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ABDOMINAL SWELLING IN A YOUNG MALE- A RARE CASE OF SOFT TISSUE GIANT CELL TUMOR

Oncology/Radiotherapy

Kisa Syeda Junior Resident, Department of Radiotherapy, Jawahar Lal Nehru medical College and hospital, Aligarh Muslim University, Aligarh, Uttar Pradesh.

Faraz Badar* Senior Resident, Department of Radiotherapy, Jawahar Lal Nehru medical College and hospital, Aligarh Muslim University, Aligarh, Uttar Pradesh. *Corresponding Author

Bushra Siddqui Assistant Professor, Department of Pathology, Jawahar Lal Nehru medical College and hospital, Aligarh Muslim University, Aligarh, Uttar Pradesh.

Mohsin Khan Assistant Professor, Department of Radiotherapy, Jawahar Lal Nehru medical College and hospital, Aligarh Muslim University, Aligarh, Uttar Pradesh.

ABSTRACT

Giant cell tumor of soft tissue (GCT-ST) origin is a rare entity. Very few cases have been described, mostly in thigh, liver and thorax. Here we describe a case report of GCT-ST arising in the iliac region in a 24yr old male, who presented with a painless abdominal swelling. Initial CT scan imaging suggested a solid cystic mass in right lower quadrant of abdomen. The diagnosis of GCT was established by histopathological examination of excised specimen.

KEYWORDS

Osteoclastoma, Giant cell tumor of soft tissue

INTRODUCTION

Giant cell tumor is one of the most common benign bone tumors, representing 4-10% of all primary bone tumors and 15-20% of all benign bone tumors [1]. They occur in young adults aged between 20–40 years with a high recurrence rate and a potential for aggressive behaviour [2]. Although benign, these tumours exhibit locally aggressive behaviour and possess the potential to metastasize.1 to 5% of cases show metastasis and there is a positive correlation between the occurrence of metastases and local aggressiveness and recurrence [3]. The most frequent site of metastasis is lungs [4]. Salm and Sissons first described 10 soft-tissue tumors that had histologic structures identical to those of giant cell tumors of bone. The authors called these tumors giant cell tumor of soft tissue (GCT-ST) [5]. In 2002, GCT-ST was addressed in the 3rd edition of the WHO Classification of Tumors of Soft Tissue and Bone and it is one of the four entities classified as fibrous histiocytic tumors in the latest edition [6]. GCT-STs represent the soft tissue analogue of giant cell tumors of the bone because of their histological and immunohistochemical similarities. Most of the reported tumors have been in the extremities with the thigh being the most commonly affected site [7]. Primary GCT-STs of other sites is extremely rare, with only four cases of primary mediastinal GCT-ST have been reported in literature [8]. Here we report an uncommon case of recurrent GCT-ST localised in the iliac region.

CASE REPORT

A 24 year old male presented to the OPD with an abdominal lump. The lump was painless from beginning and slowly progressed in size over the course of two years. There were no associated features like fever, weight loss, change in bowel habits, vomiting. His past history revealed that his left thumb developed swelling and was subsequently amputated by orthopedics team in the same institution 14 years back. The histopathological examination revealed the excised specimen to be GCT of left thumb (detailed records were lost).

On physical examination his general condition was fair with karnofsky performance status score of 70. There was a hard fixed mass palpable in right iliac fossa ~ 12x14cm approximately in size, with no overlying skin changes, no local rise in temperature or tenderness on palpation. Left thumb was absent (post amputation) and healthy stump was present. No peripheral lymph nodes were palpable.

All blood parameters were normal and he suffered from no comorbid condition like diabetes, hypertension or thyroid disorders. His CECT Abdomen revealed a large well defined solid cystic lesion in right lower quadrant of abdomen extending from level of L2 vertebral body to lower border of S3 vertebra (- APxTRxCC~ 14x18x17 cm). The lesion was described as smoothly displacing and splaying the external oblique, internal oblique and transversus abdominis muscle. Medially the lesion was closely abutting right iliac blade with cortical irregularity adjacent to soft tissue component (Figure 1)

As part of metastatic workup, patient underwent CT scan of thorax which revealed multiple (total 7 in number), well defined homogenously enhancing soft tissue density lesions involving pleura and parenchyma of both lungs (largest of size ~ 31x16 mm, APxTR in superior segment of lingular lobe) which were suspected to be metastasis. (Figure 2)

Patient underwent fine needle aspiration from swelling with but the report revealed presence of only blood products. Subsequently the mass was excised. Per-op examination revealed a 20x 17 cm mass of variable consistency in the iliac region, containing 1-1.5 litres of dirty fluid with bony tissue within the wall of capsule. His post operative period was uneventful and he recovered well.

On histopathology, the gross size of specimen was 19x11x2 cm. Outer surface had obvious hemorrhage and necrosis, admixed with smooth looking areas. Inner surface was greyish-white with congested blood vessels with central hard area. (Figure 4[A]). Microscopic examination showed a cystic tumor with scattered osteoclastic multinucleated giant cells and mononuclear stromal cells. Foci of osseous metaplasia was also seen. The tumors cells were infiltrating surrounding tissue (Figure 4 [B] and [C]). Surrounding tumor margins were negative. The final impression was Giant cell tumor.

Since the entire tumor tissue was resected out with negative margins and patient did not have any respiratory complaint, he was put on follow up with a 3 monthly evaluation with CECT thorax, with an intent of taking action only if a radiological/clinical progression in symptoms was observed. Currently, the patient is on follow up and asymptomatic.

DISCUSSION

This case is unusual in many aspects. First, the patient was diagnosed with GCT of thumb which was amputated. Then ~12yr later he developed an abdominal lump for which he underwent excision, revealing Giant cell primary of soft tissue. Along the course of the illness, he also developed pulmonary nodules which are highly suspicious of metastatic disease as per the radiological report.

Soft tissue GCT was first described by Salm and Scission and their work was followed shortly in 1972 by a report from Guccion and Enzinger in which they described a series of 32 neoplasms rich in osteoclast like giant cells, which they called malignant giant cell tumor of soft parts [9]. In 1993, Nascimento described 10 tumors that had histologic features identical to those of giant cell tumor of bone and similar to the tumor description of Salm and Sissons. He called this entity giant cell tumor of soft parts. Two of the 10 patients died of lung metastasis, and this entity was considered a low-grade sarcoma [10]. Majority of the tumors reported in the literature are located in the lower extremity, thigh being the most common site. But GCT-STs have been

seen at various other rare locations. Similar to our report GCT-STs of various sites have been reported. GCT in a mass of the liver was reported in a 88yr old female in China by Chen et al [11]. A recurrent primary mediastinal GCT-ST was reported in a case study in China [12]. Kishi et al in 2018 reported a case of retroperitoneal primary GCT in a 78yr old male. In 2000, Oliveira et al. reported 22 cases of primary GCT-STs [13], and they demonstrated that only 1 patient died of local recurrence and distant metastasis, but the other patients survived after complete resection of tumors[13]. Metastasis occurs rarely in Giant Cell Tumor of Bone (GCTB), and it most commonly develops in the lungs. The factors that contribute to increase risk of metastasis in GCTB may include the primary site of the tumor, history of multiple local recurrences, and modality of treatment of the primary tumor [14]. The primary treatment modality includes complete surgical excision with negative margins. Incomplete surgical excision is usually followed by local recurrence. Biological behaviour is unpredictable. Giant cell tumor of the soft tissue has shown a lower mean local recurrence rate compared to GCT of bone but has a higher metastatic and death rate. Therefore, close clinical follow-up is recommended [15]. In this report, we provide important information regarding diagnostic imaging (plain film, CT, MRI, Hiistopathology) from a case of recurrent metastatic GCT-ST at the very rare site i.e., abdomen. We feel that GCT-ST is a distinct entity histologically identical to its osseous counterpart as a primary neoplasm within the soft tissue. Prognosis of GCT-ST varies and the biological aggressive course for its local recurrence cannot be predicted. complete surgical excision and a long period of follow up are essential in these cases. These neoplasms should be distinguished from other giant cell rich soft tissue tumors with which these may be confused. We have kept the patient on follow up after excision of the primary tumour and he is planned for 3monthly evaluation of local site as well as lung metastasis to look for signs of local recurrence or progression of lung metastasis.

CONCLUSION

This is the first report of GCT-ST arising in the abdomen. GCT-ST is a tumor of low-grade malignant potential, identical clinically and morphologically to giant cell tumor of the bone. however, the tumor in our case was huge and invaded into surrounding areas with possible metastatis to the lung.

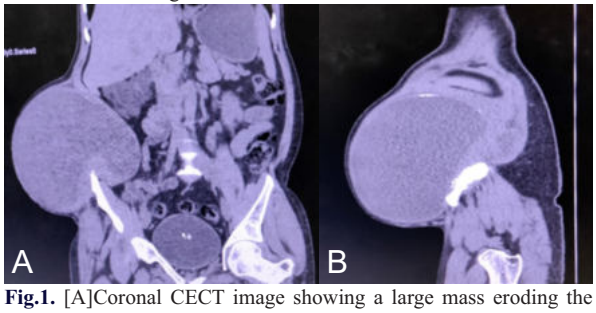

Fig.1. [A]Coronal CECT image showing a large mass eroding the Rt.Iliac Blade.[B] Saggital section showing large mass in Rt.iliac region.

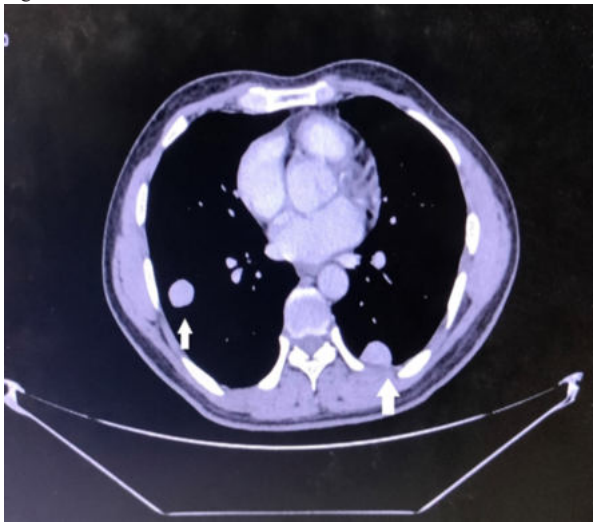

Fig 2. Axial section of CECT thorax: White arrows indicating suspicious pulmonary nodules.**A**

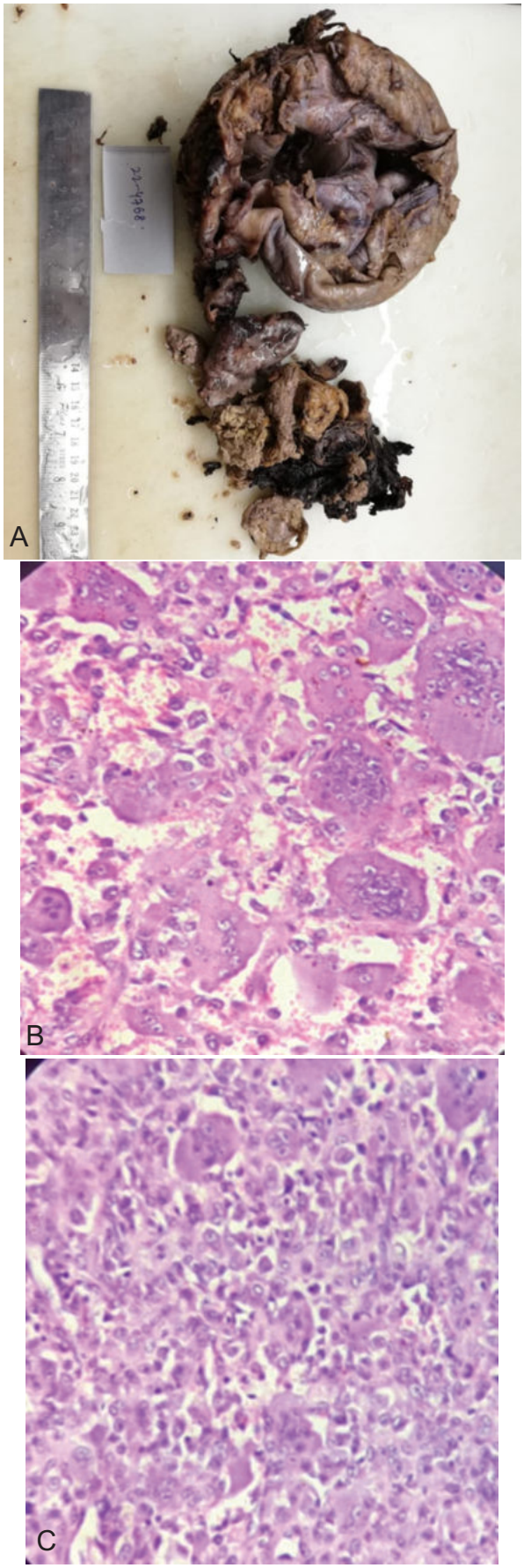

Figure 4:[A] Gross image of the excised tumor from the iliac region. [B]& [C] Histopathology slide from soft tissue show Multinucleated Giant cells within the soft tissue and cells with high mitotic activity.

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