Synchronous Pancreatic Solid Pseudopapillary Neoplasm Masquerading as Extralymphatic Involvement on ^{18}F-fluoro-2-deoxy-D-glucose Positron Emission Tomography/Computed Tomography in a Case of Hodgkin's Lymphoma

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Abstract

Solid pseudopapillary neoplasm (SPN) is a rare histopathologic variant of pancreatic tumors. Franz first described this tumor as a "papillary tumor of the pancreas, benign or malignant." In 1996, the World Health Organization named this tumor as SPN of the pancreas. It has a female preponderance with a male-to-female ratio of 1:9. A 30-year-old female who is a known case of lymphocyte-rich classic Hodgkin's lymphoma underwent ^{18}F-fluoro-2-deoxy-D-glucose (FDG) positron-emission tomography/computed tomography (PET/CT) for initial staging which showed intense metabolic activity in bilateral enlarged cervical and splenic hilar lymph nodes. Furthermore, intense metabolic activity was noted in hypodense lesion in the tail of the pancreas, and she was reported to be having Stage IIIE disease. Post chemotherapy, ^{18}F-FDG PET/CT showed disappearance of all previously metabolically active lymph nodes but persisting metabolically active lesion in tail of the pancreas. Hence, we reported as complete metabolic response of Hodgkin's lymphoma as per the Lugano criteria with suspected synchronous primary in the tail of the pancreas. Post distal pancreatectomy, histopathological examination and immunohistochemistry revealed the pancreatic lesion as SPN. SPN of the pancreas itself is a rare tumor and the presence of SPN in a patient with Hodgkin's lymphoma as synchronous primary is very rare. Due to the high density of mitochondria and the hypervascular nature of the tumor, there is an accumulation of ^{18}F-FDG in SPN tumor cells. Patients with SPN usually have a very good prognosis after surgery. The five-year survival rate is as high as 95%–97%.

Keywords: *^{18}F-fluoro-2-deoxy-D-glucose positron-emission tomography/computed tomography, Franz tumor, Hodgkin's lymphoma, solid pseudopapillary neoplasm*

Introduction

Solid pseudopapillary neoplasm (SPN) of the pancreas is a rare histopathologic variant of pancreatic tumors accounting for nearly 1%–2% of all exocrine pancreatic tumors.[1] Franz first described this tumor in 1959 as a "papillary tumor of the pancreas, benign or malignant." Since then, this tumor was referred to as Franz tumor and also by various other synonyms.[2] In 1996, the World Health Organization renamed this tumor as SPN of the pancreas. It is seen more commonly in females, with a male-to-female ratio of 1:9. Pancreatic body and tail are the most common sites of presentation. These tumors mostly occur in young women during the second to fourth decade of their life.[3,4]

Case Report

A 30 year old female patient presented with bilateral neck swelling of 6 months' duration associated with fever, loss of appetite, and significant weight loss of >10% in 6 months. She had no other complaints and no comorbidities. Biopsy and immunohistochemistry from cervical lymph nodes revealed lymphocyte-rich classic Hodgkin's lymphoma. ^{18}F-fluoro-2-deoxy-D-glucose (FDG) positron emission tomography/computed tomography (PET/CT) done for initial staging [Figure 1] showed intense metabolic activity in bilateral enlarged cervical and splenic hilar lymph nodes, largest measuring 3.0 cm × 3.6 cm with a maximum standardized uptake value (SUV_{max}) of

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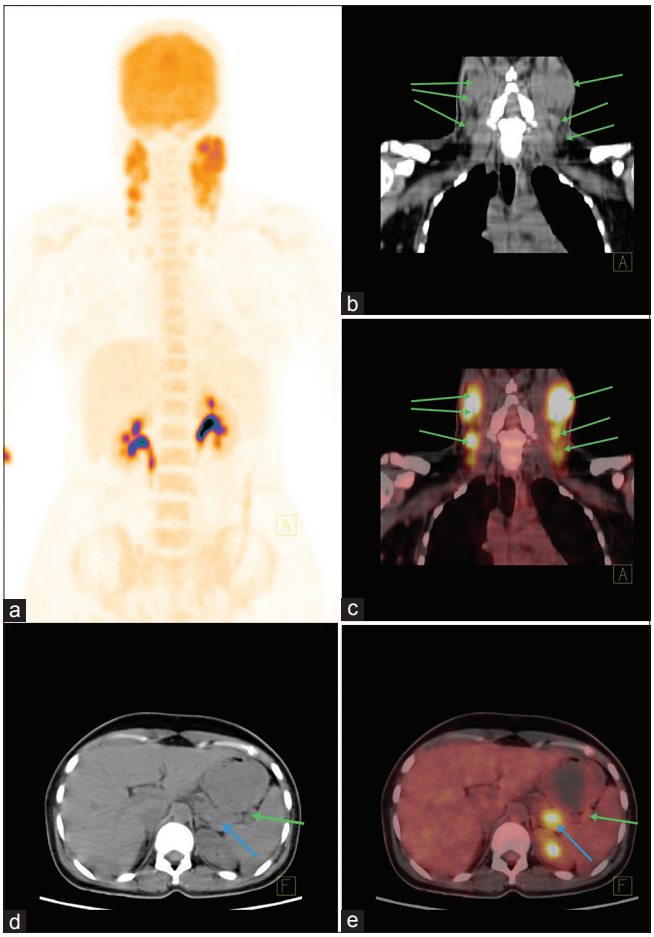

Figure 1: Initial staging ^{18}F-fluoro-2-deoxy-D-glucose positron emission tomography/computed tomography. (a) Maximum intensity projection image. (b and c) Coronal computed tomography and fused positron emission tomography/computed tomography images showing metabolically active bilateral cervical lymphadenopathy. (d and e) Axial computed tomography and fused positron emission tomography/computed tomography images showing metabolically active splenic hilar lymph node (Green arrow) and hypodense lesion in the tail of the pancreas (Blue arrow)

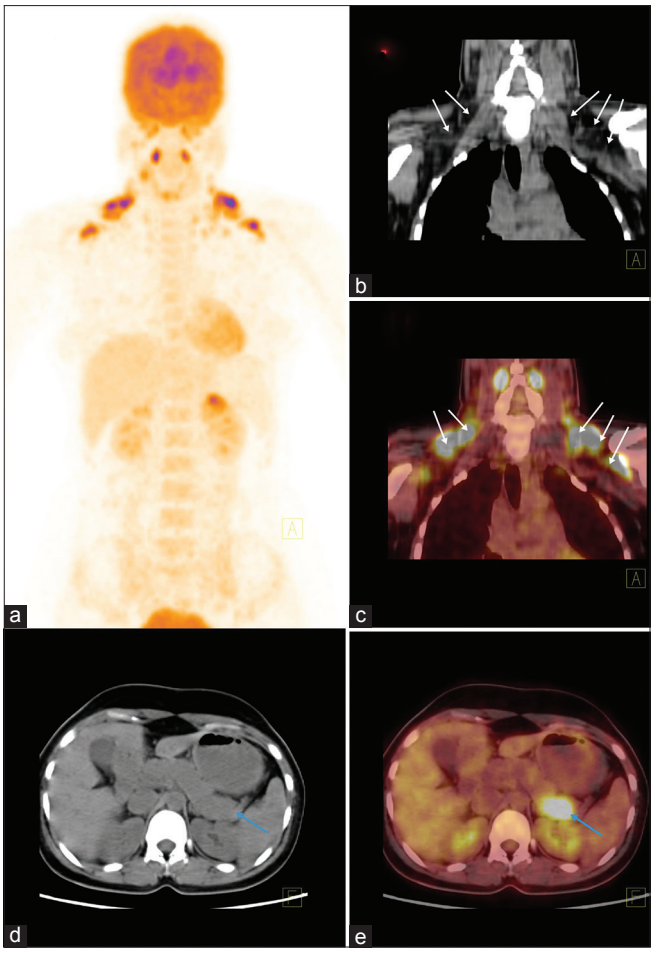

Figure 2: End of treatment ^{18}F-fluoro-2-deoxy-D-glucose positron emission tomography/computed tomography. (a) Maximum intensity projection image. (b and c) Coronal computed tomography and fused positron emission tomography/computed tomography images showing disappearance of all previously metabolically active cervical lymph nodes. Apparent bilateral metabolic activity in the cervical region is due to brown fat uptake (White arrows) which corresponds to fat density on computed tomography image (d and e) Axial computed tomography and fused positron emission tomography/computed tomography images showing disappearance of previously metabolically active splenic hilar lymph node but persistent metabolically active hypodense lesion in the tail of the pancreas (blue arrow)

15.7. Furthermore, intense metabolic activity was noted in hypodense lesion in the tail of the pancreas measuring 3.1 cm × 2.9 cm with SUV_{max} of 9.9, and she was reported to be having Stage IIIE disease (pancreatic involvement contiguous to splenic lymph nodes). She was treated with six cycles of ABVD which she tolerated well. End of treatment ^{18}F-FDG PET/CT [Figure 2] showed disappearance of all previously metabolically active lymph nodes but persisting metabolically active lesion measuring 3.4 cm × 3.1 cm in the tail of the pancreas with SUV_{max} of 12.9. Hence, we reported as complete metabolic response of Hodgkin's lymphoma as per the Lugano criteria with suspected synchronous primary in the tail of the pancreas.

Later, the patient underwent distal pancreatectomy and splenectomy. Intraoperatively, a 3 cm × 4 cm hard lump was seen in the distal pancreas. The lump was focally adhered to Gerota's fascia, and there was no omental or mesenteric lymphadenopathy and no liver metastases. Postoperative histopathological examination [Figure 3a and b] revealed a well-circumscribed cellular tumor arranged as pseudopapillary structures with focal areas showing sheets of cells and intervening areas of hemorrhage. The tumor cells showed intense nuclear positivity with β-catenin [Figure 4], moderate nuclear positivity with progesterone receptor, diffuse cytoplasmic positivity with alpha-1 antitrypsin, alpha-1 antichymotrypsin, nonspecific esterase, and vimentin confirming SPN of the pancreas.

Discussion

SPN of the pancreas itself is a rare tumor, and the presence of SPN in patient with Hodgkin's lymphoma as synchronous primary is very rare. To the best of our knowledge, this is the first case report in the available literature showing the presence of SPN in patient with Hodgkin's lymphoma as synchronous primary.

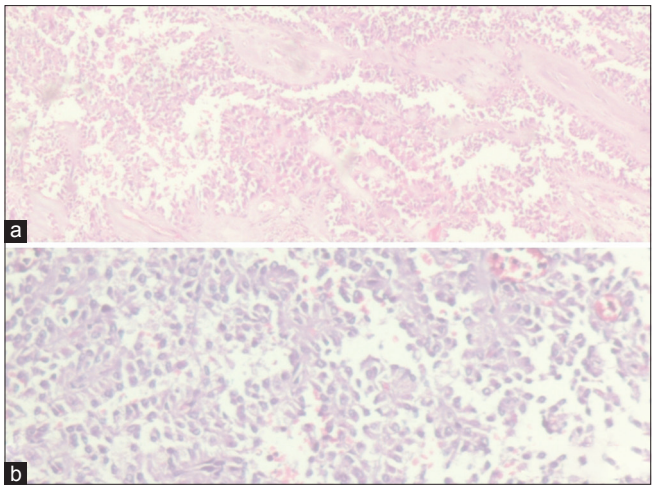

Figure 3: (a and b) Images show minimally cohesive, uniform monotonous cells lining delicate capillary-sized blood vessels giving a pseudopapillary architecture (H and E, ×100 and × 400)

SPN is frequently asymptomatic or minimally symptomatic. They can be visualized by many imaging modalities such as ultrasonography, CT, and magnetic resonance imaging.[5] Pathologically, SPN is usually a large, encapsulated mass composed of a mixture of cystic, solid, and hemorrhagic components. Both a capsule and intratumoral hemorrhage are important hallmarks in the diagnosis because these features are rarely found in other pancreatic neoplasms.[6]

^{18}F-FDG PET/CT is a good, noninvasive diagnostic imaging modality for differentiating pancreatic carcinoma from benign conditions, especially chronic pancreatitis. Malignancy in pancreatic masses can be suspected based on the ^{18}F-FDG uptake pattern with focal increased tracer uptake favoring malignancy and diffuse tracer uptake indicating benign pathology rather than relying on a single cutoff threshold of SUV_{max}.[7]

Genetically, the role of β-catenin in the pathogenesis of non-Hodgkin's lymphoma and SPN is documented. The key event in the WNT signaling pathway is the stabilization of β-catenin. In the absence of WNT signals, phosphorylation of specific serine and threonine residues in the amino-terminal region of β-catenin occurs. This phosphorylation marks β-catenin for ubiquitination and degradation by the proteasome. Signaling by WNT proteins results in the accumulation of β-catenin which will translocate to the nucleus. Here, it interacts with T-cell factor transcription factors to drive the transcription of target genes.[8] However, the role of β-catenin in the pathogenesis of Hodgkin's lymphoma is not known. Hence, the diagnosis of SPN in this patient of Hodgkin's lymphoma is mostly an incidental finding, and no genetic relationship can be attributed.

SPN is considered mostly as a benign neoplasm with a low-malignant potential. Local invasion to the adjacent structures or metastasis to other organs has been reported with 15%–20% of SPNs. Distant spread if present, is mostly to the liver and peritoneum.[9] SPN was described as malignant if they demonstrate extrapancreatic invasion, distant metastases, pancreatic parenchymal invasion, peripancreatic fat tissue infiltration, lymph node involvement, capsular invasion, or perineural or vascular invasion.[10]

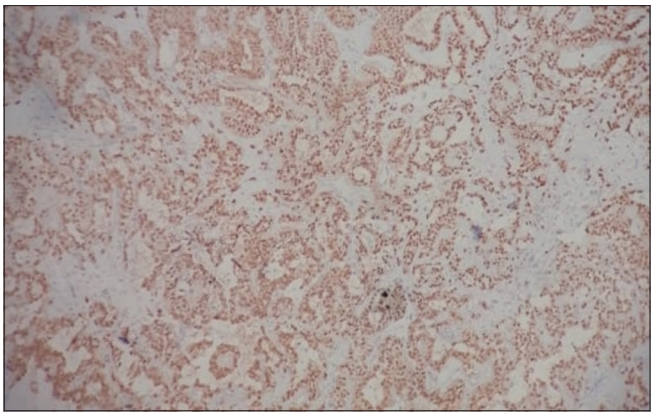

Figure 4: Immunohistochemistry with β-catenin showing intense nuclear positivity

The limited available information on the role of ^{18}F-FDG PET/CT in SPN reveals that SPN has a high uptake of 2-^{18}F-FDG, despite its benign nature. Normally, glucose transporter-1 (GLUT-1) and hexokinase-II (HK-II) play key roles in FDG uptake and trapping in the tumor cells. However, immunohistochemical staining showed that SPN tumor cells had a poor expression of GLUT-1 and moderate expression of HK-II, although they were rich in mitochondria. Due to the high density of mitochondria and the hypervascular nature of the tumor, there is an accumulation of FDG in SPN tumor cells. SPN tumors either at a benign or malignant stage can accumulate FDG more and was thus difficult to differentiate them from adenocarcinoma and neuroendocrine tumors of the pancreas based on only high SUV_{max}.[11]

Patients with SPN usually have a very good prognosis after surgery. More than 95% of patients with SPN limited to the pancreas are cured by complete surgical excision. The five-year survival rate is as high as 95%–97%, with an estimated 10-year survival rate of approximately 93%.[12]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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