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Inguinal and scrotal extramammary Paget's disease: ^{18}F-FDG PET/CT imaging

Dear Editor,

An 87-year-old man presented to our institution for investigation of an intertriginous rash, involving the left inner thigh, scrotum, and perineum, which had been neglected for a few years. The lesion was diagnosed as extramammary Paget's disease (EMPD). Due to the potential for EMPD to be associated with gastrointestinal and genitourinary malignancies, a thorough clinical and imaging evaluation was performed, the results of which were negative. The patient opted for symptomatic care only. Two years later, he returned to our institution with soft tissue swelling and edema of the left lower extremity, scrotum, and penis, with a nodular scrotal lesion and bilateral inguinal adenopathy (Figure 1). Positron emission tomography/computed tomography (PET/CT) showed ^{18}F-fluorodeoxyglucose (FDG)-avid lesions of the left scrotum, left inguinal lymph node, left pelvic lymph node, T12 vertebra, ribs, and left scapula (Figure 2). Biopsy of the left inguinal adenopathy showed signet-ring cells. Radiation therapy was initiated, resulting in partial improvement, followed by chemotherapy with carboplatin and paclitaxel. Unfortunately, the patient died, due to disease progression, at five months after the ^{18}F-FDG PET/CT imaging.

EMPD is a rare intraepithelial adenocarcinoma that typically gives rise to a pruritic rash at sites with numerous apocrine glands, such as the perineum, axilla, eyelids, scalp, and buttocks[1]. The disease occurs predominantly in patients over 50 years of age. In the Caucasian population, females are more affected than are males, whereas there is a predominance of males among EMPD patients in the Asian population[2]. The diagnosis of EMPD is based on the identification of Paget's cells with prominent nuclei and abundant lightly stained cytoplasm on hematoxylin-eosin staining[1,3]. The disease can arise from two major pathological mechanisms[4]: as an *in situ* intraepithelial adenocarcinoma which has the potential for local invasion and subsequent metastasis; and as pagetoid spread of a visceral

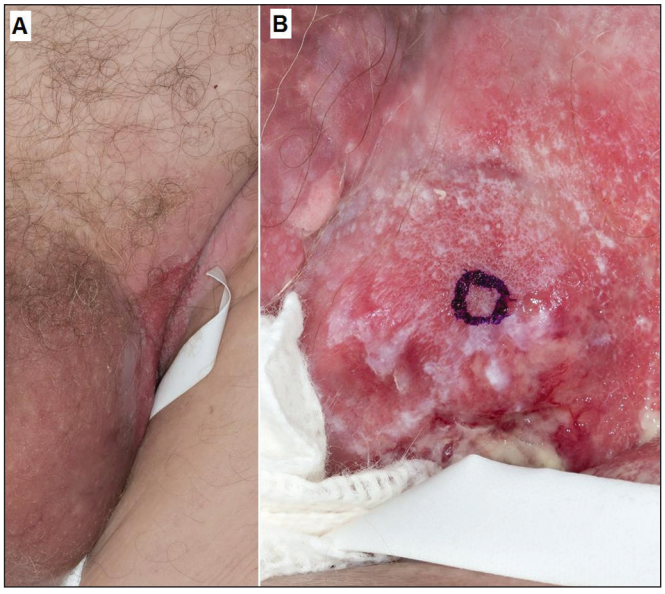

Figure 1. Photographs of a patient with EMPD. **A:** Lesions of the scrotum and left inguinal region. **B:** Close-up of the left inguinal lesion with a beefy red center and macerated whitish border.

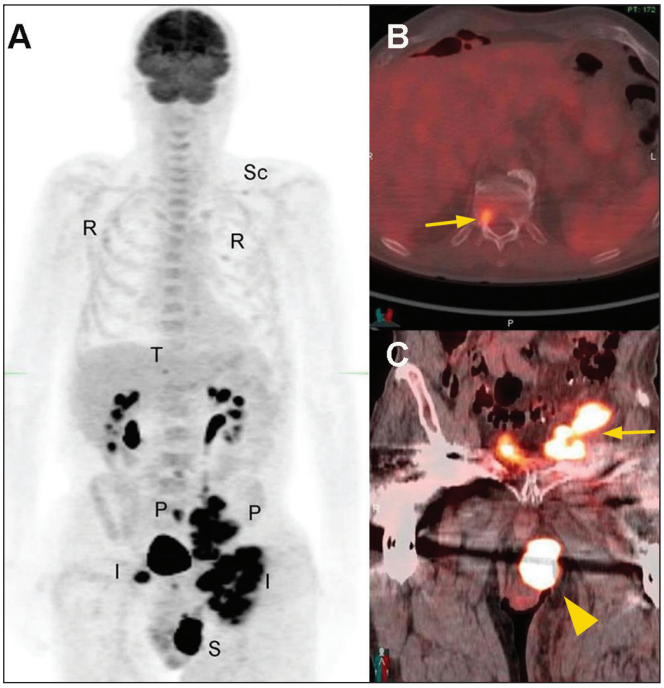

Figure 2. Maximum-intensity projection ^{18}F-FDG PET/CT (**A**) showing the left scrotal lesion (S), bilateral inguinal adenopathy (I), and pelvic adenopathy (P), as well as osseous metastases involving the T12 vertebra (T), multiple ribs (R), and the left scapula (Sc). Fused axial and coronal ^{18}F-FDG PET/CT images showing the T12 vertebra metastasis (arrow in **B**), together with the left scrotal lesion and left pelvic lymph node metastasis (arrowhead and arrow, respectively, in **C**).

malignancy. Isolated EMPD, without a coexisting internal primary lesion, is usually an indolent, slow-growing cancer that rarely metastasizes. Rare invasive EMPD has a propensity to metastasize to inguinal nodal basins[5]. EMPD involving the external genitalia has a strong association with gastrointestinal and genitourinary adenocarcinomas[4,6]. A small subset of invasive EMPD cases show signet-ring cell morphology with extracellular mucin. Immunohistochemical analysis establishes the distinction between signet-ring cells intrinsic to EMPD and those originating from coexisting visceral neoplasms[7,8]. Poor prognostic factors include dermal invasion, nodular skin lesions, lymph node involvement, and distant metastasis[3]. Given the multiple presentations of EMPD and their varying prognoses, there is a need to identify distant metastases and the primary visceral tumor: that effort is facilitated by functional ^{18}F-FDG PET/CT imaging[1–6].

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Congenital lobar emphysema

Dear Editor,

A 34-year-old asymptomatic woman underwent a chest radiography examination as an admission requirement for a new job. The X-ray showed focal hyperlucency in the left upper lobe of the lung (Figure 1A). High-resolution computed tomography (HRCT) was performed to confirm the findings (Figures 1B and 1C). The HRCT findings were characteristic of congenital lobar emphysema (CLE).

The evaluation by imaging methods in pediatrics has been the subject of a series of recent publications in the radiology literature of Brazil[1–6]. CLE is characterized by hyperinflation of one or more lung lobes in the absence of extrinsic bronchial obstruction[7]. It is a rare disease and its incidence is 20–30 cases/1000 births, most commonly affecting a single lobe of the lung (typically the left upper lobe), although multiple lobes or specific lobar segments may be involved[7,8]. The disease has a variety of causes, including bronchial cartilage deficiency (bronchomalacia) and endobronchial lesions, resulting in narrowing of the airway lumen and obstruction with air trapping, as well as progressive lobar overexpansion, usually with compression of the remaining areas of the ipsilateral lung[9].

CLE is generally diagnosed during early infancy, presenting with persistent progressive respiratory distress. It is known that CLE can occur in association with other malformations, especially cardiac malformations, which are present in 20% of cases[7]. In rare cases, it is diagnosed in adulthood and must be differentiated from other causes of localized pulmonary hyperlucency, because the treatments differ[9]. In such cases, the patients are usually asymptomatic and the disease can go unnoticed, resulting in underestimation of the true incidence of this condition.

Conventional chest X-rays are typically used in order to establish the diagnosis of CLE, showing a unilateral hyperlucent hemithorax. This finding is also present in a variety of other conditions, which include tension pneumothorax—the main differential diagnosis on routine chest radiography[7]—as well as

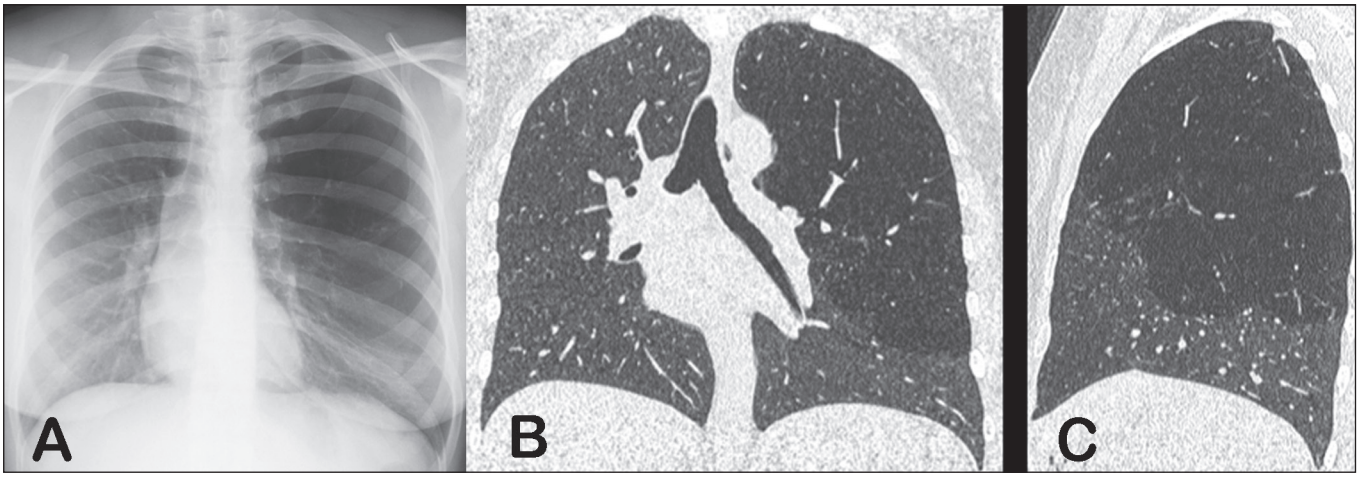

Figure 1. Anteroposterior chest X-ray (**A**) showing radiolucency and hyperinflation of the upper two thirds of the left lung. HRCT with coronal and sagittal reconstructions (**B** and **C**, respectively) showing hyperinflation of the left upper lobe of the lung, as well as vessel attenuation.