Case Report

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Extra Gastrointestinal Stromal Tumour (EGIST) of Chest Wall Detected on ^{18}F-FDG PET/CT: A Case Report and Brief Review of Literature

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Running Title ^{18}F- FDG PET/CT in Extra gastro Intestinal Tumor of Chest Wall

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ABSTRACT

Gastrointestinal stromal tumors (GIST) are rare tumors accounting for less than 1% of primary neoplasia of the digestive tract. They can also occur outside the GIT, where they are called extra gastrointestinal stromal tumors (EGIST), which are extremely rare tumors. GISTS and EGISTS have similar histo-pathological and molecular profiles. Though these are known to be FDG avid, some of the atypical GISTS can be FDG negative or only minimally FDG avid.

Here, an unusual case of a 57-year-old female with FDG avid solitary chest wall mass is presented, which after biopsy and immune-histochemistry was proven as EGIST. She underwent en block resection of the tumor and is presently on adjuvant treatment with Imatinib mesylate. This case report highlights the importance of considering EGIST as a differential diagnosis of a solitary chest wall mass and the utility of 18F-FDG PET/CT in its management.

Introduction

Gastrointestinal stromal tumors (GIST) are rare neoplastic lesions of mesenchymal origin, accounting for less than 1% of the primary neoplasia of the digestive tract [1]. Extra-gastrointestinal stromal tumors (EGIST) are the GIST that develop in extra-gastrointestinal locations, without primary GIT involvement, and account for approximately 10% of the total GIST cases [1-3]. thereby implying that primary EGIST are very rare entities.

EGISTS are known to occur mostly in the omentum and mesentery but there have been reported cases of EGISTS occurring at other regions such as the pancreas, spleen, prostate and even in the pleura [4-6]. The diagnosis of EGIST can be difficult, especially when in unusual locations, because of their rare prevalence. When the findings of conventional imaging like CT/MRI are ambiguous, 18F-FDG PET/CT can be helpful for characterization of such lesions [7]. Though GISTs are usually considered to be FDG avid but occasionally they may be non-FDG avid and may not demonstrate FDG uptake [7,8]. The sensitivity and positive predictive value for the detection of GISTs

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by FDG PET/CT have been described as 86 and 98%, respectively, and false-negative PET/CTs are mostly related to small lesions or non-FDG avid lesions [9]. Hence, final diagnosis can only be confirmed after biopsy and immuno-histochemical analysis.

In addition to its useful role in diagnosis in and initial staging of GIST, FDG PET/CT has been used for pre-treatment/baseline malignancy risk assessment of these tumors, into low and high-risk categories for guiding treatment decisions. Furthermore, FDG PET/CT also allows early assessment of treatment response in cases of baseline FDG positive tumors [11,12].

Case Presentation

A 57-year-old female presented with a complaint of progressively increasing pain in the upper chest in the midline for the last 2 months. This pain was moderate in intensity and continuous in nature. She also complained of difficulty in breathing and swallowing, and significant weight loss of approximately 8 kg over the last 3 to 4 months.

She was a non-smoker with no other co-morbidities, no history of previous surgery or relevant family history.

On local examination of the chest, there was a mild fullness in the midline along the sternal region with some tenderness. However, no obvious mass/swelling, ulceration, or discharge from overlying skin was seen.

In view of her complaints of dysphagia and significant weight loss, an upper GI endoscopy was ordered, which turned out to be a normal study. Subsequently, an ^{18}F-FDG PET/CT was ordered to look for any possible underlying primary tumor or to look for any other pathology causing her symptoms.

^{18}F- FDG PET/CT findings

The ^{18}F-FDG PET/CT images revealed a heterogeneous soft tissue mass in the presternal chest wall. This mass measures approximately 5.6cm (T) X 2.3cm (AP) X 5.7cm (CC), and contains a few tiny specks of

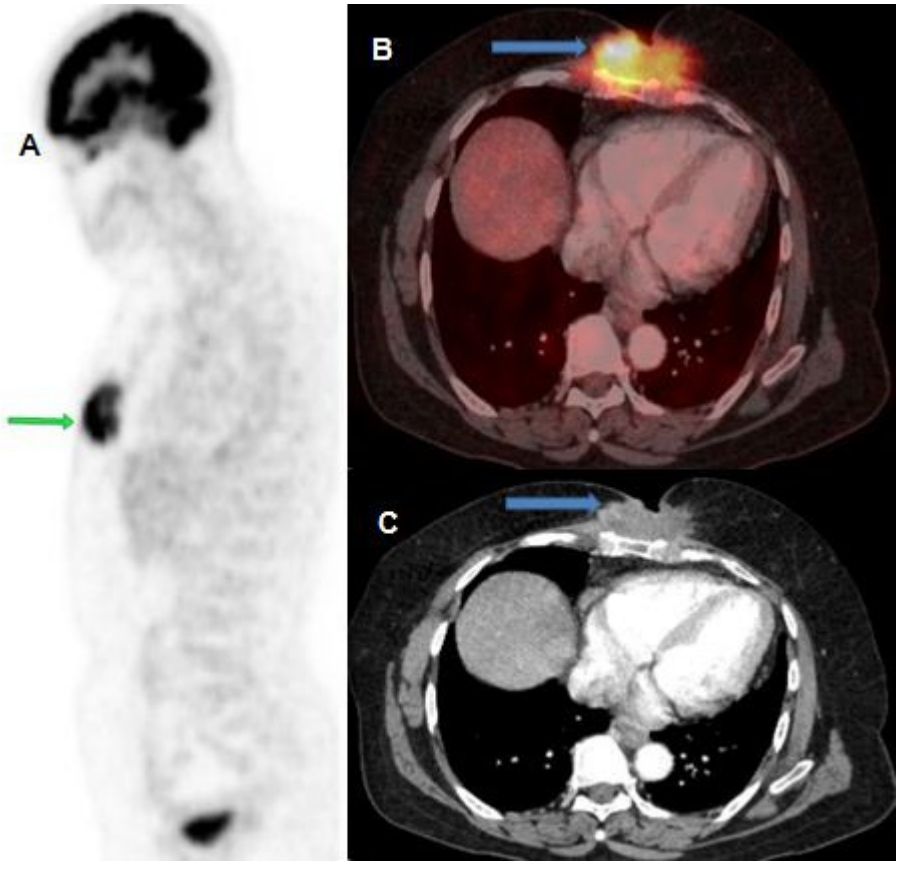

Fig. 1. A) **The PET images** show mildly increased FDG uptake in the presternal region**. Fused axial PET/CT** images reveal an FDG-avid soft tissue lesion in the presternal region, abutting the sternum at places. **Axial CECT images** demonstrate a heterogeneous soft tissue mass with a few tiny calcifications in the presternal chest wall. The mass is encasing the medial end of the left clavicle, with evidence of its erosion.

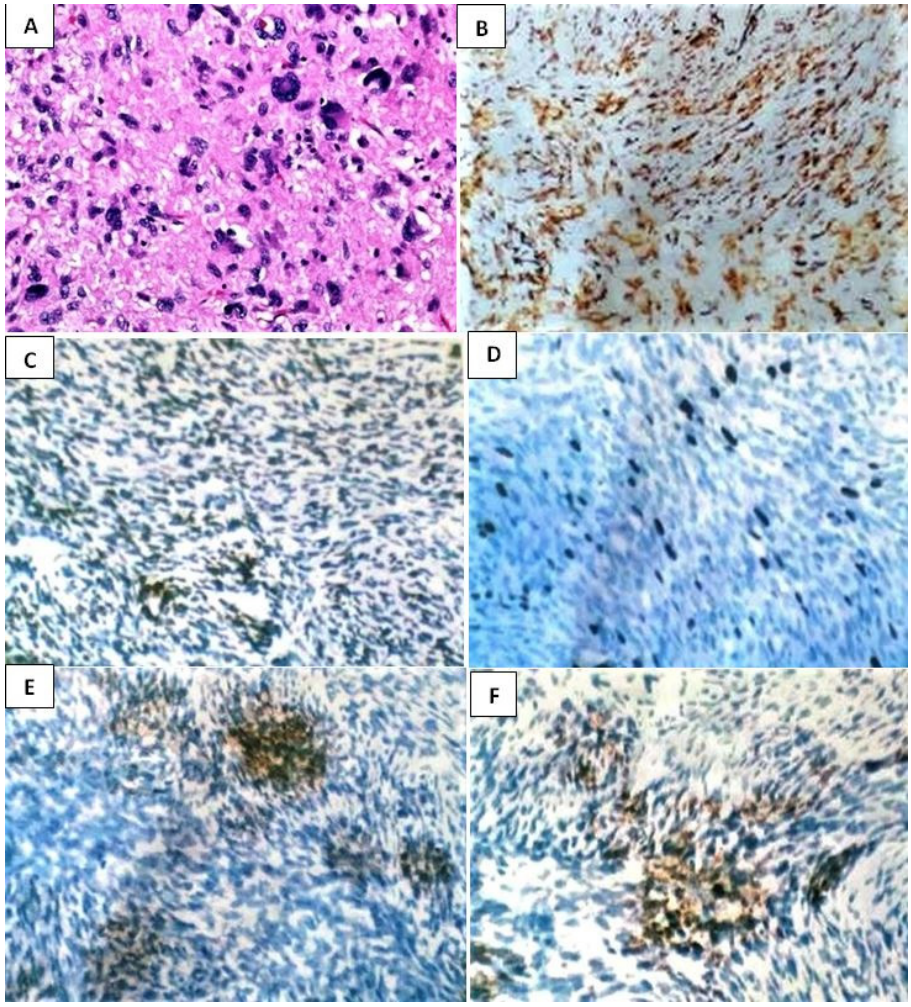

Fig. 2. (A) The histopathological examination of the resected tumour revealed spindle cells with elongated oval to spindle nuclei arranged in intersecting fascicles (H &E; 100X). Immuno-histochemical staining (B to F) demonstrated positivity for CD34 (B), SMA ©, Ki-67 (D), CD-117(E), and DOG-1 (F) respectively (200X).

calcification. It is abutting the sternum, but without any evidence of erosion. The lesion demonstrated moderate FDG avidity, with an SUVmax of 7.9 (compared to a liver blood pool SUVmax of 3.1 and mediastinal blood pool SUVmax of 1.8). No abnormal lesions or FDG uptake were observed elsewhere in the rest of the body.

A guided FNAC of the lesion was ordered, and the findings were suggestive of a spindle cell neoplasm. In light of the FNAC and FDG PET/CT findings, the patient underwent radical excision of the tumor.

The histo-pathological examination revealed a spindle cell lesion arranged in intersecting fascicles. The spindle cells had elongated oval to spindle nuclei with fine chromatin and moderate eosinophilic cytoplasm.

Immuno-histochemistry was positive for SMA (focal), CD34 (diffuse), DOG-1 (focal), CD117 (focal) and negative for CK, S-100, Desmin, and STAT-6. The Ki-67 labelling index was 12-14%. The overall features were suggestive of an extra-intestinal stromal tumor of the chest wall.

Adjuvant therapy with Imatinib (400mg/day) was subsequently initiated for her, and she is currently under close follow-up.

Discussion

GISTs, originating from GI mesenchymal tissue, constitute a small percentage of soft tissue tumours in the abdomen. On the other hand, EGISTs are considered mesenchymal tumours that originate outside the GI tract, accounting for less than 5% of GIST.[3] EGISTs have been reported to occur in various locations, including the omentum, mesentery, pancreas, spleen, prostate, and even the pleura. [3-6] The diagnosis of a primary EGIST in the chest wall

is an extremely rare occurrence. There is only one previous case report that describes a primary EGIST in the chest wall [13].

Conventional imaging methods such as CT or MRI often yield ambiguous results when the EGIST is located in an unusual region, such as the chest wall, as in the case of this patient. Further investigation is necessary to characterize this lesion and to rule out metastasis from a primary tumor elsewhere as a cause for such atypical lesions. FDG PET/CT is particularly indicated in cases with ambiguous CT or MRI results and has also been used for staging purposes [8]. However, there have been reported cases where GISTS with typical morphological criteria on CT were found to be FDG negative or showed low-grade FDG uptake [14].

The final diagnosis of EGIST requires and relies on biopsy and immuno-histochemical studies. These tumors have immuno-histochemical and molecular profiles similar to GIST [15]. CD117 (c-KIT receptor) and CD34 are both important EGIST biomarkers [2,15]. In 95% of the tumors, there is a somatic mutation of CD117 (c-kit), and its discovery in the immuno-histochemical staining characteristically defines the GIST/EGIST [2].

Beyond its role in diagnosis and staging, 18F-FDG PET/CT has also been used for risk assessment of malignant potential in GIST. The use of a cut-off value of SUVmax 3.0 has been shown to yield a sensitivity and specificity of 85.7% and 62.5%, respectively, for classifying patients into low-risk and high-risk malignancy groups. When SUVmax is more than 3.0, the tumor must be resected even if it measures less than 2 cm, because of its high malignant potential [7,10].

FDG PET/CT, using the Choi criteria, has been described as a more sensitive and specific method to assess early response to the Tyrosine kinase inhibitor, Imatinib mesylate [11,12]. Tumor size alone on CT/MRI imaging is unreliable for assessing the early response to imatinib mesylate treatment [11]. Consequently, when neo-adjuvant imatinib therapy is considered, a baseline PET/CT should be performed [16]. However, FDG PET/CT cannot be used for therapy monitoring in baseline FDG-PET negative tumors [11].

In this case, the results of the 18F-FDG PET/CT scan suggested that this anterior chest wall mass was probably malignant and at the same time it was not a metastatic lesion from an underlying primary cancer elsewhere in the body. The possibility of a chest wall sarcoma or plasmacytoma was initially considered. However, the diagnosis of chest wall EGIST was subsequently confirmed after histo-pathological and IHC work up.

The definitive management of GIST/EGIST includes en block resection of the tumor to achieve negative margins followed by adjuvant treatment with tyrosine kinase inhibitors to reduce the likelihood of recurrence, as was done in this case.

Conclusion

EGISTs are rare tumors with a wide range of clinical presentations. A chest wall EGIST is an extremely rare tumor that is difficult to diagnose and characterize on conventional imaging alone. In cases with ambiguous prior imaging, ^{18}F-FDG PET/CT can help in characterizing the lesion, not only ruling out any other possible underlying primary malignancy that could have led to a similar appearing metastatic lesion, but also allowing staging of such a lesion. However, the exact diagnosis of EGIST is based only on biopsy and immuno-histochemical analysis. Furthermore, ^{18}F-FDG PET/CT also has value in malignancy risk categorization, thus aiding in management decisions for such EGISTs. A baseline ^{18}F-FDG PET/CT scan also provides a more effective reference compared to CT alone for early assessment of treatment response. Therefore, in patients with both suspected or diagnosed EGIST, ^{18}F-FDG PET/CT scan is a valuable tool that can provide important information and should be considered in their clinical management. However, in EGISTs with a negative baseline scan, further ^{18}F-FDG PET/CT scans are not indicated for treatment response assessment.

Ethical Considerations

Compliance with ethical guidelines

There were no ethical considerations to be considered in this article.

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Conflict of Interests

The authors state that they have no conflicts of interest.

Abbreviations

CECT: contrast enhanced computed tomography

MRI: Magnetic resonance imaging

^{18}F-FDG: ^{18}Flourine - Flouro Deoxy glucose

PET: Positron emission Tomography

GIST: Gastro intestinal stromal tumour

EGIST: Extra Gastro intestinal stromal tumour

FNAC: Fine needle aspiration cytology

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