

Extensive Invasive Extramammary Paget Disease Evaluated by F-18 FDG PET/CT

A Case Report

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Abstract: Extramammary Paget disease (EMPD) is a rare cutaneous, intraepithelial adenocarcinoma. Because of its rarity, little is known about the value of fluorodeoxyglucose (FDG) positron emission tomography/computed tomography (PET/CT) in evaluating this disease. Our case report aims to increase current knowledge of FDG PET/CT in EMPD as a noninvasive imaging tool for assessing the extension of the disease and detecting distant metastases.

We reported a 64-year-old Chinese man who presented with a slowly progressive, ill-margined erythematous lesion with a crusted, eroded, and scaly surface involving multiple sites of penis, scrotum, left pelvic wall, hip, groin, and thigh for >4 years, which became extensive in the past 1 year. He was referred for an FDG PET/CT examination to further evaluate the lesions. A following skin biopsy was performed to obtain a definitive histological diagnosis.

FDG PET/CT imaging revealed mild FDG uptake at the extensive cutaneous lesion with subcutaneous invasion, involvement of lymph nodes, and multiple intense FDG-avid of skeletal metastases. According to the appearance of FDG PET/CT, a provisional diagnosis of advanced cutaneous malignancy was made. Histopathology findings indicated characteristic of EMPD. The patient was treated with radiation therapy and died from complications 2 months after the last dose of radiotherapy.

Our case highlighted that a whole-body FDG PET/CT should be incorporated into the diagnostic algorithm of EMPD to give a comprehensive assessment of this disease.

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Abbreviations: EMPD = extramammary Paget disease, FDG = fluorodeoxyglucose, PET/CT = positron emission tomography/computed tomography.

INTRODUCTION

Extramammary Paget disease (EMPD) is a rare cutaneous malignancy. The disease is often limited to the epidermis and mucosa, without invasion.¹ In rare instances, EMPD is

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reported to become invasive, occasionally infiltrating the dermis and even be highly metastatic to the regional lymph nodes and distant sites.^{2,3} Factors such as dermal invasion, involvement of regional lymph node, and distant metastases are indicators of poor prognosis and closely related to the management of the disease.⁴ Fluorodeoxyglucose (FDG) positron emission tomography/computed tomography (PET/CT) is of proved value in nodal staging and detection of distant involvement for malignant tumors. However, to our knowledge, the value of FDG PET/CT on evaluating EMPD has occasionally been reported in the literature.⁵⁻⁷

In this report, we describe a case of an old man with extensive invasive EMPD, in whom F-18 FDG PET/CT was valuable in assessing the extension of the disease and detecting distant metastases.

CASE REPORT

A 64-year-old Chinese man presented with a slowly progressive, ill-margined erythematous lesion with a crusted, eroded, and scaly surface involving multiple sites of penis, scrotum, left pelvic wall, hip, groin, and thigh for >4 years, which became extensive in the past 1 year (Figure 1). The obvious presenting features included pruritus and painful burning sensation. He also complained of swelling in the left thigh for >3 months and a history of muscular atrophy of disuse in the



FIGURE 1. Clinical features of the patient with extensive invasive EMPD. Ill-margined infiltrative erythematous lesion with a crusted, eroded, and scaly surface is covering on the extensive sites of penis, scrotum, left pelvic wall, hip, groins, and thigh. EMPD = extramammary Paget disease.

right leg due to encephalitis when he was 2 years old. He was admitted with complaints of progressive pain in shoulder, chest, and back for nearly 2 months' duration.

He was referred for an F-18 FDG PET/CT imaging to further evaluate the lesions. Whole-body PET/CT imaging was performed 60 minutes after the intravenous injection of 370Mq (10mCi) F-18 FDG and 8 hours fast using a PET/CT scanner (Discovery™ ST-16; General Electric Medical Systems, Milwaukee, WI, USA). The anteroposterior 3-D maximum intensity projection (Figure 2A) image demonstrated extensive and invasive abnormal FDG-avid lesions in the body. Selected coronal (Figure 2B) and transaxial (Figure 2C–M) views of CT (left column), PET (middle column), and PET/CT (right column) fusion images revealed mild FDG uptake at the extensive cutaneous lesion with subcutaneous invasion (Figure 2B–E), involvement of lymph nodes (Figure 2 F–I), and multiple intense FDG-avid of skeletal metastases (Figure 2J–M). It was highly

suggestive of malignancy, and a provisional diagnosis of advanced cutaneous malignancy was made.

These appearances of FDG PET/CT were not unique to EMPD and could also be seen in other cutaneous malignancies, even in an inflammatory or infective disease. Therefore, biopsy of the FDG-avid lesion was suggested to make a differential diagnosis. A following skin biopsy was performed to obtain a definitive histological diagnosis. Hematoxylin and eosin stain showed that the tumors were composed of cells with prominent nuclei and abundant lightly stained cytoplasm (suggesting Paget cells) (Figure 3). These pathologic findings were consistent with the EMPD.

EMPD has been traditionally managed with surgery, especially in the early stage of this disease. In our case, wide local excision is not appropriate because of the advanced stage of the disease that was successfully assessed by FDG PET/CT imaging. The use of radiation therapy was considered in this

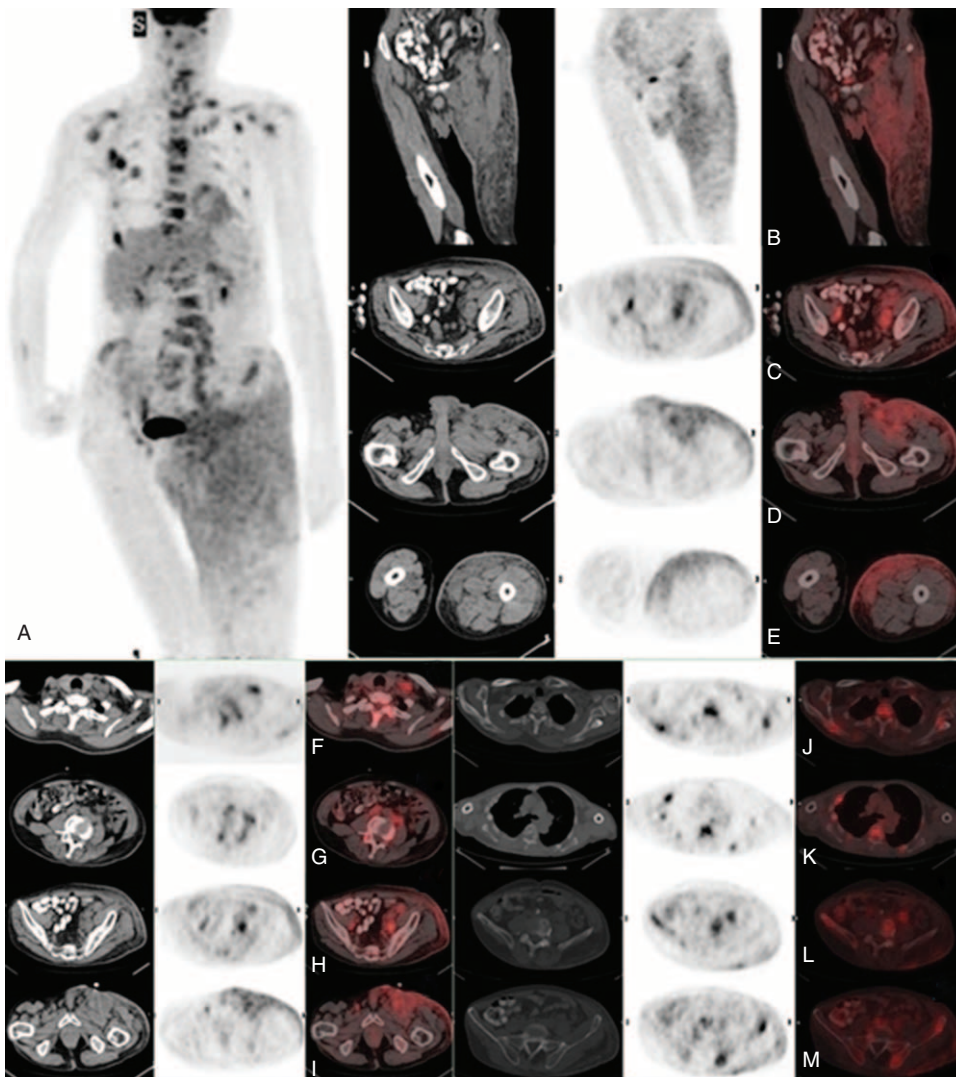


FIGURE 2. Whole-body FDT PET/CT images demonstrated the presence of multiple FDG-avid lesions in the (A) anteroposterior 3D-MIP. The selected (B) coronal and (C–M) transaxial images revealed mild FDG uptake at the extensive cutaneous lesion with (B–E) subcutaneous invasion, involvement of (F–I) regional lymph nodes, and (J–M) multiple intense FDG-avid of skeletal metastases. 3D-MIP = 3-dimensional maximum intensity projection, FDG = fluorodeoxyglucose, PET/CT = positron emission tomography/computed tomography.

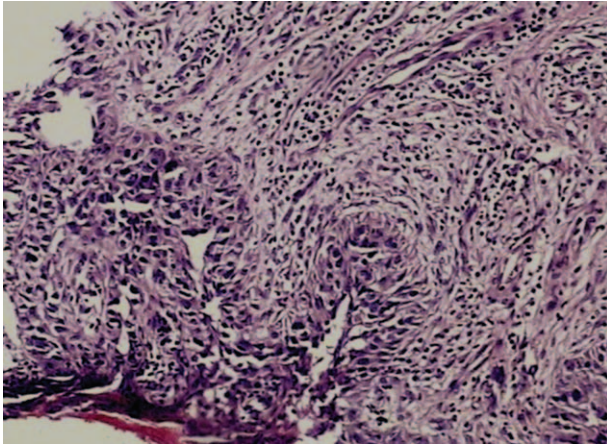


FIGURE 3. Histological assessment of EMPD. Hematoxylin and eosin stain ($\times 200$) showed that the tumors were composed of cells with prominent nuclei and abundant lightly stained cytoplasm (suggesting Paget cells). EMPD = extramammary Paget disease.

case. After receiving 4 weeks of radiotherapy, the patient suffered from anemia, hypoproteinemia, and sustained high fever probably caused by systemic metastases and died from complications 2 months after the last dose of radiotherapy.

Institutional approval was given by the head of the department, and informed consent was given by the patient.

DISCUSSION

EMPD is a rare cutaneous, intraepithelial adenocarcinoma involving primarily the epidermis but occasionally extending into the underlying dermis.² The clinical features of EMPD can be nonspecific and that misdiagnosis as an inflammatory or infective condition (eczema, psoriasis, or moniliasis) is very common. It was originally described in 1889 by Crocker,⁸ and it is most often observed in areas of apocrine gland-bearing, including the scrotum, penis, vulva, and perineal and perianal region. Other rare sites of involvement, such as eyelids, axilla, external auditory canal, and prostate have been reported.^{9,10} EMPD is often limited to the epidermis and mucosa¹; however, it can also become invasive, infiltrating the dermis and even metastasizing to the regional lymph nodes and distant sites, which is also uncommon.

Factors such as dermal invasion, presence of nodules in the primary lesion, and regional lymph node involvement are indicators of poor prognosis.⁴ The prognosis is good when the disease is confined to the epidermis and when it does not metastasize. However, in the presence of dermal invasion, the prognosis is poor.¹¹

Moreover, EMPD is sometimes associated with a concurrent underlying internal malignancy in nearly 20% of cases.^{12,13} Therefore, a thorough search for the metastatic lesions and underlying malignancy should be recommended before the treatment of EMPD.^{12,14,15}

Whole-body F-18 FDG PET/CT has the ability to identify foci of abnormally high metabolism, and FDG PET/CT has established itself as a valuable noninvasive imaging tool for diagnosing and staging, as well as a prognostic indicator of oncological patients; however, little is known about the use of FDG PET/CT imaging in evaluation of EMPD. Although increased FDG uptake on skin can also be nonspecific, and it

can mimic malignancies (cutaneous lymphoma, melanoma, and metastases from other internal malignancies), and an inflammatory or infective disease,^{16–21} our case indicated that FDG PET/CT is useful in evaluating EMPD by providing the extent of disease, lymph node involvement, and distant metastases, which is closely related to diagnosis, staging, management, and prognosis of this disease.

CONCLUSION

Extensive invasive EMPD is an extremely uncommon condition. Although the impact of FDG PET/CT on the staging and management of EMPD should be further evaluated, our case suggested that a whole-body FDG PET/CT should be incorporated into the diagnostic algorithm of EMPD to give a comprehensive assessment of disease and probably exclude the presence of coexisting internal malignancy.

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