Abstract

Primary chest wall tumors are rare and primary osteosarcoma of the chest wall is considered as an even rare among the primary chest wall malignant tumors. The main presentation is rapidly expanding painful mass with elevation of alkaline phosphatase. We present a case of a 34 years old male who was found to have an incidental asymptomatic large chest wall mass with normal alkaline phosphatase level. He underwent several radiological diagnostic modalities which showed the extent and delineation of the mass. Complete excision of the mass was achieved and the chest wall defect was reconstructed with Prolene mesh. The histopathology confirmed the diagnosis of osteosarcoma of the chest wall.

Key words

Osteosarcoma, chest wall tumors, primary chest wall tumor, chest wall reconstruction

Introduction

Chest wall tumors challenge clinicians with respect to diagnosis and management. The spectrum of these lesions are wide, some are benign, malignant or even non neoplastic (1). The malignant lesions are classified into eight diagnostic categories: muscular, vascular, fibrous and fibrohistiocytic, peripheral nerve, osseous and cartilaginous, adipose, hematologic, and cutaneous (2). Of the osseous origin is the osteosarcoma. It is considered as a rare malignant primary chest wall tumor (3). It usually arises form the ribs, scapula and the clavicle (2).

Most of the patients with malignant chest wall tumors present with rapidly expanding mass. Pain is the most common symptom (4). The radiological modalities will aid in the diagnosis. They will show suggested features pertaining to one of the categories mentioned earlier but the need for tissue diagnosis is far important in regard to the best course to management. Plain radiography will show the lytic bony lesions as well as the extent of destruction. CT scan will give a better visualization of the bony destruction and the tiny calcification within the tumor tissue. MRI has a superior role in the delineation of the tumor with better assessment of the extent of invasion (1). The next step in the management is obtaining tissue diagnosis, and that is achieved by fine needle aspiration, true cut, incisional or excisional biopsies.

The tumor has to be resected surgically with safety margins. Complete surgery is an essential component of cure (5). The issue of chest reconstruction has improved the management and outcome of such tumors by reducing the morbidity and mortality. The reconstruction depends on main factors that would be decided up in the preoperative assessment. Reconstruction of the chest wall includes restoration and stabilization of the skeletal defect as well as soft tissue coverage. In general, this procedure can be performed in one operation, with short hospitalization (6).

Case Report

We present a case of a 34 years old male who presented to the plastic surgery team for release of skin tethering on the right cheek. He was diagnosed 4 years earlier as a case of basal cell carcinoma which was excised completely with no evidence of metastasis found...
at that time. The patient did not have any other medical history. He was a smoker for almost 15 years. The patient did not have any complaints regarding the respiratory system or any other system. His physical exam showed decreased air entry mainly on the left lower lung field mainly posteriorly with stony dullness on percussion. His routine laboratory investigations were within normal as well as his pulmonary function tests. chest x-ray showed a retro-cardiac shadow consistent with a posterior mediastinal mass. (Fig. 1) Chest CT scan confirmed the presence of the mass in the posterior mediastinum, with evidence of rib destruction and involvement of the posterior chest wall muscles. (Fig. 2) There was a line of cleavage between the mass and the aorta, heart, lung and esophagus. It was difficult to comment on the diaphragm involvement.

Fig. 1. Chest X-Ray PA and lateral views showing a large well defined posterior mediastinal mass violating the posterior part of the left 9th rib.

Fig. 2. Chest CT scan soft tissue and bone windows: A large well defined soft tissue density mass located at the left lower posterior chest wall with calcification spots and involving the posterior part of the left 9th rib.
but there was an element of indentation on the diaphragm by the mass. Chest MRI (Fig. 3) showed proper delineation of the soft tissue involvement especially the diaphragm. The results confirmed the absence of any diaphragm involvement. True cut biopsy revealed undifferentiated round cell tumor with dense chromatin. The patient underwent exploratory thoracotomy. A left posteriolateral incision was done at the level of 8th intercostal space. Three ribs were excised with their periosteum (one normal rib above and below the lesion). The mass was not attached to the lung and it was large in size measuring 15x10x10 cm. Complete excision of the mass was achieved (Fig. 4). Chest wall reconstruction was done by covering the defect with Prolene mesh and the muscles were closed primarily over the defect (Fig. 5). Histopathology showed a tumor composed of spindle cells arranged in interlacing fascicles with nuclear pleomorphism. There were areas of mitosis and necrosis. Hyaline material consistent with osteoid tissue was seen in-between and around the neoplastic cells. The immunostaining was strongly positive for Vimentin, but weakly positive for EMA, Actin, Myosin, NSE and chromogranin. The final diagnosis was malignant mesenchymal tumor consistent with osteosarcoma grade II-III (Fig. 6). The patient had uneventful post-operative course and he was referred to a specialized oncology center for chemotherapy but for social reasons he went to his native country.

Fig. 3. Chest MRI Coronal and axial T2: A large low signal intensity mass displacing the left copula of diaphragm and extending to the posterior chest wall muscle. Associated left pleural effusion is seen

Fig. 4. Operative image, showing the mass arising from the posterior chest wall with line of cleavage between the mass and the diaphragm with no evidence of lung invasion.

Fig. 5. Chest wall defect after complete excision of the mass and approximation of anterior part of the thoracotomy with defect in the postero-lateral aspect. It was covered later by Prolene mesh.
Discussion

Chest wall tumors are rare. They usually arise from various benign or malignant lesions. This fact led to some difficulties in diagnosis and therapy. Around 50% of these lesions are malignant and most of these are either metastatic or as a result of direct invasion from adjacent structures, mainly the lung, pleura and the mediastinum. The chest wall tumors are classified into 4 major groups: primary, metastatic, extension form adjacent structures or non neoplastic lesions. The primary lesions are sub-classified into either malignant or benign and these are subdivided based on the origin whether originating from bone and cartilage or soft tissue.

The approach to these tumors involve an accurate diagnosis by either fine needle, true cut, incisional or excisional biopsies. Wide surgical resection with adequate safety margin is a major part in the management. The appropriate reconstruction of the chest wall has led to the improvement in management of such cases by reducing the morbidity and mortality. Osteosarcoma of the chest wall constitute 10% of all primary chest wall tumors. It accounts for almost 3% of all sarcomas. It is considered as an aggressive malignant tumor with poor prognosis. It affects teenagers and young adults with slight increase among males. Patients usually present with rapidly enlarging painful mass. But can be completely asymptomatic as reported in this case. Usually alkaline phosphatase is elevated but could be normal as in this case.

The radiological investigations can be a useful tool in regard to site, size, extent, degree of bone destruction and degree of invasion and state of lymph node involvement. In osteosarcoma, chest radiography may show lytic or sclerotic osteoid bone matrix in the lesion. An important feature on CT scans is the spatial distribution of areas of mineralization, which is greatest at the center of the lesion and least at the periphery. MRI may depict tumor mineralization, which may have signal intensity higher than that of muscle on T1-weighted images. Mixed but predominantly high signal intensity is observed on T2-weighted images. Large cystic components also may be depicted. The tumor shows heterogeneous enhancement after administration of intravenous contrast material.

By gross examination, these tumors are large, lobulated, extending through the bone and adjacent soft tissues. Under histopathological examination, it will show either one of three elements; predominantly bone, cartilage or fibrous.

These tumors do metastasize and it is reported that 34% of patient have metastasis at time of presentation. The recommended treatment is wide surgical resection with adequate safety margins and adjuvant chemotherapy. It has been largely discussed what can be considered a safe resection margin. McCormack et al. suggested that in sarcomatous tumors, resection should include the superior and inferior sound ribs and intercostal muscles besides all the affected tissues. King et al. recommended a 4 cm free margin for highly aggressive primary.
tumors and 2 cm for metastases or benign or low-grade malignancies to avoid local recurrences. Following McCormack’s recommendations (8,9), in 2001 Warzelham et al. (11) found a 64% 3-year and 58% 5-year survival rates in chest wall sarcomas. Stabilization of chest wall with prosthesis is considered to decrease the need of prolonged mechanical ventilation and improve postoperative pulmonary function (6,7,12). The size and site of the lesion influence the selection of the prosthesis. A rigid one seems necessary for sternal and parasternal defects but soft prosthesis are enough in anterolateral defects when compared to posterior and anterior ones (13,14). Losken et al. reported that mesh closure was required more often for lateral defects when the prognosis remains poor and unfortunately the 5 years survival is around 15% with a median of 12 months (3).

References