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PHOTO QUIZ

A Sézary cryptogram

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CASE REPORT

A 77-year-old women was admitted to the haematology department because of fever. Her medical history consisted of a cutaneous T cell non-Hodgkin's lymphoma (Sézary syndrome) diagnosed in 2003, for which she had received several lines of chemotherapy, but was currently only taking prednisone 5 mg twice daily. The day before presentation she had collapsed after which she started to complain of headaches. Physical examination revealed no neurological abnormalities, in particular no signs of meningitis. There were no signs of a urinary, abdominal or pulmonary infection. The skin showed features of the Sézary syndrome, but also a large red swelling (20 cm in length) on the inner left thigh. This lesion was sharply demarcated, and painful on palpation. The laboratory results showed an elevated C-reactive protein of 62 mg/l (normal value <5 mg/l) and a leucocyte count of 9.4×10^9/l (with 72% neutrophils and 21% lymphocytes). A chest X-ray showed no abnormalities, and cerebral scanning CT revealed no intracerebral haemorrhages or lesions. The lesion was surgically drained, as shown in *figure 1*. Purulent material was sent to the microbiology laboratory for microscopic investigation and culture. The microscopy is shown in *figure 2*.

Figure 1. *Skin lesion*

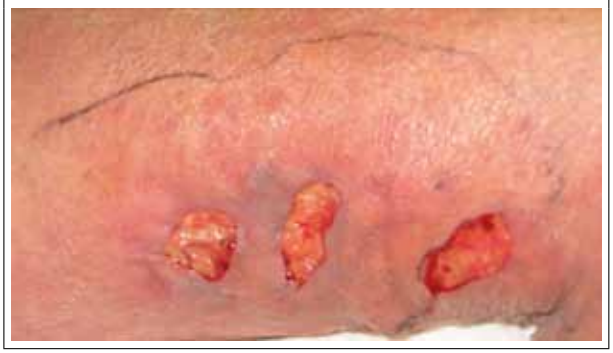


Figure 2a. *Indian ink staining*

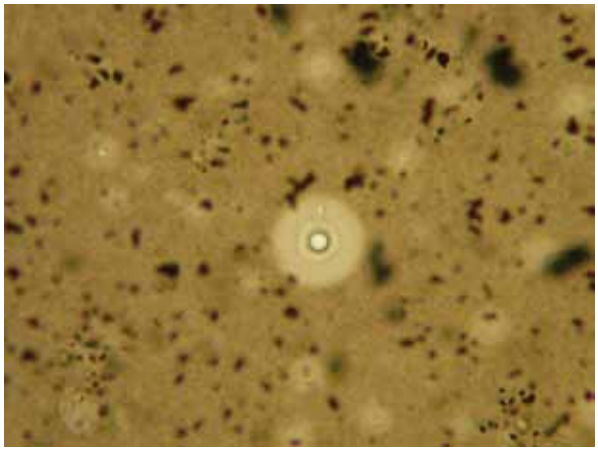
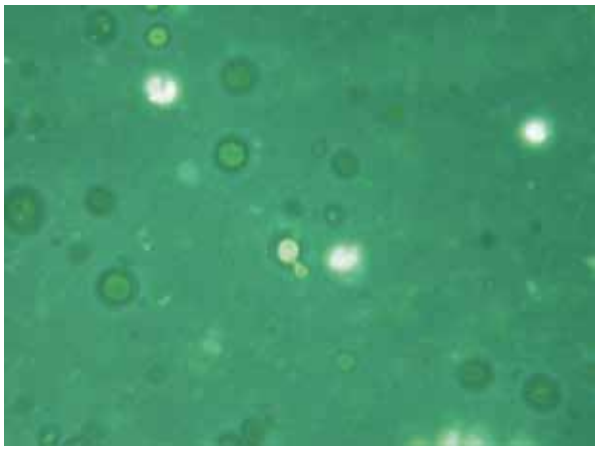


Figure 2b. *Blankophor P staining*



WHAT IS YOUR DIAGNOSIS?

See page 349 for the answer to this photo quiz.

ANSWER TO PHOTO QUIZ (PAGE 346)

A SÉZARY CRYPTOGRAM

DIAGNOSIS

The Indian ink staining (*figure 2a*) showed round yeast cells surrounded by a halo, and a blankophor P stain showed narrow-based budding (*figure 2b*), both consistent with cryptococcosis. The culture finally revealed *Cryptococcus neoformans*. Blood cultures taken on admission also became positive, and the serum cryptococcal antigen titre was 1:1024. Within two days after admission the patient developed progressive neurological signs, including headache, drowsiness, palsy of the facial nerve, and nausea. A lumbar puncture was performed and the cerebral fluid contained *Cryptococcus*. So we concluded that this patient suffered from a disseminated cryptococcal infection with fungaemia, meningo-encephalitis, and skin abscesses.

Only one previous case of a patient with a Sézary syndrome suffering from a cutaneous cryptococcal infection (*Cryptococcus albidus*) has been described, underlining the rareness of the complication.[1] In addition, also in other haematological malignancies *Cryptococcus neoformans* infections, such as pneumonia and meningitis, have been reported only sporadically.[2] Sézary syndrome is characterised by the classic triad of generalised erythroderma (>80% of the body surface area), generalised lymphadenopathy or other systemic manifestations of the lymphoma, and the presence of more than 5% of large atypical T cells with cerebriform nuclei (called Sézary cells).[3] Nowadays, the diagnosis is largely based on the combination of the typical clinical picture with erythroderma and flow cytometry of the blood showing a clone of T cells with a CD4+ phenotype with aberrant surface marker expression. Patients with Sézary syndrome suffer from compromised specific cellular immune responses. The pathogenesis of this immune deficiency is regarded to be multi-factorial, but the use of prednisone was probably the most important factor causing the immunocompromised status in this patient.[4,5]

Our patient was initially treated with a combination of liposomal amphotericin B and fluconazole, followed by the combination of flucytosine and fluconazole because of unsatisfactory clinical recovery. She recovered with improvement of the neurological signs. Long-term maintenance therapy with fluconazole 400 mg, once daily, was started at discharge, because of the persisting immune deficiency, resulting from the necessity to continue prednisone for the Sezary syndrome.

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