She was conditioned for an allogenic HLA identical bone marrow transplant from a sixteenth-cousin with a total body irradiation (12 Gy) and cyclophosphamide (60 mg/kg body weight × 2); as prophylaxis for graft-versus-host disease, she received cyclosporin A (1.5 mg/kg body weight every 12 h, starting on day -1) and a course of methotrexate, according to the Seattle protocol. On the day of the bone marrow infusion, she developed fever and was put on broad-spectrum antibiotics and a regimen with amphotericin B, which was added later. Her histological study and chest X-ray were normal. She had, meanwhile, progressive weight gain, abnormal liver function test (increased of bilirubin and γ-glutamyl transaminase) and deterioration of renal function. By day +10 she had ascites, tender hepatomegaly and oedema of her lower limbs, all suggestive of hepatic veno-occlusive disease. A thoraco-abdominal computed tomography (CT) scan was carried out which showed hepatosplenomegaly, ascites and retroperitoneal lymphadenopathy. Soon after the transplant she developed severe mucositis. On day +13 she was dyspnoeic and a chest X-ray showed consolidation in the lower left lobe, left pleural effusion and a poorly defined nodule in the upper right lobe. Two days later, the nodule enlarged and consolidation at the base of both lungs with bilateral pleural effusion were observed. Because of the suspicion of pulmonary aspergillosis, the dose of amphotericin B was increased.

On day +20, neutrophil engraftment was detected (neutrophil count >0.5 × 10^9/L). On day +22, the patient presented respiratory distress syndrome and was transferred to the Intensive Care Unit, where she eventually required mechanical ventilation, and died six days later (day +30).

Necropsy showed right pulmonary aspergillosis, radiation pneumonitis with severe diffuse alveolar damage, hepatomegaly with severe cholestasis and venous wall oedema, visceral congestion, and intense generalised mucositis.

The role that the precedent of toxic oil syndrome played in the post-transplant course of this patient is a matter of speculation.

Criteria for the diagnosis of toxic oil syndrome

1. Intake of cooking oil sold in bulk, and/or other cases among relatives
2. Interstitial-alveolar pattern on chest X-rays during the first 4 months
3. Esophagitis (>≥500 eosinophils/μl), myalgia, or rash during the first 4 months
4. Pulmonary hypertension, hepatic disease, scleroderma, sicca syndrome, polyneuropathy, joint contractures, Raynaud disease, muscle cramps, chronic lung disease

For diagnosis, either
- at least two of criteria 1, 2, or 3 or
- at least criteria 1, 2, or 3, and two or more of the features included in criterion 4 should be fulfilled.

Proptosis, skull infarction and epiperal haematoma in sickle thalassemia

Sir,

Infarction of the long bones is a common complication of sickle cell disease. However, there are less than 10 cases of orbital bone infarction associated with sickle cell disease in the English literature. A combination of orbital and skull bone infarction with associated epiperal haematoma in sickle cell disease has been reported only once in the world literature, with our case being the second. This case is unique in that it is sickle thalassemia and hence occurred at a later age.

A 35-year-old man, admitted with painful limitation of eye movements and proptosis of the left eye of eight days duration, had absent extraocular movements in all fields of gaze with the cornea, sclera, fundus, pupils and visual acuity being normal. The patient had a haemoglobin of 8.1 g/dl and sickle cells on a peripheral smear. Sickleting test was positive and haemoglobin electrophoresis revealed Hb S >48.8%, Hb F ≥49.0% and Hb A2 <2.2%, consistent with a diagnosis of sickle thalassemia. A bone marrow biopsy depicted areas of infarction. The left orbital ultrasonography demonstrated a mass with soft tissue echotexture in the upper temporal quadrant of the left eye, displacing the eye ball inferomedially. A computed tomography (CT) scan of the orbit showed an non-enhancing soft tissue mass without any adjacent bone erosion, destruction or intracranial extension (figure). CT scan of the brain showed an epidural haematoma and swelling secondary to it. The patient received hydroxyurea, blood transfusion and analgesics. After 14 days he regained full movements of the eye and proptosis disappeared.

Sickle cell disease is rarely associated with periorbital and skull bone infarctions.

Though intracranial bleeding occurs in sickle cell disease, a combination of epiperal haematoma and periorbital and skull infarction has only been reported once before.

One should include periorbital bone infarction due to sickle cell disease in the differential diagnosis of periorbital swelling. It is often difficult to differentiate bone infarction from oseomyelitis and a 99mTc MDP bone scintigraphy is useful in demonstrating activity-deficient areas in infarction and hyperactive areas in osteomyelitis. This is important as both can coexist in sickle cell disease and clinical differentiation is often difficult. The combination of an orbital CT scan and orbital ultrasonogram are useful in differentiating the various orbital problems in such cases.

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Figure CT scan of the left orbit showing non-enhancing soft tissue mass

Periorbital swelling: causes

- trauma
- infarction
- exophthalmic goitre
- nephritis
- lacrimal gland involvement
- glaucoma
- angioneurotic oedema
- bleeding due to blood dyscrasias
- leukemic infiltration
- metastatic neuroblastoma
- cavernous sinus thrombosis