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Case Report

Axillary Fibromyxoid Sarcoma with an Aspergillus Niger in a Tumor Cavity Compressing the Chest Wall: A Rare Tumor with Fungus

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ABSTRACT

Axillary tumors constitute about 0.11% of all malignancies. More rare tumors arise from the soft tissue component of the body that includes muscles, nerves, or fibrous tissues. Low-Grade Fibromyxoid Sarcoma (LGFMS) is a rare, malignant, soft tissue tumor often seen in young and middle-aged adults. A number of cases are found in the literature of LGFMS that can arise from different locations like thigh, breast, head, neck, masseter muscle, uterine cavity, and mesentery, but a huge axillary fibro-myxoid sarcoma with Aspergillus Niger fungal infection within the sarcoma cavity is not yet reported. We present here a young male with a two-year history of painful swelling in the right axillary region which was foul-smelling with a discharging sinus containing pus. The patient was empirically treated with anti-tuberculous drugs and was also operated twice. The patient was admitted, and a chest computed tomography scan with contrast was done which showed an asymmetrical right chest with a large heterogeneous mass in the right posterosuperior aspect of the chest wall extending into the axillary region. Operatively the draining sinus along with the mass was excised and sent for histopathology. A 17-18x8 cm hard calcified capsule was removed. On cross-section, a thick hard cavity laden with debris was noted. Immunohistopathology confirmed a low-grade fibro-myxoid sarcoma. Therefore, a discharging chest wall sinus should be reviewed. Broad differentials including tuberculosis must be ruled out especially after effective anti-tuberculous therapy. The risk of metastasis is significantly reduced with efficient excision and aggressive surgical intervention reducing further risk of future complications.

Keywords: Sarcoma, low-grade fibro-myxoid sarcoma (lgfms), aspergillus niger.

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Introduction

Axillary tumors both benign and malignant are a rare entity. These can arise primarily from the axillary structures or can also metastasize from distant sites [1]. One variant of fibrosarcoma is that of low-grade fibro-myxoid sarcoma (LGFMS), which has an inherent aggressive metastatic potential [2]. A rarer complication is that of an associated fungal infection within the sarcoma cavity as present in our case. After a thorough literature search, we could not find any case with Fibromyxoid sarcoma with Aspergillus Niger infection in the tumor cavity which is an unusual incidental finding.

Case Report

A 30-year-old male with no associated co-morbid presented with a two-year history of painful right axillary swelling with a pus draining foul-smelling sinus. The patient complained of a reduction in the range of motion with pressure-like effects. He was empirically treated for tuberculosis with anti-tuberculous drugs and was also operated in the same region twice previously. Multiple biopsies were done which remained inconclusive of the etiology. On thorough clinical examination, a hard, mildly tender, foul-smelling brownish fluid discharging sinus, and adherent to overlying skin at sinus area was seen in the Right axillary region with an intact neurovascular status of the upper limb. On further investigation, a chest x-ray showed an asymmetrical right hemithorax with soft tissue swelling in the right axillary area (Figure 1). A contrast-enhanced chest computed tomography scan showed an asymmetric, large, heterogeneous soft tissue mass of about 12.1 x 6.3 x 11.0 cm was seen in the right posterosuperior aspect of chest wall reaching into the axillary region (Figure 2).

The walls of the mass appeared thick with an enhancing internal rim and soft-tissue attenuation and multiple air loculations were seen. Multiple enlarged axillary lymph nodes were seen on the right-side largest measuring 1.3 x 1.2 cm. The mass was seen to indent pectoralis major and poster laterally inseparable from subscapularis muscle. It was causing compression of 2nd, 3rd & 4th rib anteromedially while posteriorly was resulting in rib cage deformity. The discharge from the sinus was sent for drug sensitivity and tuberculosis workup, which was negative for TB while showing growth of Gram-positive cocci. He was started on an appropriate antibiotic thereafter. The mass appeared ovoid and 17.0-18.0 cm in length and 8.0cm in width

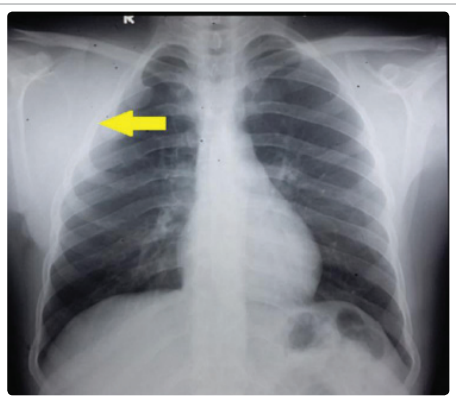

Figure 1: Chest Radiograph showing soft tissue swelling in the right axillary area.

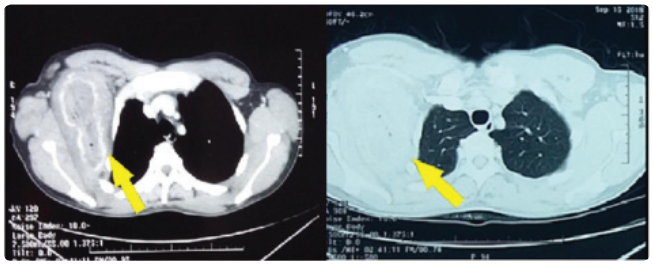

Figure 2: A Contrast-enhanced CT chest showing soft tissue density lesion in right posterosuperior aspect of the chest wall causing chest wall compression (yellow arrows). CT Computed Tomography.

with a hard-calcified capsule. The sinus was carefully excised as it was extending deep into the axilla, subclavian and axillary vessels. On cross-section the mass had a hard-outer capsule with debris (Figure 2,3). Histopathology of the mass showed a moderately differentiated neoplastic lesion composed of fusiform spindle-shaped cells with abrupt transition to myxoid areas and stroma with collagen rosettes. However, increased cellularity, atypia or mitotic activity was not seen. About 20 lymph nodes were excised and showed benign reactive changes. Immunohistochemical staining was positive for MUC-4, EMA, and ASMA. While S100, Desmin, CD34, Cytokeratin Ae1/Ae3 & STAT6 were negative suggesting a low-grade Fibro-myxoid sarcoma. Fungal culture was also sent, and the patient was started on antifungal therapy. Immune status was checked, and a thorough history was gathered. Metastatic disease was ruled out using a Positron Emission Tomography Scan. The patient was symptom-free on his one-month follow-up.

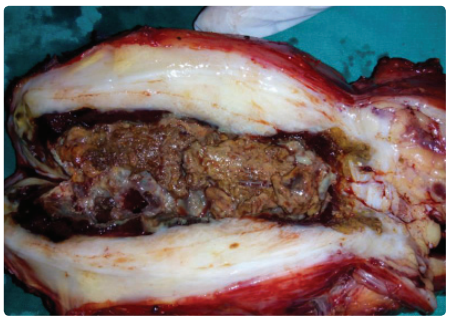

Figure 3: A Cross-section of the tumor cavity showing collected fungus and debris.

Discussion

Malignant axillary tumors account for 0.11% tumors and are a rare finding corresponding to their location [1]. According to the American Cancer Society, soft tissue tumors are a rare variety arising from soft tissues such as muscles, nerves, fat, and fibrous tissues. There are about 50 different types of soft tissue tumors, of which less than five percent occur approximately ten years after radiation exposure. Other risk factors include familial cancers such as neurofibromatosis, Li Fraumeni syndrome, Gardner syndrome, tuberous sclerosis [3]. Low-Grade Fibromyxoid Sarcoma (LGFMS) is a rare, malignant, soft tissue tumor often seen in young and middle-aged adults. Several cases of LGFMS have been reported in the literature found in different

sites in the human body like thigh, breast, head, neck, masseter muscle, uterine cavity, mesentery. Up till now, only one study has been done in Pakistan regarding abdominopelvic and retroperitoneal low-grade fibro-myxoid sarcoma and their immunostaining patterns but a huge LGFMS has not yet been noticed.

An incidence of LGFMS has been reported to be 0.18 per million, with approximately 10% of all soft tissue sarcomas, originating from the deep layers and occurring in both the genders with the same frequency, although male predominance is reported in some case reports [4]. Histologically, Low-grade Fibromyxoid sarcoma comprises spindle cells in a whirling pattern with collagenized and myxoid areas. This heterogeneous histological appearance results in diagnostic difficulties and ultimately affects treatment strategies [5]. This tumor was first identified in 1987, Size of which may vary from one to 23 cm with a median size of about 9.4 cm as reported in various studies. A huge mesenteric low-grade Fibro-myxoid sarcoma of size 30 x 50 cm was reported so far but giant Axillary fibro-myxoid sarcoma of about 11 x 17 cm in size was not reported earlier. There is a tendency of local recurrence or distant metastasis several years after complete excision which precludes the aggressive behavior of tumor [6,7]. In our literature search unusual axillary fibro-myxoid sarcoma with these dimensions and fungal infection in tumor cavity was not found.

The most appropriate treatment of LGFMS is extensive and aggressive surgical removal with long term follow up for detection of recurrence and late metastasis [8]. Recently, after a long term follow up, some studies showed 64% recurrence, metastasis in 45%, while death from disease in 42% in positive/uncertain margins. Reported recurrence was around 15 years later; median 3.5 years); Site of metastases was usually to lungs, pleura, chest wall (up to 45 years later; median 5 years) [9].

Fungal infection is a common entity in immunocompromised states such as in cancer patients especially in hematologic malignancies, post-radiation, after Chemotherapy, HIV, and Chronic granulomatous disease. Most common fungus found in cancer patients are Candida species, but less commonly invasive aspergillus infection is associated with malignancies [10]. Aspergillus species are ubiquitous molds with about 100 different species found in organic matter. Most common Aspergillus species are Aspergillus fumigatus and Aspergillus Niger and, less frequently, by Aspergillus flavus and Aspergillus clavated [11]. Only few case reports were found in literature about sarcoma with invasive aspergillus infection i.e., Myeloid sarcoma (MS) which is a tumor mass of myeloblasts & rare extramedullary myeloid tumors that may occur de novo, or coexist with or follow the presentation of Acute Myeloid Leukemia (AML) via hematogenous spread but fungus in sarcoma cavity is not reported [12]. Owing to the nature of this tumor, there are very less chances of an overt metastasis. Hence, local resection despite the size remains an adequate treatment option without any adjuvant chemo radiotherapy [12].

Conclusion

Discharging sinus along the chest wall should always be evaluated and considered for other diagnoses as in our case there were long persistent symptoms with increased severity at presentation and no improvement despite antituberculous therapy and some previous surgical interventions. After prompt workup excisional biopsy of axillary mass and subsequently confirmed histopathologic diagnosis is needed for further radiotherapy or chemotherapy, hence, rare tumors can be found at rare sites.

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