Interesting image

An unusual case of metastatic extramammary Paget’s disease of the vulva identified by $^{18}$F-FDG PET/CT

Un caso inusual de la enfermedad de Paget vulvar metastásica identificada con $^{18}$F-FDG-PET/TC

G. Treglia$^a$,*, E. Giovannini$^b$, F. Bertagna$^c$, L. Giovanella$^a$, M. Malaggese$^d$

$^a$ Department of Nuclear Medicine and PET/CT Centre, Oncology Institute of Southern Switzerland, Bellinzona, Switzerland
$^b$ Nuclear Medicine, Private Practice, Rome, Italy
$^c$ Department of Nuclear Medicine, Spedali Civili and University of Brescia, Brescia, Italy
$^d$ Department of Gynecology, Fondazione Giovanni Paolo II, Campobasso, Italy

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A 61-year-old female patient with history of complicated inflammatory bowel disease underwent surgical excision of a vulvar extramammary Paget’s disease (EMPD). Surgical margins were clean. Three years after surgery, the patient underwent fluorine-18-fluorodeoxyglucose positron emission tomography/computed tomography ($^{18}$F-FDG PET/CT) for restaging due to the onset of vaginal recurrence demonstrated at gynecological examination and histology.

Before $^{18}$F-FDG injection, the patient had fasted for at least 6 h; at the time of the radiopharmaceutical injection she presented

A whole-body maximum intensity projection (MIP) $^{18}$F-FDG PET image (A), performed for restaging in a patient who underwent surgical excision of a vulvar extramammary Paget’s disease three years before, showed multiple areas of increased radiopharmaceutical uptake in the abdomen and pelvis. Fused PET/CT images in axial projection (B–G) showed increased $^{18}$F-FDG uptake corresponding to a vaginal mass (green arrow), multiple abdominal and pelvic lymph nodes (yellow arrows) and two liver lesions (red arrows). Histology on lymph nodal and liver lesions showed the presence of a metastatic extramammary Paget’s disease.

* Corresponding author.
E-mail address: giorgiomednuc@libero.it (G. Treglia).

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glucose blood levels corresponding to 93 mg/dL. Images were acquired 1 h after intravenous injection of 265 MBq of $^{18}$F-FDG according to the body mass index. Urinary catheterization was also performed.

$^{18}$F-FDG PET/CT showed multiple areas of abnormal radiopharmaceutical uptake in the abdomen and pelvis corresponding to a vaginal mass, multiple abdominal and pelvic lymph nodes and two liver lesions (Fig. 1).

Based on these PET/CT findings, the patient underwent histological examination of the liver and abdominal and pelvic lymph nodal lesions. Histological examination showed the presence of neoplastic infiltration of tumoral cells compatible with EMPD. A final diagnosis of metastatic EMPD was performed. Therefore the patient was addressed to chemotherapy.

EMPD is a rare malignancy and occurs most commonly in the anogenital region in older Caucasian patients. It can arise either as a primary intraepithelial adenocarcinoma with potential for invasiveness or as the secondary spread of an underlying internal malignancy to the skin surface.1–3

In most cases of primary EMPD, the tumor is confined in the epidermis and is generally slow growing. Although local recurrence is common after inadequate excision, systemic metastases are seldom reported in the literature. Surgery is the mainstay of treatment of EMPD, but systemic chemotherapy also has been used to treat advanced EMPD.1–3

Imaging is crucial to evaluate the presence of metastases in EMPD. Only few reports described the usefulness of $^{18}$F-FDG PET or PET/CT in staging EMPD localized to the anogenital region.1–3 This functional imaging method has been able to visualize both the primary tumor1 and the metastases2 in patients with EMPD.

In our case $^{18}$F-FDG PET/CT has been very useful in restaging an EMPD originating from the vulva showing multiple metastases in the abdomen and pelvis.

References