Primary mediastinal giant cell tumor

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Abstract

Giant cell tumor of soft tissue is a rare tumor first described by Salm and Sissons in 1972 as being a distinct entity.[1] Histologically, it is composed of multinucleated giant cells dispersed among mononuclear cells, and is indistinguishable from its bone equivalent.[2] The majority of these tumors have been reported to occur in the lower extremity.[2,3] We describe a case of giant cell tumor of soft tissue within the posterior mediastinum. The only other report of a primary mediastinal giant cell tumor of soft tissue in the English literature was published by Fu *et al.* in 2002, in which they described two patients with posterior mediastinal masses.[4]

Case Report

A 53-year-old man with a history of hypertension presented with increasing left anterior chest pain. He was seen by a cardiologist and had a treadmill stress test, which was normal. A chest radiograph revealed a large left mediastinal mass (Figure 1). A follow-up non-contrast chest CT scan showed a large left posterior mediastinal mass with compression of the adjacent lung (Figure 2). There was pressure erosion and sclerosis of the left posterior fifth rib, suggestive of a slowly growing mass. The mass contained areas of low density suggesting necrosis. It was unclear on the CT scan if the mass was arising from the rib, pleura, or soft tissue. One week later, the patient had a bronchoscopy, which was negative for malignant cells. A PET-CT scan showed patchy hypermetabolic uptake within the mass consistent with malignancy without evidence of metastatic disease. A CT-guided core biopsy of the mass was performed, which revealed a giant cell tumor.

A left lateral thoracotomy was performed with gross resection of the tumor via a left lower lobectomy and partial left chest wall resection. Gross examination revealed a 13 cm mass within the extra-pleural left chest wall with compression of the adjacent lung and ribs (Figure 3). Fragments of the fourth, fifth, sixth, and eighth ribs showed no evidence of tumor. Microscopic examination demonstrated the mass to be composed of a mixture of round and oval mononuclear and multinucleated osteoclast-like giant cells with a background of rich vascular stroma (Figures 4 and 5). The tumor also showed areas of cystic degeneration and hemorrhage. There was no increase in the mitotic index. Immunohistochemically, the mass was positive for CD68 and CD34, and was diagnosed as a giant cell tumor of soft tissue. Five mediastinal lymph nodes were resected without evidence of malignancy. The patient had an uneventful postoperative course and received postoperative radiation treatment to the left chest wall. A follow-up non-contrast chest CT scan did not show evidence of recurrent disease or metastases.

Discussion

Giant cell tumor of soft tissue is a rare tumor, which demonstrates a spectrum of benign to malignant characteristics. Microscopically, these tumors are composed of multinucleated giant cells evenly dispersed among mononuclear cells, and are histologically indistinguishable from their bone equivalent.[2] In 1972, Salm and Sissons were the first to describe a series of 10 patients with giant cell tumors of soft tissue.[1] In their study, the tumors demonstrated benign features without metastases. However, there was local recurrence in two patients. In the same year, Guccion and Enzinger reported a series of 32 patients with soft tissue tumors similar to those studied by Salm and Sissons, but with malignant features and widespread metastases.[3] These tumors were called "malignant giant cell tumors of soft parts." In 2000, O'Connell *et al.* described a series of 18 patients with giant cell tumors of soft tissue that demonstrated a spectrum from benign to malignant behavior determined by the cytolog-

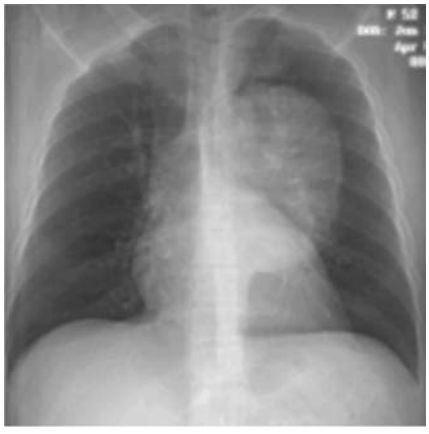

Figure 1. Frontal chest radiograph demonstrating a large left mediastinal mass.

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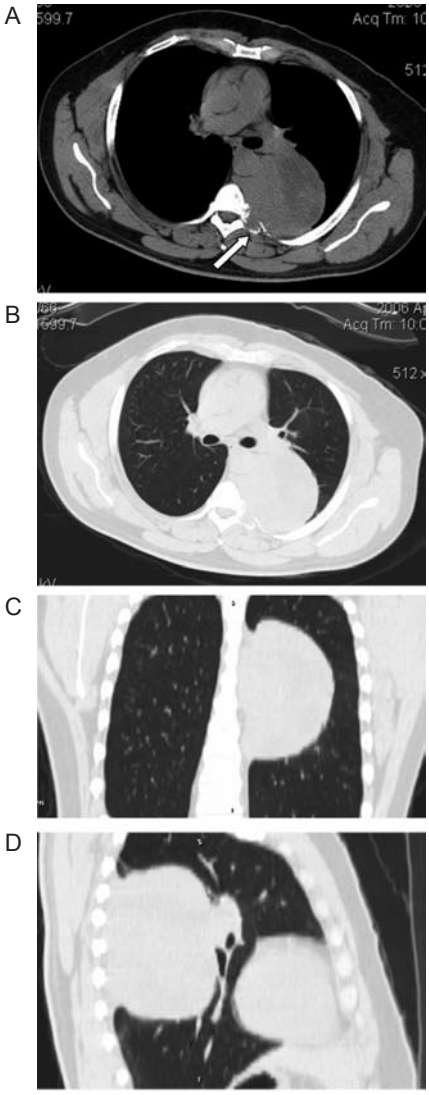

Figure 2. Non-contrast axial computed tomography scan showing A: soft tissue, and B: lung windows; C: coronal, and D: sagittal reformations. There is a large heterogeneous left posterior mediastinal mass with sharp lateral margins causing compressive atelectasis of the left lower lobe. There is pressure erosion of the adjacent posterior rib (A: arrow).

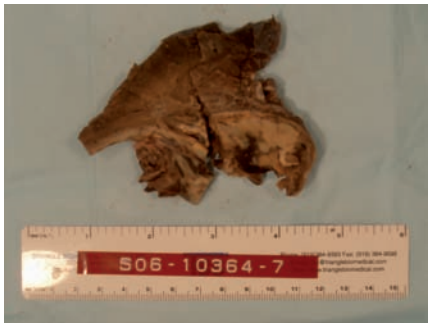

Figure 3. Gross specimen of the giant cell tumor of soft tissue.

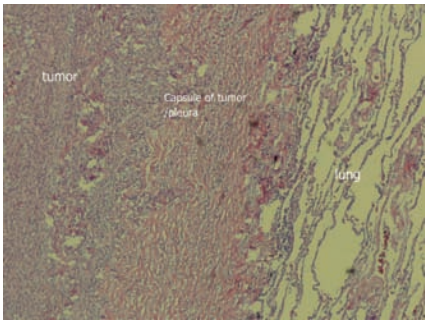

Figure 4. Low-power microscopic examination demonstrating the tumor, tumor capsule, and uninvolved lung.

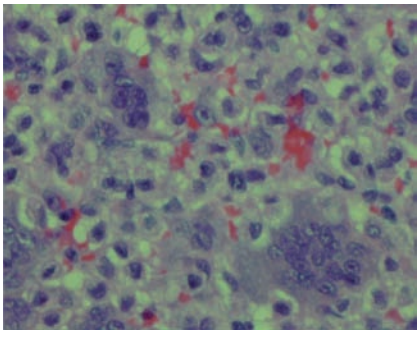

Figure 5. Microscopic examination at 600X magnification demonstrating the mass to be composed of a mixture of round and oval mononuclear and multinucleated osteoclast-like giant cells.

ical appearance of the mononuclear cells and mitotic activity.[5]

Giant cell tumors of soft tissue can be seen at any age, but usually affect middle-aged adults of both sexes. These tumors can occur in both the superficial and deep soft tissues. The majority of these tumors reported in the literature have been located in the lower extremity, particularly in the thigh.[2,3] Other sites of involvement include the upper extremities, trunk, and rarely the skin.[6] The most common clinical presentation is a painless soft tissue mass.[5]

In a series of 22 patients with giant cell tumor of soft tissue, Oliveira *et al.*[2] described the recurrence rate to be 6.2%, which was less than the 25% reported recurrence rate of giant cell tumors of bone.[1] However, the metastatic and death rates were determined to be higher in giant cell tumors of soft tissue.[1] Immunohistochemically, the giant cell tumor of soft tissue in this case report expressed CD68 and CD34. Conversely, the giant cell tumors of soft tissue reported in the literature expressed only CD68.

This case report illustrates a primary giant cell tumor of soft tissue located within the posterior mediastinum. The only other report of a primary mediastinal giant cell tumor of soft tissue in the English literature was published by Fu *et al.* in 2002, in which they described two patients with posterior mediastinal masses.[4] Our case is very similar to those described by Fu *et al.*, as the cells all had low mitotic activity and the patients had a favorable clinical course without metastases or recurrence of the tumor.

In conclusion, giant cell tumors of soft tissue are rare tumors that demonstrate a spectrum of benign to malignant potential. Although the majority of these tumors are located in the lower extremity, they may occur rarely in the posterior mediastinum and present as a posterior mediastinal mass.

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