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Stewart-Treves Syndrome in a Patient with Breast Cancer on FDG-PET/CT Imaging

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Abstract: *Lymphangiosarcoma is a rare malignant tumor which occurs as a complication of chronic lymphedema. 18-F-Flourodeoxyglucose (FDG) Positron Emission Tomography/Computed Tomography (PET/CT) is usually used in a variety of cancer patients for staging, re-staging and treatment response. We present a rare case of lymphangiosarcoma with a history of right mastectomy, axillary lymphadenectomy and chemoradiotherapy 14 years ago in a 73-year-old woman who presented with a mass on her right lymphedematous arm. FDG-PET/CT demonstrated the second primary tumor and its subcutaneous spread.*

Keywords: *Stewart-Treves syndrome, Angiosarcoma, Lymphedema, Breast Cancer*

1. Introduction

Angiosarcoma is a rare entity in breast cancer patients. 18-F-Flourodeoxyglucose (FDG) Positron Emission Tomography/ Computed Tomography (PET/CT) is usually used in breast cancer patients for staging, re-staging and treatment response. Lymphangiosarcoma also called as Stewart-Treves Syndrome can be develop in the following years as a complication after therapy of breast cancer. Imaging and staging are of special importance at this stage. FDG PET-CT imaging can be useful in these patients. In this case report, the role of FDG PET/CT in the demonstration of angiosarcoma and its distribution in a patient with metastatic breast cancer is presented.

2. Case Report

A 73-year-old woman who presented with a mass on her right lymphedematous arm was sent to our department for FDG-PET/CT imaging for restaging. She had a history of mastectomy and axillary lymphadenectomy and also chemo radiotherapy for right breast cancer 14 years ago. Her arm had become persistently swollen in the recent weeks and a clinical diagnosis of lymphedema was made, initially, until she developed an ulcerated mass lesion. Biopsy of the lesion was angiosarcoma.

Immunohistochemical studies revealed positivity for CD34, CD31, and a high Ki-67 proliferation rate (70%). FDG-PET/CT scan was performed for restaging. Following 12 h of fasting, while blood glucose level was 102 mg/dl, 8.77 mCi 18 F-FDG i.v. was injected. After 60 minutes, the patient was imaged in the 3D mode 2.3 minutes per bed from the calvarium to the footwell. Obtained images were evaluated after attenuation correction with low dose nondiagnostic CT. Right breast was not seen secondary to operation. No pathologic focus was found in the left breast, left and right axilla and right breast operation area on PET-CT scan. Lymphedema was seen in the right arm. PET-CT scan demonstrated an approximately axial 57x48 mm heteroge- nous lobulated mass extending to skin and subcutaneous tissue and with borders seper- ated from bone and muscle structures in the right arm with a SUVmax of 17.05 (Figure 1). In the vicinity of the mass, satellite nodules with the largest size of about 2.5 cm were observed. Fibrotic changes were observed in right apical lung region secondary to radiotherapy. No additional pathologic focus was detected on whole body imaging. Accor-

ding to PET-CT findings amputation of right arm was performed.

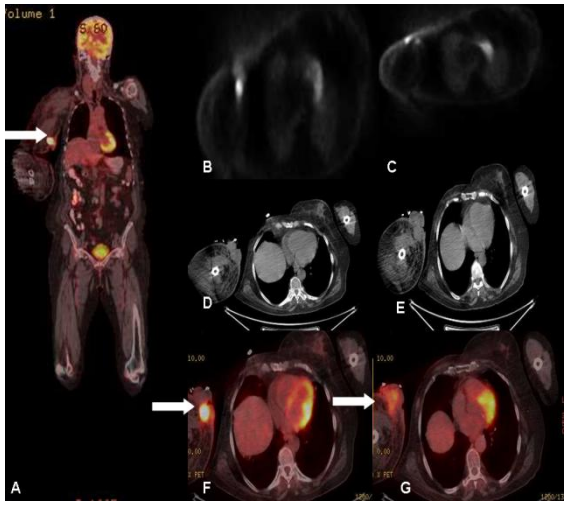

Figure1. *Coronal fusion image (A) of a 73-year-old woman who presented with a mass on her right lymphedematous arm, affected by chronic lymphedema of the upper extremity. PET-CT revealed a mass lesion with an lobulated contour showing intense FDG uptake (SUVmax: 17.05) with periferal satellite nodules in the right arm. Axial PET (B, C), CT (D, E), and fusion images (F, G) showed the secondary primary mass. One of the satellite nodules was observed at the level of the right anterior humerus head. .Additional wide resection could not be performed for local control and right arm amputation was performed instead of wide resection to the patient according to PET-CT findings.*

3. DISCUSSION

Lymphangiosarcoma is a very rare vascular neoplasm with poor prognosis occur as a complication of chronic congenital or acquired lymphedema. Lymphangiosarcoma arising in lymphedematous tissue have been reported in the literature in a variety of reports. As seen in this case, it is most frequently associated with postmastectomy lymphedema also called as Stewart-Treves syndrome (1). Stewart-Tre ves syndrome is an angiosarcoma that occurs because of chronic lymphedema. In most cases, lymphangiosarcoma is a compli- cation after mastectomy with axillary node dissection and postoperative radiation. Patients usually have a significant history for breast cancer and radical mastectomy, 5–15 years before sarcoma presentation. The mean onset between radical mastectomy and lymphangiosarcoma is 11 years (2). Stewart-Treves syndrome accounts for approximately 5% of angiosar- comas (3). Lymphangiosar- coma can excepti- onally arise in congenital hereditary lymphe- dema also called as Milroy syndrome and Meige syndrome and nonhere- ditary lymphe- dema such as congenital, praecox, or forme tarde lymphedemas (4-6). In the majority of cases in literature, purplish papules or nodules, necrosis or ulceration have occurred in upper extremities, 10 to 20 years after radical mastectomy, and radiotherapy (7-11). L Cui et. al reported 10 cases in a case report and review of literature (12). In a recent case report and review of the literature (13) the authors reported that there are approximately 400 cases of Stewart-Treves syndrome reported in the literature, most in postmastectomy patients who are predominantly women. PET-CT imaging is a valuable method in restaging of breast cancer, especially in distant metastases. Although, reports describing FDG PET-CT findings of lymphangiosarcoma are very rare (14-18). As a complication after therapy of breast cancer, lymphangiosarcoma can be develop in following years and imaging is also of special importance at this stage. The most common site of metastasis are the lungs, followed by liver, bone, soft-tissue structures and lymph nodes in lymphangiosarcoma if untreated (2, 19-21). For the demonstration of subcutaneous spread and planning of a surgical procedure, F-fluorodeoxyglucose positron emission tomography may be used (2, 22). In this case report, the role of FDG PET-CT in the demonstration of angiosarcoma and its subcutaneous spread in a patient with known metastatic breast cancer is presented. FDG PET-CT demonstrated the second primary tumor and its distribution. There was no distant metastasis in this case. High SUVmax values were obtained in proportion to aggressivity of the second malignancy. One of the satellite nodules was observed at the level of the right anterior humerus head in PET-CT images. Additional wide resection could not be performed for local control and right arm amputation was performed instead of wide resection to the patient according to PET-CT findings. PET-CT was found useful in this case especially for subcutaneous spread of the disease and excluding distant metastasis.

4. CONCLUSION

Stewart-Treves syndrome is a rare complic- ation of postmastectomy lymphedema with very poor prognosis. Patient's survival can be improved by early diagnosis and knowing the disease distribution in the diagnosis. FDG PET-CT imaging as a whole body imaging procedure can be used for demonstrating both subcutaneous spread and distant metastasis.

There are only few case reports on this entity. More reports on PET-CT imaging in lymphangiosarcoma patients are needed.

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