Tender skin plaques exuding yellow oil

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A man presents with tender skin plaques exuding a yellow oily discharge. What is the cause of this and how can it be treated?

A 47-year-old man was admitted to hospital with a six-week history of multiple tender plaques localised to his upper back, abdomen, buttocks and thighs. The plaques exuded a yellow, viscous, oily discharge (Figure 1). He had developed a fever and malaise. A deep skin biopsy revealed interstitial neutrophilia undergoing leucocytoclasis between the collagen bundles in the dermis. There was intense oedema with a mixed inflammatory infiltrate within the deeper dermis and an associated separation of the underlying fat outlined by a granulomatous reaction (Figure 2).

Differential diagnosis

Panniculitis (inflammation of the subcutaneous tissue) may be a presentation of a variety of differing processes that can destroy the fat and may reflect systemic disease.

- **Infectious panniculitis** may present clinically as an indurated cellulitis, which may be associated with fever or disseminated intravascular coagulation. Background immunosuppression may be present. Bacteria, fungi or mycobacteria may be localised to the panniculus. Deep skin biopsy will reveal intense neutrophilia with abscesses or suppurative granulomas. Organisms may be visible but usually require culture for identification.
- **Connective tissue disease panniculitis**, particularly lupus erythematosus, may present as erythematous nodules and plaques that are particularly localised over the cheeks and shoulders. The lesions may ulcerate, and they often heal with skin depression, reflecting lipoatrophy. Skin biopsy demonstrates hyaline fat necrosis, lymphoid infiltration and lymphocytic vasculitis.
- **Pancreatic panniculitis** presents as tender erythematous nodules and plaques that may be associated with arthritis and hypereosinophilia. Circulating pancreatic enzymes produce diffuse fat necrosis with neutrophilia and calcification. Ulceration with oil discharge may be seen. Pancreatitis or pancreatic carcinoma may induce this reaction.

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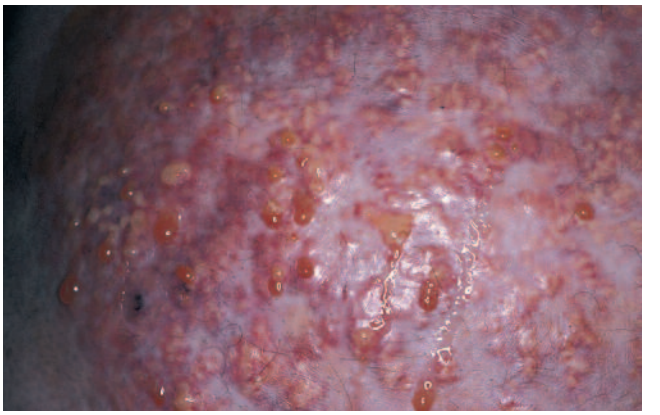

Figure 1. Diffuse plaque with prominent yellow oily discharge on the patient's buttock.

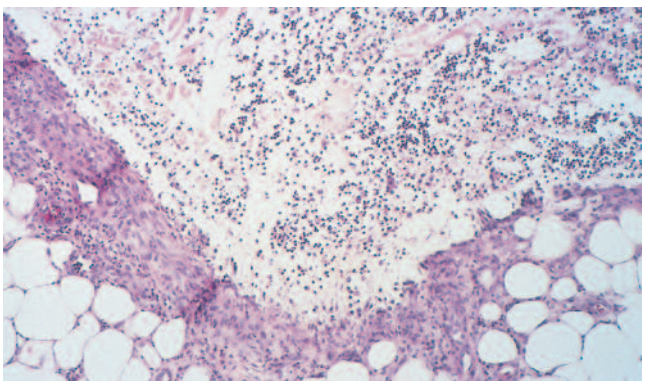

Figure 2. Skin biopsy demonstrating deep dermal oedema with a mixed inflammatory infiltrate. Dissection of the fat is outlined by a granulomatous reaction.

- **Alpha-1-antitrypsin (α1AT) deficiency panniculitis** is the correct diagnosis. It may be precipitated by local trauma or infection. This is a rare form of panniculitis seen in individuals who usually have a homozygous phenotype for this serum protease inhibitor. The low level of α1AT results in poorly regulated activity of neutrophil enzymes, resulting in lipolysis and elastolysis. Individuals with α1AT deficiency may develop emphysema, liver cirrhosis or arthritis. Measuring serum α1AT levels and determining the genetic phenotype will confirm the diagnosis.

Treatment

Dapsone or doxycycline are effective treatments. In severe recalcitrant cases, recombinant α1AT infusions may be used but these are expensive.

Keypoint

Panniculitis with oily discharge is an indicator for measuring α1AT levels because treatment can be directed at reversing the fat destruction. MT