Clinical Case Report

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Purulent lupus panniculitis unmasked by FDG-PET/CT scan

A case report

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Abstract

Rationale: Lupus panniculitis (LP) is a unique variant of cutaneous lupus erythematosus. Clinical manifestations are typically mild and include erythema, nodules, and small ulcers. In certain cases, diagnosing LP may be challenging. Skin overlying the typical subcutaneous inflammation may appear normal, and bacterial superinfections of the skin sometimes mask the underlying LP. It has been suggested that a computed tomography (CT) scan may help to identify obscure LP lesions. Here, we report a case of a 54-year-old woman with an unusually severe form of LP, in which the full disease extent was only revealed by a fluorodeoxyglucose positron emission tomography (FDG-PET)/CT scan.

Patient concerns/Diagnoses/Interventions/Outcomes: Our patient initially presented with a bacterial infection of the skin. After initial improvement with antibiotic treatment, new erythematous lesions and sterile subcutaneous pus collections developed. An FDG-PET/CT scan revealed extensive subcutaneous inflammation at sites that had appeared normal during physical examination and on CT scan. As the subcutaneous lesions showed a remarkably linear pattern on FDG-PET/CT scan, the patient was suspected of having LP. After confirmation of this diagnosis by a deep-skin biopsy, our patient was treated with systemic glucocorticoids. Eventually, our patient succumbed to complications of LP and its treatment.

Lessons: Our case demonstrates that clinical manifestations of LP are not always mild and that timely diagnosis is needed. Furthermore, we show that obscure LP lesions are more readily identified on an FDG-PET/CT scan than CT scan.

Abbreviations: CRP = C-reactive protein, CT = computed tomography, DNA = deoxyribonucleic acid, FDG-PET = fluorodeoxyglucose positron emission tomography, LP = lupus panniculitis.

Keywords: FDG-PET/CT scan, infection, lupus panniculitis, panniculitis

1. Introduction

Lupus panniculitis (LP), also termed lupus erythematosus profundus, is a rare chronic form of cutaneous lupus erythematosus.[1] Symptoms are typically mild and may include erythematous plaques, subcutaneous nodules, and small ulcerations.[2,3] The subcutaneous lesions may adopt a characteristic linear configuration in rare cases.[4] Diagnosing LP may be challenging in certain cases. LP lesions are not always detected during physical examination, as the overlying skin may appear normal. Furthermore, bacterial superinfections of the skin can occur, which masquerade the underlying subcutaneous inflammation of LP.[5] We here report for the first time that a fluorodeoxyglucose positron emission tomography (FDG-PET)/computed tomography (CT) scan may reveal such concealed LP lesions. In a patient with an unusually severe and purulent form of LP, which has not been reported previously, an FDG-PET/CT scan demonstrated extensive subcutaneous inflammation at sites that had mostly appeared normal on physical examination and CT-scan. As these lesions showed a typical linear pattern on FDG-PET/CT scan, a diagnosis of LP was suspected. A deep-skin biopsy eventually confirmed this diagnosis.

1.1. Consent

The patient's husband signed the necessary documents to consent to the use of data for teaching and publication.

2. Case report

A 54-year-old woman with type 2 diabetes presented with an ill-defined erythema on her right foot and right hand. The erythema had developed 2 weeks earlier. The right foot also showed dry necrosis of the fourth toe, whereas a pus collection was observed on her right hand. She also complained of left shoulder pain. Examination of the shoulder revealed slight swelling but no erythema or limitation of movement. She had no fever or other remarkable symptoms. Blood testing revealed a C-reactive

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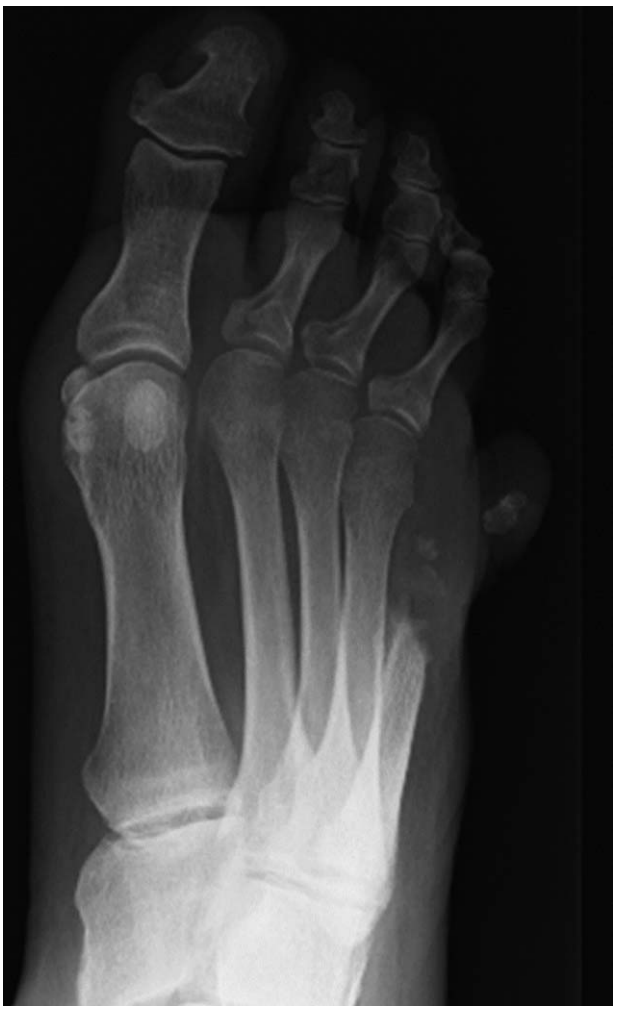

Figure 1. X-ray of right foot showing metatarsal osteomyelitis. Extensive destruction of the fifth metatarsal bone is present.

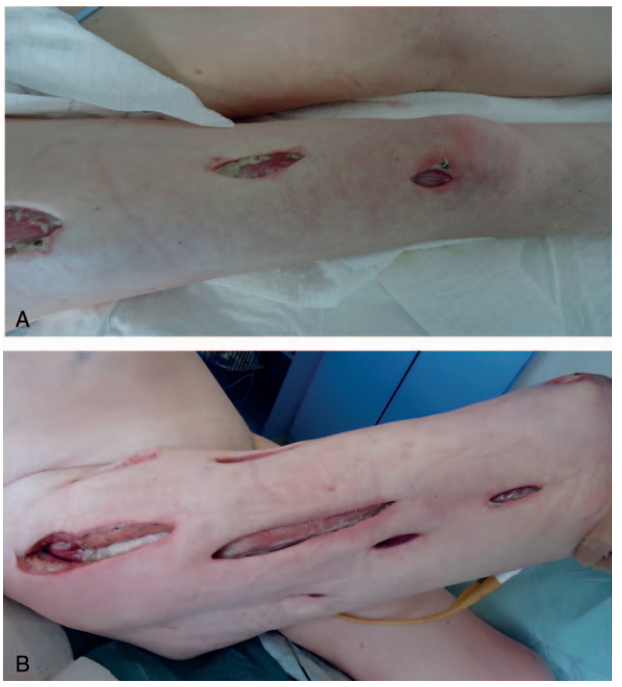

Figure 2. Appearance of upper leg after surgical drainage of subcutaneous pus. (A) Ventral view and (B) lateral view of multiple surgical skin defects on the right upper leg. The remaining skin appears relatively normal on inspection.

protein (CRP) level of 182 mg/L and leukocyte count of 10.6×10^9/L with >10% immature neutrophils. X-ray of her right foot showed extensive destruction of the fifth metatarsal bone (Fig. 1). Ultrasound of the left shoulder demonstrated subcutaneous edema, but no synovitis or bursitis. Cultures obtained from the blood and the pus of the right hand tested positive for *Staphylococcus aureus*. At this stage, our patient was diagnosed with a diabetic foot with extensive osteomyelitis caused by *S. aureus* that had disseminated to the skin of her right foot and hand. After empiric treatment with clindamycin and ciprofloxacin, the culture results prompted a switch to monotherapy with flucloxacillin. A transmetatarsal amputation of the fourth and fifth metatarsals was performed, and an infectious endocarditis was ruled out by transesophageal echocardiography. The patient initially seemed to respond well to the treatment, as indicated by a decrease in CRP levels to 44 mg/L.

After 4 weeks of antibiotic treatment, however, she gradually developed new erythematous lesions on both upper legs and her right hemithorax. When a small skin defect above her left knee discharged pus, a CT scan of her legs was performed to search for a subcutaneous abscess. No abscess was observed, but disappearance of subcutaneous fat was noted in a very small area above the left knee. Subsequently, the right upper leg developed a diffuse, fluctuant swelling. This finding prompted the surgeon to perform exploration of the right upper leg (Fig. 2). Upon incision, large amounts of subcutaneous pus were discharged from an area ranging from the knee to the hip. Importantly, the fascia appeared intact. Since the pus was regarded as evidence for a bacterial infection, the antibiotic regimen was replaced by meropenem and clindamycin. Eventually, neither gram stains and cultures nor deoxyribonucleic acid (DNA) sequencing of the pus revealed a microbial agent. During the workup, an FDG-PET/CT scan was performed to find a source for the presumed bacterial dissemination.

Although the FDG-PET/CT scan showed no bacterial dissemination source, it revealed extensive subcutaneous inflammation of both upper legs, the right hemithorax, left shoulder, and right hand (Fig. 3). The full extent of the subcutaneous inflammation was unexpected, as many sites had been unremarkable during physical examination (Fig. 2). The FDG-PET/CT findings were consistent with either fasciitis or panniculitis. As our patient was not critically ill and the fascia of the right leg had appeared normal during surgical exploration, we favored a diagnosis of panniculitis. We also noted that the subcutaneous lesions showed a remarkable linear pattern, which has only been reported in 1 type of panniculitis: LP. A deep-skin biopsy indeed demonstrated findings consistent with LP: lobular panniculitis with a lymphocytic infiltrate and hyaline fat necrosis without vasculitis. The only differential diagnosis of this histological pattern, a subcutaneous panniculitis-like T-cell lymphoma, was ruled out by additional findings: that is, lack of atypical lymphocytes, no typical localization of lymphocytes around fat cells, limited presence of histiocytes, and lack of clonality. Antinuclear antibodies were absent in the serum of our patient. Thus, our patient was diagnosed with an unusually severe and purulent form of LP.

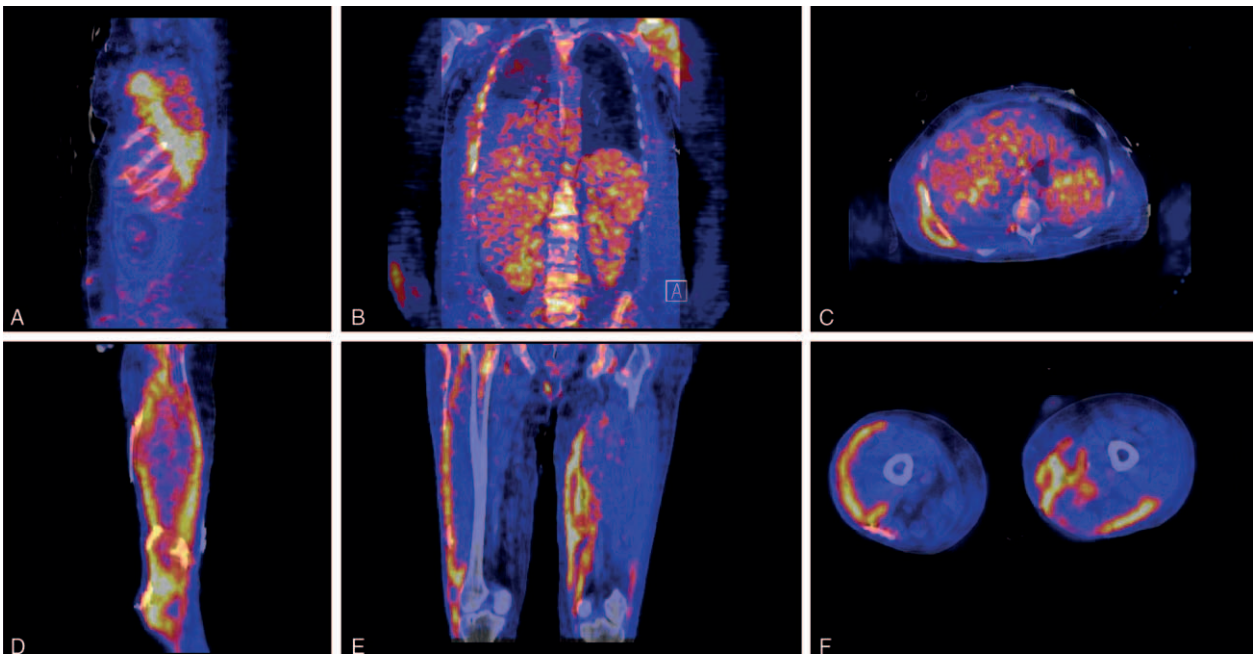

Figure 3. Fluorodeoxyglucose positron emission tomography/computed tomography scan showing extensive subcutaneous inflammation. (A) Sagittal view of the right hemithorax and (B) right upper leg. (C) Frontal view of the thorax/abdomen and (D) upper legs. (E) Transversal view of the lower chest/upper abdomen and (F) upper legs. Inflammatory subcutaneous lesions with a linear pattern were identified on the right hemithorax and both upper legs. In addition, subcutaneous inflammation was present at the left shoulder and right hand.

After LP was diagnosed, treatment with 1000 mg intravenous methylprednisolone was given for 3 days, followed by oral prednisolone 60 mg daily. After 2 months of treatment, our patient required hemodialysis as a result of progressive diabetic nephropathy and acute tubular necrosis resulting from septic shock due to a bacterial superinfection of the skin lesions. Hydroxychloroquine, other disease-modifying antirheumatic drugs, and biologicals were not given due to renal failure and the already prominent infection risk posed by the extensive skin defects. After 12 months, the prednisolone dose had been tapered to 10 mg daily. Her skin defects had healed poorly, despite various skin transplantations. Eventually, she developed recurrent episodes of hematemesis due to a nonhealing peptic ulcer, upon which she refused further medical treatment. Several weeks after discontinuation of hemodialysis, our patient died at home.

3. Discussion

Panniculitis is characterized by inflammation of subcutaneous fat. It should be distinguished from more superficial skin infections (i.e., cellulitis and erysipelas) on the one hand and deeper infections of the fascia (i.e., fasciitis) and muscle (i.e., myositis) on the other. Various forms of panniculitis have been described (Table 1). A deep-skin biopsy is critical to demonstrate inflammation of subcutaneous fat and to determine the type of panniculitis. Important considerations in the classification of panniculitis are the localization of the infiltrate (i.e., lobular vs septal), the presence of vasculitis, and the type of immune cells that predominate in the infiltrate.[6] In our case, we observed a lobular and lymphocytic infiltrate without vasculitis. These findings are consistent with LP.[3] The only differential diagnosis of this histological picture, a subcutaneous panniculitis-like T-cell lymphoma, was ruled out by additional immune phenotyping of infiltrating cells.[7]

LP is a rare form of cutaneous lupus erythematosus.[1] Approximately 50% to 90% of LP patients have detectable antinuclear antibodies serum titers, and 10% to 40% of LP patients fulfil classification criteria for systemic lupus erythematosus.[2,3] In some cases, LP may precede the development of systemic lupus erythematosus.[8] LP patients typically present with erythematous subcutaneous plaques, nodules, or small ulcers, although the overlying skin may sometimes appear

Table 1

Classification of panniculitis based on deep-skin biopsy findings[6].

Type	Infiltrate	Example
Lobular without vasculitis	Nearly absent	Sclerosing panniculitis
	Lymphocytes	Lupus panniculitis
	Neutrophils	Infective panniculitis, α1 antitrypsin deficiency
	Histiocytes, granulomatous	Subcutaneous sarcoidosis, gout panniculitis
Lobular with vasculitis		Vasculitis nodularis, rheumatoid panniculitis
Septal without vasculitis		Erythema nodosum, scleroderma panniculitis
Septal with vasculitis		Polyarteritis nodosa

Examples of different types of panniculitis are shown.

normal.[3–5,8] LP shows a predilection for the upper arms, shoulders, face, scalp, and buttocks.[2,3] Unlike other forms of panniculitis, LP lesions can adopt a linear configuration.[9] LP lesions are usually chronic but mild. Our patient suffered from a remarkably severe form of LP. We are not aware of prior reports on LP associated with extensive pus formation.

LP may be easily mistaken for a bacterial skin infection, especially when a secondary infection is present.[5] In the current case, the presence of subcutaneous pus was regarded as a clear sign for a bacterial infection. In retrospect, several findings had argued against a bacterial skin infection. Although an *S. aureus* was isolated from the initial cultures, subsequent gram staining, cultures, and even bacterial DNA sequencing were all negative. The subcutaneous edema around the left shoulder and the subtle disappearance of subcutaneous fat on CT might have been the first subtle clues for an underlying panniculitis. Moreover, the subcutaneous inflammation clearly outweighed the dermal inflammation and pointed toward panniculitis. Overall, our case underscores that pus is not specific for bacterial infections and may also occur in an autoimmune disease.

Eventually, the FDG-PET/CT scan was critical in diagnosing LP in our patient. The FDG-PET/CT scan revealed extensive inflammation of subcutaneous tissue, even at sites that had appeared mostly normal on physical examination and CT scan. The linear configuration of the lesions readily led to the diagnosis of LP. To our knowledge, this is the first report indicating a role for FDG-PET/CT scanning in diagnosing LP. Two earlier case reports suggested that subcutaneous LP lesions may be detected by a CT scan.[10,11] However, CT findings in our patient were unremarkable, despite the presence of extensive subcutaneous inflammation. Our report therefore suggests that an FDG-PET/CT scan might be more appropriate for identifying LP lesions than a CT scan.

In case series, LP is commonly treated with oral glucocorticoids and hydroxychloroquine.[3] This treatment is gradually tapered over a period of several months, but 75% of patients may develop a relapse after complete tapering.[3] In sporadic cases, LP has been treated with other agents such as methotrexate, ciclosporin, infliximab, and rituximab.[3,12–14] To date, no randomized controlled trials have been performed in patients with this rare disease. Our patient was treated with glucocorticoids only. Renal failure and a high infection risk precluded the use of hydroxychloroquine and other immune-modulating agents. Unfortunately, the glucocorticoids not only affected the healing of her extensive skin defects, but also that of her peptic ulcer, which eventually led to the decision to discontinue all medical treatments.

In conclusion, we describe a patient with an usually fulminant form of LP. Our case underscores that pus is not always a sign of bacterial infection, but may occur in autoimmune disease as well. When subcutaneous inflammation exceeds dermal inflammation, panniculitis should be suspected and lead to a deep-skin biopsy. Obscure subcutaneous lesions of LP are readily identified on an FDG-PET/CT scan.

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