

Conventional Giant Cell Tumor of the Rib with Pulmonary Metastases: A Case Report and Review of Literature

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ABSTRACT

We report a 25-year-old female with a giant cell tumor originating from the anterior arc of the rib who presented with bilateral pulmonary metastases. She underwent extensive resection of the thoracic wall, attached right middle lobe and right upper lobe metastases. She was treated with Interferon- α -2b (INF) followed by systemic chemotherapy. After 23 months, she had no complaint and no significant disease progression was detected through imaging studies. (*Tanaffos*2010; 9(2): 64-68)

Key words: Giant cell tumor, Rib, Pulmonary metastasis

INTRODUCTION

Giant cell tumor (GCT) is an aggressive recurrent tumor with a low metastatic potential (1-3). This tumor tends to involve the epiphyses of the long bones. It is rare for a GCT to appear in the ribs. Giant cell tumor occurs slightly more often in females than in males (4). Most patients are in the 3rd and 4th decades of life (5). Pain, mass, local tenderness and decreased motion in the adjacent joint are the most common clinical symptoms. Eighty percent of GCTs of the long bones occur after skeletal maturity, and 75% of them develop around the knee joint (6-9). The rib is a rare site for a GCT with an incidence below one percent (7). GCTs are eccentric lytic lesions with no matrix production. They have

poorly defined borders and they are juxtaepiphyseal with a metaphyseal component. Treatment of the GCT of the bone is surgical excision (1,3,4,10-13).

CASE SUMMARIES

A 25-year-old female patient presented with a one-year history of cough and pain in her right hemithorax. A few months later, she found a bulging mass in the anterior part of her right hemithorax.

On physical examination, her performance status was $\frac{1}{4}$. She had a firm mass of about 12x12x12 cm in the right upper quadrant of her right breast. In addition, breath sounds were decreased at the right side. No pallor, lymphadenopathy or organomegaly was detected. Limbs examination was normal.

Chest X-ray showed opacification in the right hemithorax and scanty pulmonary nodules (Fig. 1). Spiral computed tomography (CT) scan of the chest demonstrated rib expansion due to a large calcified mass originating from the anterior chest wall ribs

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with intrathoracic extension (Fig. 2). It had non-homogeneous enhancement and pressure effect over the right heart and mediastinal structures. Also, multiple bilateral pulmonary parenchymal nodules were seen. Spiral CT of the abdomen and pelvis was normal. A bone scintigraphy study revealed a specific calcified or osteoblastic mass in the right hemithorax without distant metastasis.

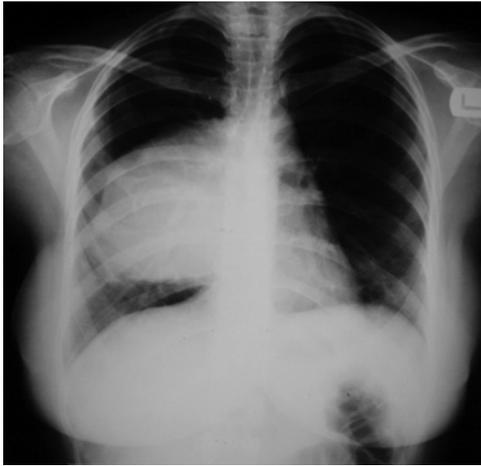


Figure 1. Chest X-ray shows a huge opacification in the right hemithorax.



Figure 2. Chest CT scan shows a calcified chest wall mass originating from the anterior arc of the rib. Pulmonary metastasis is also evident.

The blood test results including serum levels of alkaline phosphatase (196 IU/L), calcium (10.5 mg/dl) and phosphorus (3.5 mg/dl) were within the normal limits. The diagnosis was made through an incisional biopsy which revealed a giant cell rich tumor characterized by numerous giant cells with more than 10 nuclei distributed evenly in a vascular stroma containing oval cells with nuclei akin to giant cell nuclei, no cellular atypia or abnormal mitotic figures and areas of reactive bone formation, findings that were compatible with those of a conventional GCT (Fig. 3).

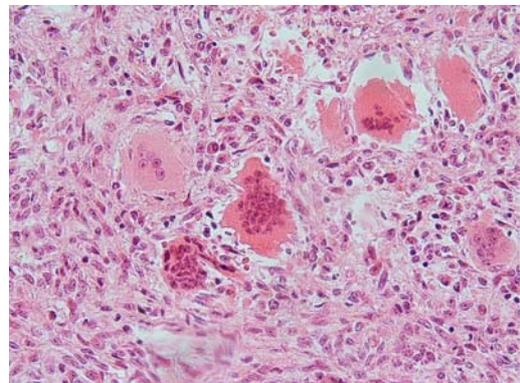


Figure 3. Multinucleated giant cells with nuclei akin to stromal cell nuclei.

The patient initially underwent surgical resection of the chest wall tumor and anterior portion of the 2nd to 4th ribs along with removal of the attached lung tissue and wedge resection of lung metastases. The tumor was 10 x 10 x 7cm in size with a tan-yellow capsule and gray-brown to hemorrhagic soft solid cystic cut surface (Fig. 4).



Figure 4. An encapsulated tumor with solid-cystic gray-brown to hemorrhagic cut surface.

Histological findings confirmed the diagnosis of a conventional benign GCT. Similar findings were obtained from the lobectomy specimen and resected lung nodules.

Treatment was started with 3 MU IFN- α -2b, thrice weekly. Since the metastatic lesions of the lungs were stable after 3 months of INF therapy, we discontinued IFN and started chemotherapy with a combination of adriamycin and ifosfamide. After 4 courses of chemotherapy, lung CT scan showed a stable disease, therefore no further medical treatment was recommended.

Since then, outpatient follow-up evaluation with chest X-ray and/or CT scan has been performed every 4 months. Eighteen months after the termination of chemotherapy, we had no local recurrence and the pulmonary metastatic lesion had minimal progression.

DISCUSSION

Occurrence of a GCT in the axial skeleton is considered a rare incidence and to date, only a few cases of GCT of the rib have been reported (14, 15). Even in cases with ribs involvement, most lesions were located in the posterior arc of the ribs. Only a small number of lesions were reported to originate from the anterior arc (16).

In this article, we reported a case of GCT originating from the anterior arc of the rib. Because of its rarity, GCT arising from the rib is difficult to diagnose, especially when it is located in the anterior arc (17).

Among laboratory findings in these patients, serum acid phosphatase value is suggested to be a useful marker for diagnosis of GCT and for evaluation of the efficacy of treatment. In one study, this value was high in 56% of patients and returned to normal after resection of the tumor (18). In our patient, we did not have an initial value for serum acid phosphatase; but it was normal two weeks after

surgery.

Initially, this tumor can be studied by plain radiographs, CT scan and MRI. The goal of these imaging studies is to determine the nature and location of the tumor and evaluating the presence of probable pulmonary metastasis, although it is not a common finding at the time of diagnosis. Bone scan is useful for detection of other sites of skeletal disease (19).

The histopathological diagnosis of chest wall tumors cannot be made based on evaluation of the samples collected through fine-needle aspiration (20). Open biopsy is considered the most appropriate method to make definite diagnosis of GCT (21, 22). In this case, we had to consider some other differential diagnoses especially when studying the incisional biopsy specimen such as "giant cell rich osteosarcoma" which was ruled out due to the absence of cellular atypia. In addition, the tumor had reactive bone formation rather than neoplastic osteoid formation which is a significant diagnostic finding in osteosarcoma. Other differential diagnoses including "giant cell rich soft tissue sarcomas" were ruled out due to the absence of malignant tumor morphology. Absence of metaplastic bone without osteoblastic rimming ruled out "fibrous dysplasia" that can be a giant cell rich lesion.

The preferred treatment for most patients with potentially completely resectable tumors is surgery (4,12,13,23-26). Wide resection may be employed for biologically aggressive extensive lesions. Patients with tumors that are not amenable to surgical resection, particularly those with locally aggressive or recurrent tumors are treated with moderate-dose radiotherapy (27-29). Chemotherapy has a limited role in the treatment of GCT (8,30,31). Since these are vascular tumors, the use of antiangiogenesis agents could be a reasonable approach. In this regard, at MD Anderson Cancer Center, some patients have been treated with IFN with either 3 million units SC

everyday for 6-12 months or 10 million units SC every Monday, Wednesday and Friday for 6-12 months (30). We treated our patient with IFN followed by chemotherapy with no remarkable response. The reason for the lack of response to IFN could be the low dose of IFN administered or its early termination (3 months duration).

Another interesting finding in this case was pulmonary metastasis that is rare in conventional benign GCT (32). The occurrence of pulmonary metastases in GCT of the bone, which was first reported by Finch et al. in 1926, is well documented (33). The frequency of metastases ranges from 1% to 9% and the majority of them affect the lungs (8,30,31,33).

Complete excision of metastases has been very successful with good long-term survival rates, but those with an inoperable disease may die from metastasis (9, 34-36). Although rare, there are several reports where the metastases have completely regressed spontaneously or have remained static for years (34-36).

Eighteen months after the last chemotherapy, the patient had no evidence of local or regional recurrence and remaining pulmonary metastases were asymptomatic and static.

Metastatic disease in giant cell tumor does not carry the same poor prognosis as it does in other tumors. Therefore, therapy should be directed at achieving adequate local control and if possible, complete excision of the metastatic lesion. Although there is lack of compelling evidence, but antiangiogenesis agents may have a role in the treatment of this tumor and further studies are awaited in this regard.

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