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Case Report
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Primary Mediastinal Giant Cell Tumor: Case Report and Review of Literature

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Abstract

Giant cell tumor of soft tissue is a rare non-cancerous, however aggressive tumors, first described in 1972. The majority of them have been described to occur mainly in the lower extremity, particularly within the thigh, in both the superficial and the deep soft part. The most frequent clinical presentation is a painless soft tissue mass and both sexes are equally affected, at any age. In this study, we describe a case of 20-year-old lady, with giant cell tumor of soft tissue within the posterior mediastinum, which is an uncommon site of occurrence. At the best of our knowledge this is the sixth case described in the literature with mediastinal involvement from Giant cell tumor.

Keywords: Giant cell tumor; Soft tissue; Mediastinum; CT

Introduction

Giant cell tumor of soft tissue is a rare non-cancerous, however aggressive tumors, first described in 1972, by Salm and Sisson [1], as being a distinct entity. These tumors are even rare but have been noted in multiple anatomic sites over extremities, trunk, head and neck, superficial and deep fascia, tendon sheaths, and skeletal muscle [2,3]. Such tumors can take place in the superficial and the deep soft tissues. The majority of them have been described to occur mainly in the lower extremity, particularly within the thigh [2-4]. However, possible involvement involving the upper extremities, trunk, and rarely the skin was reported [5-7]. The most frequent clinical presentation is a painless soft tissue mass [5,6]. The major part of these tumors, as reported in the literature, can affect both sexes, at any age, but customarily act on middle-aged adults.

In this study, we describe a case of 20-year-old Saudi lady, with giant cell tumor of soft tissue within the posterior mediastinum and the appropriate literature is thoroughly reviewed.

Case Presentation

A 20-year-old lady, with no history of comorbidity, presented with history of back pain, started in the second month of her 1st pregnancy, located initially at the upper back between the shoulders, not radiating, not associated with numbness. There was no previous or recent history of fall or traumatism. The pain had been increased gradually limiting her physical activity. She went to local hospital with this complain, where he received pain killer in form of paracetamol which partially alleviate her pain. She had no associated symptoms such as fever, weight loss, night sweats or skin rash.

At the 1st week of her 9 months of pregnancy she was developed shortness of breath. She was delivered *via* cesarean section. The baby was in a good health. She had developed massive right sided pleural effusion and a chest drain was inserted. After delivery, a Computed Tomography (CT) of chest was done. It showed large heterogeneous enhancing midline mass with areas of necrosis crossing midline in the posterior mediastinum, measuring approximately 14 cm × 8 cm × 10.0 cm, encasing the descending thoracic aorta and causing anterior displacement and mass effect on the heart. There destruction of the ninth and tenth and thoracic vertebra extending into the spinal canal with encasement and compression of the cord (Figure 1). The MRI of the whole spine showed the large infiltrate posterior mediastinal mass causing destruction of vertebral bodies of T9, T10 and T11 with epidural component measures surround the spinal cord, this results in significant spinal canal stenosis resulting in cord compression with evidence of signal alteration (Figure 2). No other lesion in the spinal canal is noted.

She underwent CT guided biopsy of the paraspinal mass. The final histology analysis showed

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Table 1: Provides clinical, histological and therapeutic data of the five published cases included in the review.

Year of publication	Ref	Age	Gender	Initial presentation	Tumor size (cm)	Mitotic rate	Rib invasion	Spinal canal invasion	Treatment	Recurrence
2002	[5]	31	F	Pain and paresthesia upper limb	8	Less than 5%	No	No	resection	No
		18	M	Incidental X Ray	2.5	Less than 5%	No	No	resection	No
2009	[8]	53	M	Chest pain	13	No increase	Yes	No	resection	No
2015	[9]	28	M	Chest pain	15	High 20% to 30%	Yes	No	Debulking	No
2017	[10]	18	F	Back pain	5	High 25% to 30%	Yes	Yes	resection	Yes

F: Female; M: Male

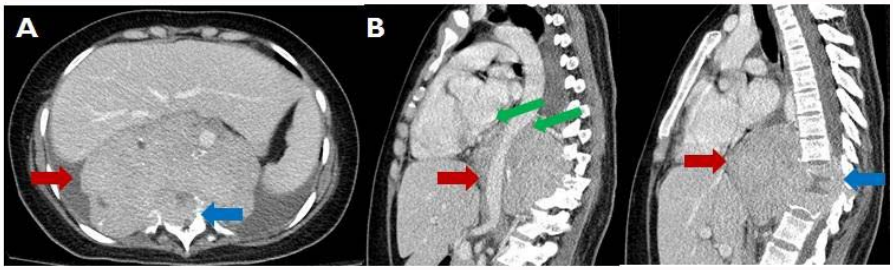

Figure 1: The Axial A and Sagittal B, CT scan images demonstrate large heterogeneous enhancing midline mass with areas of necrosis crossing midline in the posterior mediastinum (red arrow) measuring approximately 14 cm × 8 cm × 10.0 cm, encasing the descending thoracic aorta and causing anterior displacement and mass effect on the heart (green arrow). There destruction of the ninth and tenth and thoracic vertebra extending into the spinal canal with encasement and compression of the cord where (blue arrow).

blend of round and oval mononuclear and multinucleated osteoclast-like giant cells with a background of rich vascular stroma (Figure 3). Fortunately, there was no increased mitotic index. In the immunohistochemical analysis, the final diagnostic of giant cell tumor of soft tissue was proven, as the lesion was positive for *CD68* and *CD34*.

A posterior decompression surgery was done for the spinal cord with spinal fixation at level of T5 to L2.

The patient started yet on physiotherapy, after the suffering postoperative course. Dental clearance was done and we immediately started her on Denosumab 120 mg subcutaneous weakly for 4 weeks. After which we will continue with Denosumab every 28 days.

Discussion

Giant cell tumor of soft tissue are infrequent tumors that have been recognized in the literature for several decades, which demonstrates a spectrum of benign to malignant characteristics [1,2]. In the English literature, this entity was reported as distinct tumor at the first time in 1972 by Salm and Sissons [1]. In their study, they included 10 patients with giant cell tumors of soft tissue. All the cases analyzed were benign without any distant metastases. However, the authors reported two cases with local recurrence [1].

Guccion et al. [4] signed the second study in the same year and it was larger than the first, including 32 cases, but with malignant behavior and multiple metastases. Thus, it was the first description of "malignant giant cell tumors of soft tissue", in 1972 [4].

In 2000, O'Connell et al. [6] reported a relatively large series of 18 cases with giant cell tumors of soft part that revealed a spectrum from benign to malignant features determined by the cytological appearance of the mononuclear cells and mitotic index.

In the same year, Oliveira et al. [2] described a series of 22 cases with giant cell tumor of soft part. The authors compared the recurrence and the metastatic and death rates of giant cell tumors of

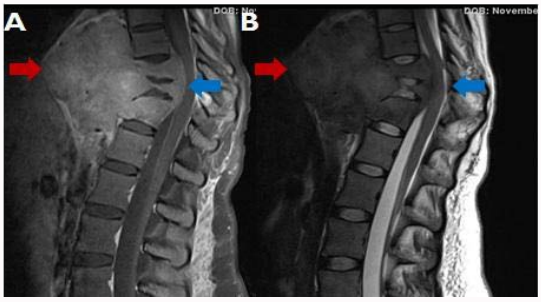

Figure 2: The sagittal enhanced T1-weighted images A and the coronal T2-weighted images B. showed the large infiltrate posterior mediastinal mass causing destruction of vertebral bodies of T9, T10, and T11 with epidural component measures surround the spinal cord, this results in significant spinal canal stenosis resulting in cord compression with evidence of signal alteration. No other lesion in the spinal canal is noted.

soft tissue and those of bone. The recurrence rate was significantly less for giant cell tumors of soft tissue estimated at 6.2% *vs.* 25% for those of bone [2]. While, the metastatic and death rates were significantly higher in giant cell tumors of soft part [2].

Giant cell tumors of soft tissue can take place in the superficial and the deep soft tissues and can affect both sexes, at any age, with predominant attract for the middle-aged adults. In our case the patient was 20-year-old lady.

The major previous described cases have been reported in the lower extremity, particularly within the thigh [2-6]. However, others sites of rare involvement have been reported in the upper extremities, trunk, and even the skin [5-7]. In this study, the giant cell tumor of soft tissue took place in the posterior mediastinum, which was infrequent site. Thus, we searched into the English literature focusing on cases of mediastinal giant cell tumor. Literature review yielded only five patients, two males and three females, reported in 2002, 2009, 2015 and 2017 [5,8-10]. The age ranged from 18 to 53 years (Table 1).

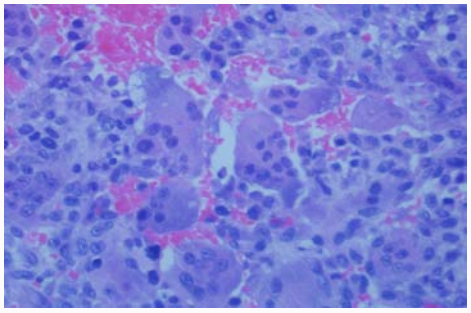

Figure 3: Microscopic examination at 600x magnification demonstrating the mass to be composed of blend of round and oval mononuclear and multinucleated osteoclast-like giant cells.

The most common clinical presentation was a painless soft tissue mass [5,8-10], as in our study. A medical imaging exam has a powerful role in detecting the exact origin and describing the concrete details of the lesion [10].

In the present study, the CT of chest showed the posterior mediastinal mass and detected the invasion of the adjacent rib, similar to that described previously by Goldberg et al. [8], Jain et al. [9] and Hu et al. [10]. There was also extension of the large lesion through the intervertebral foramen into the spinal canal, which had been described at the first time by Hu et al. [10] in 2017 and it was the first description of the MRI findings of this entity.

Conclusion

Giant cell tumors of soft tissue are very rare aggressive tumors, with spectrum of benign to malignant potential. The lower extremities, particularly the thigh remain the most common site; however, this entity can take place in the posterior mediastinum, as a painful mass. Medical imaging exams have a powerful role in detecting the exact origin and describing the concrete details of the lesion, with emphasis of MRI.

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