Incidental finding of Psuedomyxoma Peritonei in Malignant Peripheral Nerve Sheath Tumour

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Abstract
Psuedomyxoma peritonei is an infrequent clinical entity characterised by intraperitoneal mucinous/gelatinous ascites produced by the cancerous cells. It has been associated with gastrointestinal, gynaecological, lung and breast tumours. It is commonly asymptomatic and is most often detected incidentally on abdominopelvic imaging or laparoscopy. Higher histological grade of the tumour shows increased metabolic activity on ¹⁸F-Fluorodeoxyglucose (FDG) positron-emission tomography (PET) computed tomography (CT). It has been rarely reported in patients with sarcoma. We hereby present an interesting case of incidentally diagnosed pseudomyxoma peritonei on ¹⁸FDG PET-CT scan of a patient with soft tissue sarcoma of peripheral nerve sheath.

Keywords: pseudomyxoma peritonei; ¹⁸FDG PET/CT; sarcoma

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Discussion
A 46-year-old patient, with malignant peripheral nerve sheath tumour (MPNST), underwent restaging PET-CT scan. The exact origin of MPNST was not known. He received multiple lines of chemotherapy followed by radiotherapy due to relapses. Restaging CT scan showed lower abdominal omental deposits, therefore, ¹⁸FDG PET/CT was done to evaluate full extent of

Figure: Coronal PET-CT images show a large mass in the central and left hemi abdomen causing pressure effect on the adjacent structures (A, B). There is heterogeneous metabolic activity (SUV 4.4) with peripheral areas of increased tracer activity and some areas within the mass (C, blue arrow). A satellite hypermetabolic nodule (SUV 6.3) was seen in the right hemi abdomen (D, E blue arrows). Due to the large central abdominal mass, pressure effect was seen on horseshoe with right sided hydrenephrosis and reduced cortical function. (D, E green arrows).
disease. However, there was a huge mass occupying most of the central and lower abdomen with nodular areas of FDG uptake within this lesion and along its periphery (Figure A-C). Additionally, a hypermetabolic right paracolic satellite lesion was seen (Figure D-E).

Psuedomyxoma peritonei is a rare mucinous neoplasm, which spreads into the peritoneal cavity in the form of ‘jelly like’ masses. Intra-abdominal mucous accumulation can lead to not only abdominal distention but also intestinal obstruction, malnutrition, cachexia, and eventually death. Karl Rokitansky first described this in 1842. It has a female preponderance and is most commonly associated with gastrointestinal tumours especially appendiceal mucinous tumours and with ovarian tumours. There are only a few case reports in literature in which psuedomyxoma peritonei was associated with non-epithelial neoplasm. One such case of peritoneal dissemination of intrabdominal sarcoma was reported by Klingler et al. Our case was a rare presentation in terms of being a male patient with peripheral nerve sheath soft tissue sarcoma. Thus, it highlights the usefulness of 18F-FDG PET/CT in MPNST and has a mention in a few guidelines on the subject.

References