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Case Report
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Evaluation of Treatment Response in Retroperitoneal Alveolar Soft Part Sarcoma Using ^{18}F-FDG PET/CT

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Abstract

Alveolar Soft Part Sarcoma (ASPS) is a rare sarcoma predominantly presenting as a painless mass in the lower limbs. This report presents the ^{18}F-FDG PET/CT findings of ASPS in a 17-year-old woman with a painless abdominal mass. Initial ^{18}F-FDG PET/CT showed a hypermetabolic mass in the retroperitoneum and a few hypermetabolic lung nodules, suspicious of metastases. After treatment with sunitinib and surgery, a subsequent ^{18}F-FDG PET/CT revealed complete resolution of the tumors. Following the detection of recurrence, the patient underwent treatment with nivolumab/ipilimumab, leading to complete resolution as demonstrated ^{18}F-FDG PET/CT imaging.

Keywords: Alveolar soft part sarcoma; Retroperitoneum; PET/CT; ^{18}F-FDG; Chemotherapy

Introduction

Alveolar Soft Part Sarcoma (ASPS) is a rare, slow-growing tumor that predominantly affects young adults and has a unique clinical and pathological profile. Despite its rarity, its distinct clinical behavior, characterized by a high propensity for metastasis, particularly to the lungs and brain, necessitates a meticulous approach to diagnosis and management. Recent advancements in imaging techniques, especially ^{18}F-FDG PET/CT, have proven instrumental in not only diagnosing ASPS but also in monitoring treatment response and disease progression. This section will delve into the origins, epidemiology, and current understanding of ASPS, underscoring the role of ^{18}F-FDG PET/CT in optimizing patient outcomes.

Case Presentation

A 17-year-old woman presented with a painless abdominal mass in the epigastric area for three months. The ^{18}F-FDG PET/CT for initial work-up reveal a hypermetabolic mass measuring 13 cm in the right retroperitoneum. Additionally, a few hypermetabolic nodules were observed in the right lower lobe of the lung, raising suspicions of metastases. Subsequent pathological analysis confirmed the mass in the right retroperitoneum to be Alveolar Soft-Part Sarcoma (ASPS). After one month of treatment with sunitinib, the follow-up ^{18}F-FDG PET/CT revealed an increase in both the size and metabolism of the retroperitoneal tumor when compared to the initial PET/CT scan. Similarly, the metastatic lung nodules also exhibited an increase in size and metabolism. Consequently, the patient underwent tumor resection in the right retroperitoneal area, along with right nephrectomy and wedge resection of the metastatic lung nodules. Postoperative ^{18}F-FDG PET/CT images confirmed the complete resolution of the retroperitoneal tumor and metastatic lung nodules (Figure 1A-1F).

The gross pathology and microscopic examination with hematoxylin-eosin staining of the resected retroperitoneal tumor reveal large, round to polygonal cells with well-defined cell border. Immunohistochemical staining of the tumor cells revealed positive expression for Transcription Factor 3 (TFE3), providing a definitive diagnosis of Alveolar Soft-Part Sarcoma (ASPS) [1,2]. Following the surgery, the patient underwent postoperative treatment with pembrolizumab/axitinib. However, the subsequent ^{18}F-FDG PET/CT revealed a hypermetabolic lesion in the right nephrectomy bed and right paravertebral space, raising concerns about cancer recurrence. As a result, pembrolizumab/axitinib was discontinued, and the treatment was switched to nivolumab/ipilimumab. After five therapy cycles, the follow-up ^{18}F-FDG PET/CT scan to evaluate the treatment response evidenced complete resolution compared with the previous scan. Remarkably, after three years of treatment, the patient remains in good health, with no evidence of disease for the past 21 months (Figure 2A-2F).

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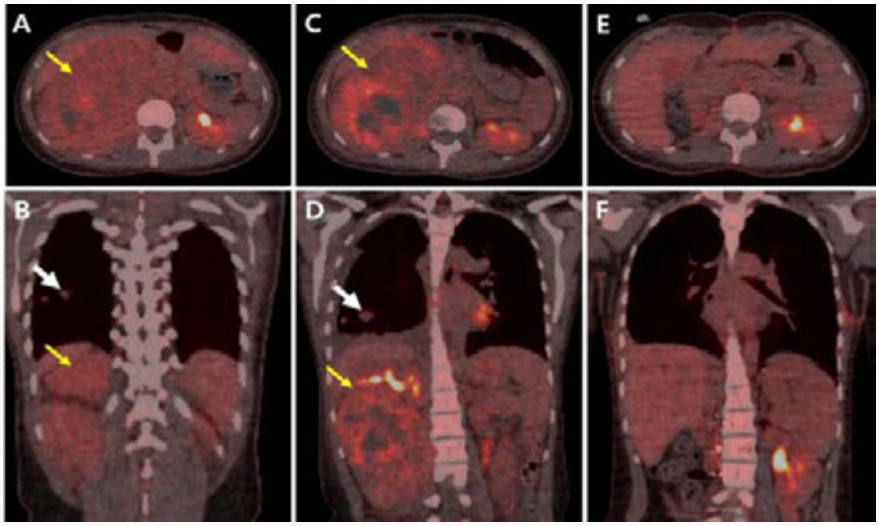

Figure 1: ^{18}F-FDG PET/CT in a 17-year-old male with ASPS: The axial (A) and coronal (B) images for initial work-up showed a 13 cm hypermetabolic mass (yellow arrow; SUVmax: 4.6) with metastatic lung nodules (white arrow; SUVmax: 1.8). On follow-up ^{18}F-FDG PET/CT (C, D) after sunitinib, the tumor (yellow arrow; SUVmax: 5.7) and metastatic lung nodule (white arrow; SUVmax: 2.3). Postoperative axial (E) and coronal (F) ^{18}F-FDG PET/CT images of the retroperitoneal tumor and metastatic lung nodules.

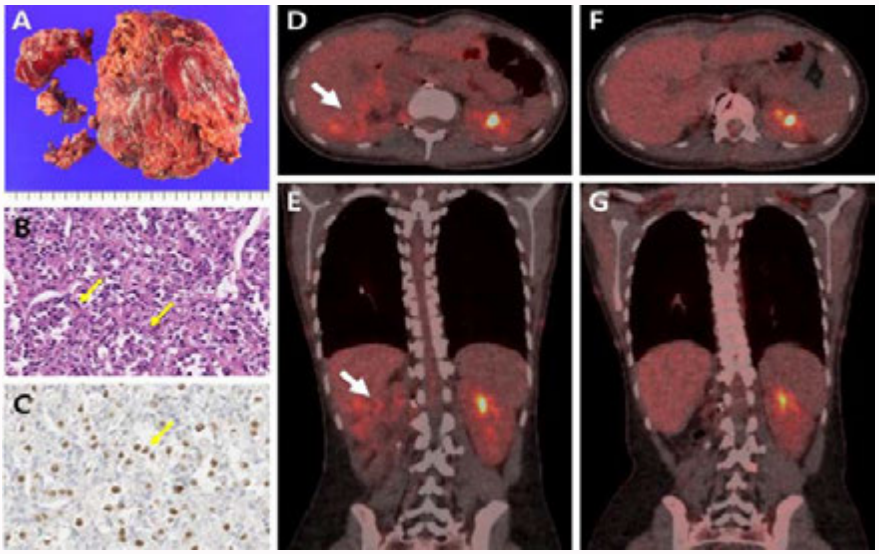

Figure 2: The gross pathology (A), microscopic examination (B) with hematoxylin-eosin staining (original magnification ×200) and immunohistochemical staining (C) of the resected retroperitoneal tumor. After surgery, the patient was treated with pembrolizumab/axitinib, ^{18}F-FDG PET/CT (D, E) revealed cancer recurrence (white arrows). ^{18}F-FDG PET/CT scan (F, G) in complete resolution after treatment with nivolumab/ipilimumab.

Discussion

ASPS, a rare malignant soft tissue tumor predominantly affecting young adults (15–35 years), usually presents as a painless mass in the lower extremities; however, it occasionally occurs in the trunk, pelvis, or retroperitoneum. Metastasis is frequently observed at the time of diagnosis, involving the lungs, brain, and bones [3-5]. Although MRI and CT findings of ASPS arising from the retroperitoneum have been documented [6,7], ^{18}F-FDG PET/CT findings are rarely reported. The application of ^{18}F-FDG PET/CT in this case exemplifies its value not only in diagnosing ASPS but also in assessing the metastatic burden, monitoring treatment response, and detecting recurrence. While ASPS is known to have variable FDG uptake [8-10], the initial and follow-up scans provided critical insights into the tumor's metabolic activity and response to therapy. This case reinforces the utility of ^{18}F-FDG PET/CT in guiding clinical decisions, from the initial work-up through to post-treatment surveillance.

Conclusion

In conclusion, this case report underscores the complexities and challenges inherent in diagnosing and treating Alveolar Soft Part Sarcoma. It reaffirms the indispensability of comprehensive diagnostic tools and tailored treatment regimens in managing this rare malignancy. The significant role of ^{18}F-FDG PET/CT in guiding treatment decisions and monitoring disease progression is highlighted, offering valuable insights into the potential pathways for improving patient outcomes.

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