Open Journal of Clinical & Medical Images

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Short Report

Open Access, Volume 2

Metastatic NUT midline carcinoma

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Received: Sep 12, 2022

Accepted: Oct 26, 2022

Published: Nov 04, 2022

Archived: www.jclinmedimages.org

Abstract

NUT Midline Carcinoma (NMC) is a rare carcinoma that presents with rearrangement to the NUTM1 (NUT) gene. NMC predominately affects adolescents and young adults with a median age of approximately 24 [1,2]. Mostly reported in pathology and oncology literature, this highly lethal cancer with a median survival time of diagnosed patients is nine months and presumably under recognized for varying undifferentiated neoplasms [3,4]. Typically, NMC presents in the midline upper airway regions of the head, neck, and thorax, with few reported cases involving solid organs and intramuscular locations [3]. To date, there is no gold standard imaging or treatment for NUT carcinoma. Within imaging literature, there are very few published cases of NMC. Here, we report the PET/CT findings of FDG-avid metastatic NMC in two atypically older patients.

Keywords: NUT midline carcinoma; NMC; NUTM1; PET/CT.

Case presentations

Patient 1: Patient 1 was a 65-year-old man with a medical history of Adult Polycystic Kidney Disease (APCKD), presenting with hemoptysis and persistent cough, for which imaging revealed an incidental 11 cm lung mass (Figures 1 and 2). Within several weeks, biopsy confirmed a diagnosis of NMC with subsequent initiation of chemotherapy. Upon initiation of treatment, the course was met with progression on varying lines of chemotherapy, eventually turning lethal. Figure 1 shows the FDG PET images of these PET/CT scans performed approximately one and eight months after diagnosis (Figure 1A-C and Figure 1D-F, respectively). The attenuation corrected coronal (Figure 1A), sagittal (Figure 1B), and maximum intensity projection (Figure 1C) PET image of the baseline scan before first-line chemotherapy demonstrates intense FDG uptake in the primary left lung lesion (Figure 1 solid arrow) and left adrenal (Figure 1 open arrowhead). Focal FDG activity in the acromioclavicular joint (open arrow) was felt to be inflammatory secondary to degenerative changes but, on subsequent imaging eight months later, thought to be metastatic given the intense increase in uptake. Subsequent attenuation corrected coronal (Figure 1D), sagittal (Figure 1E), and maximum intensity projection (Figure 1F) PET images following multiple lines of therapy eight months later showed evidence of progression with a substantial burden of FDG-avid disease on PET, including throughout the skeleton. It is important to note that osseous metastasis was not evident on cross-sectional imaging without the help of molecular imaging with PET. Further evaluation of the primary mass at baseline (Figure 2A-C) and eight months later (Figure 2D-F) demonstrated development of FDG avid ground-glass opacities (Figure 2 open arrowhead) which was felt to be lymphangitiscarcinomatosis.

Patient 2: Patient 2 was a 64-year-old female with no significant medical history who presented with complaints of acute sensation of substernal tightness following two months of cough and congestion (Figure 3). Imaging in this patient revealed a circumferential soft tissue mass involving the esophagus and trachea, causing narrowing of the trachea with

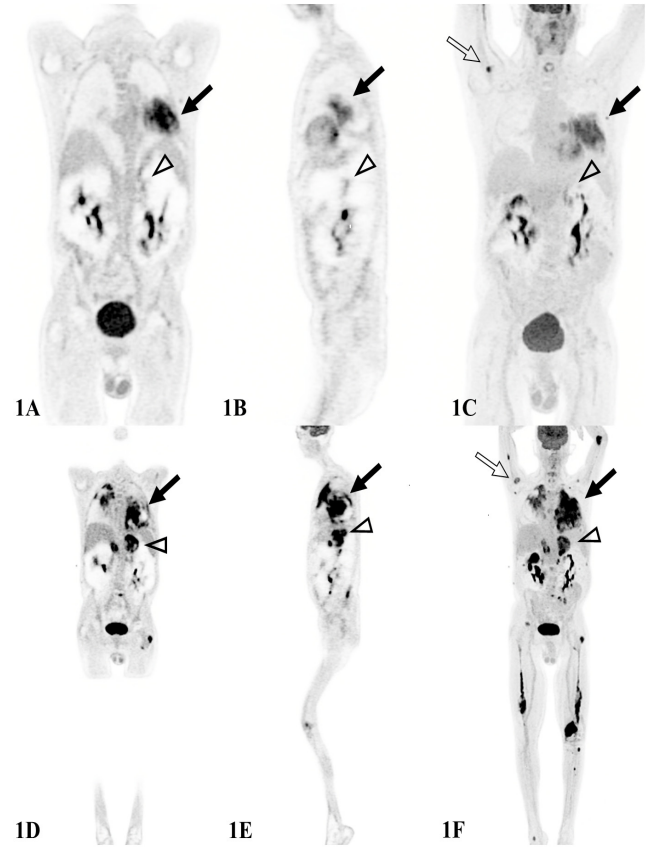

Figure 1: Patient 1 Whole body FDG PET images. Top row (1A-C) demonstrates baseline multiplanar PET images obtained 1 month after diagnosis with FDG-avid 11 cm left lung mass (solid arrow), left adrenal gland (open arrowhead), and right acromioclavicular joint (open arrow). Bottom row (1D-F) demonstrates 8 month follow-up multiplanar PET images with significant progression of intensely FDG-avid disease in the bilateral lungs (solid arrow), left adrenal gland (open arrowhead), right acromioclavicular joint (open arrow), and additional new sites.

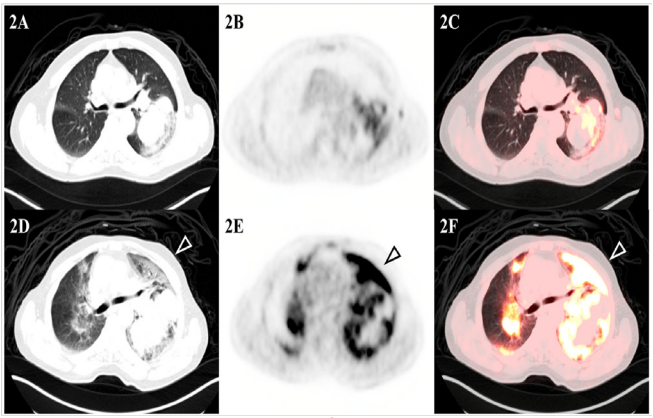

Figure 2: Patient 1 Axial FDG PET/CT images. Top row (2A-C) demonstrates baseline axial CT (2A), axial PET (2B), and axial fused PET/CT (2C) images of the chest obtained 1 month after diagnosis with FDG-avid 11 cm left lung mass. Bottom row (2D-F) demonstrates 8-month follow-up axial CT (2D), axial PET (2E), and axial fused PET/CT (2F) images with significant interval progression of FDG-avid disease. New FDG ground glass lung opacities were favored to represent lymphangitis carcinomatosis (open arrowhead).

complete occlusion of the proximal esophagus. An immediate tracheostomy tube was placed, and a biopsy was performed. Histology showed a poorly differentiated carcinoma with NUT1 expression suggestive of NUT carcinoma. Axial CT (Figure 3A), attenuation corrected PET (Figure 3B), fused PET/CT (Figure 3C), and non-attenuation corrected PET (Figure 3D) demonstrated an intensely FDG avid lesion in the superior mediastinum. Later her respiratory symptoms progressed as the mass enlarged before she could be placed on chemotherapy, ultimately leading to her death a month later.

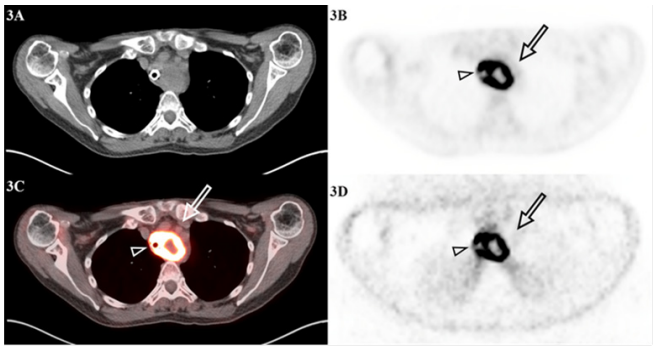

Figure 3: Patient 2 Axial FDG PET/CT images. Axial CT (3A), attenuation-corrected PET (3B), fused PET/CT (3C) and non-attenuation corrected PET (3D) images demonstrate contiguous intensely FDG avid masses in the superior mediastinum that encase the trachea (open arrowhead) and esophagus (open arrow).

Conclusion

NUT midline carcinoma is a rare and highly lethal carcinoma that is likely under recognized. In the very few published cases with FDG PET/CT as well as in the two atypically older patients presented here, these tumors can be very FDG-avid and FDG PET/CT may be useful in the diagnosis and staging of NMC.

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