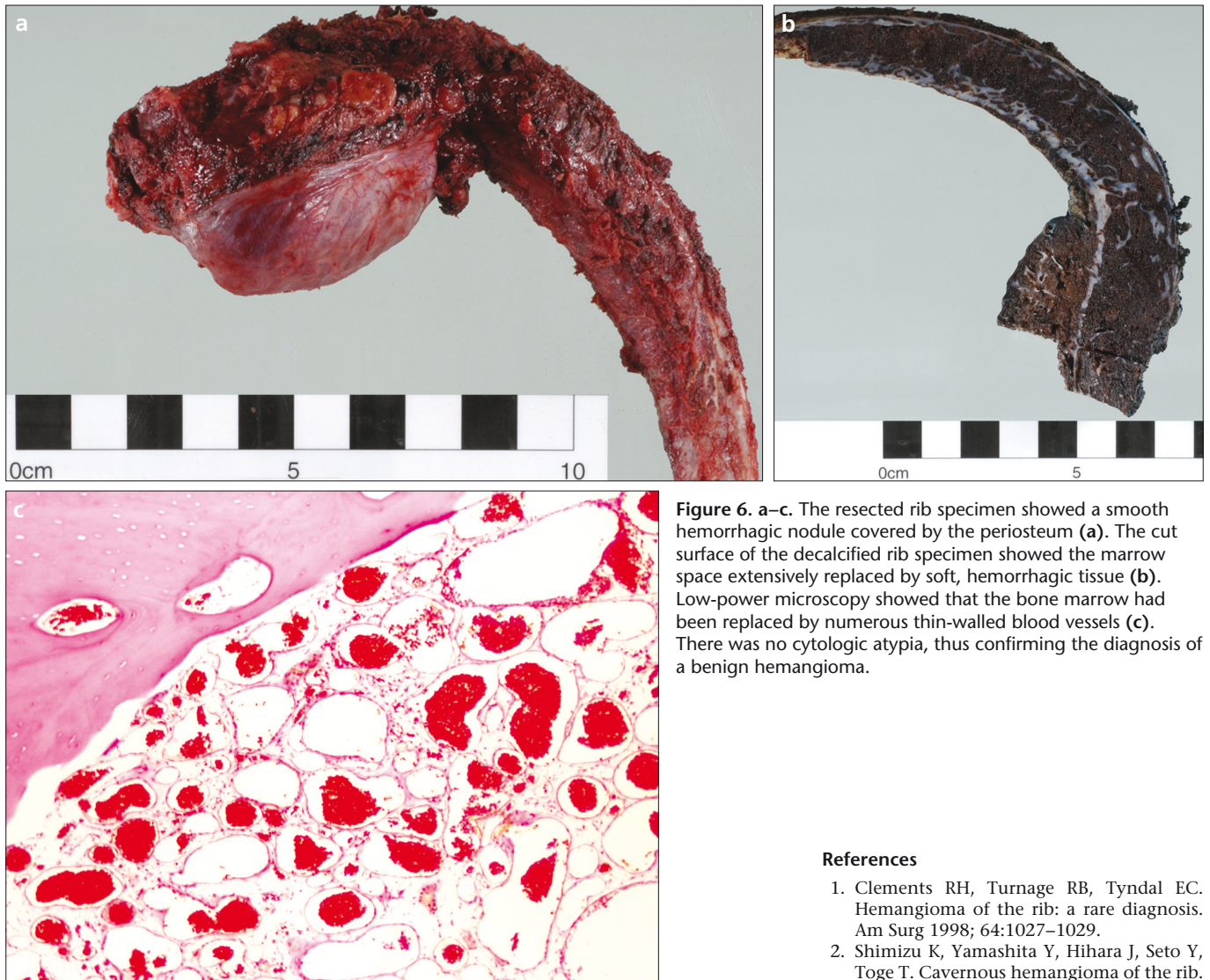


Figure 6. a–c. The resected rib specimen showed a smooth hemorrhagic nodule covered by the periosteum (a). The cut surface of the decalcified rib specimen showed the marrow space extensively replaced by soft, hemorrhagic tissue (b). Low-power microscopy showed that the bone marrow had been replaced by numerous thin-walled blood vessels (c). There was no cytologic atypia, thus confirming the diagnosis of a benign hemangioma.

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is found with an aggressive-looking rib lesion in a young person, Ewing sarcoma or an active inflammatory process needs to be excluded (8). Pain is not a reliable predictor of malignancy (1). About half of primary chest wall tumors are malignant; hence, accurate diagnosis is imperative (2). Preoperative diagnosis of a chest wall tumor by imaging alone is often not possible, and definitive diagnosis by biopsy or resection may be required (3).

With regard to deciding on a biopsy versus a resection of a solitary rib lesion, a biopsy (fine needle, core or open) of a hemangioma can result in significant bleeding (9). Moreover, many bone tumors are inhomogeneous on histological examination; hence, studying small samples (as opposed to a wholly excised specimen) can be misleading (7). A complete resection of the rib lesion, if feasi-

ble, may be the best option once other investigations confirm that the lesion is solitary. Some other management options for symptomatic hemangiomas include radiotherapy, transarterial embolization and alcohol injection (9).

In conclusion, the diagnosis of a hemangioma should be considered in cases of an asymptomatic expansile rib lesion, especially if there are radiating bone trabeculae and if MRI suggests the presence of fat within the lesion. A rib hemangioma can also show cortical disruption, which can give the false impression of aggressive behavior. A biopsy or resection may be required to establish the diagnosis because a significant proportion of primary rib lesions are malignant. A biopsy, however, can result in significant bleeding, which the person performing the procedure needs to be prepared to manage.