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A 28-Year-Old Man from Ghana With a Chronic Ulcer on His Ankle

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Clinical Presentation

History

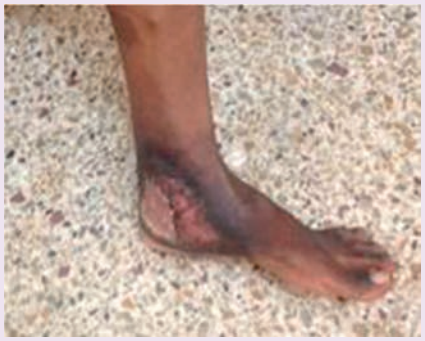
A 28-year-old West African man presents to a clinic in Ghana complaining of a painful ulcer on his left ankle. The ulcer has been present for the past 4 months and is not healing (Fig. 62.1). There is no history of prior trauma; however, he had a previous ulcer on his right ankle some years ago which took about a year to heal. He also complains of pain in his right thigh and in both knees.

Clinical Findings

The patient is short for an adult; he is pale and has a tinge of jaundice. The ulcer is on his left ankle, next to the medial malleolus; the skin surrounding it is hyperpigmented. There is tenderness in both knee joints and in his right thigh. The rest of the physical examination is normal. Vital signs: Temperature 36.6°C, pulse 88 bpm, blood pressure 110/70 mmHg.

Questions

1. What is your differential diagnosis?
2. How would you confirm the diagnosis?



• **Fig. 62.1** The patient's left leg showing an ulcer surrounded by hyperpigmented skin on the medial malleolus.

Discussion

A 28-year-old West African man presents with a chronic, painful ankle ulcer. He had a similar ulcer on the other foot a few years prior. He also complains of bone pains. He is short for an adult, pale and mildly jaundiced.

Answer to Question 1

What is Your Differential Diagnosis?

The chronic nature of the ulcer, its site and the surrounding hyperpigmentation make a venous ulcer likely. The patient's bone pain, jaundice and pallor could point to a haemolytic anaemia. Various hereditary haemolytic anaemias may be complicated by chronic leg ulceration, e.g. haemoglobinopathies (thalassaemia and sickle cell disease), spherocytosis and pyruvate kinase deficiency. In the West African context, sickle cell disease is the most likely diagnosis.

Further differentials would be tropical ulcer, diabetic ulcer or chronic osteomyelitis with a discharging sinus. Buruli ulcer, caused by *Mycobacterium ulcerans,* is usually painless. Other mycobacterial infections can also present with chronic skin ulcers. Pyoderma gangrenosum and malignant diseases also need to be considered.

Answer to Question 2

How Would You Confirm the Diagnosis?

A full blood count and a peripheral blood film should be done. In sickle cell disease, during an acute crisis, abundant sickled red cells can be seen on a blood film. Other characteristic but non-specific features include target cells, Howell–Jolly bodies, polychromasia and nucleated red cells. The presence of HbS can be demonstrated by using a simple sickle slide or solubility test. Blood is mixed with sodium metabisulphite, which will provoke sickling of cells containing HbS; this can be demonstrated on a slide. If resources allow, confirmation is by haemoglobin electrophoresis, liquid chromatography or isoelectric focusing.

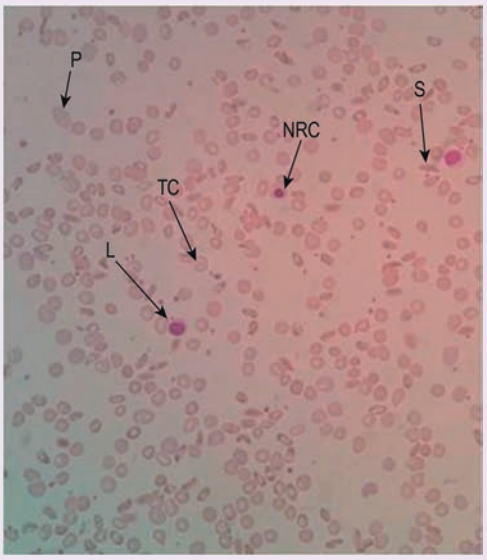
Fasting blood sugar and wound swab for culture and sensitivity as well as a radiograph of the left leg should be done to rule out other differential diagnoses and osteomyelitis.

The Case Continued…

Blood was taken and the available results are shown in Table 62.1. The patient's blood film is shown in Figure 62.2. The sickling test was positive and haemoglobin electrophoresis showed a homozygous HbSS-type. The wound swab grew *Pseudomonas* species sensitive to levofloxacin. Radiography showed a periosteal reaction with slightly sclerotic bones.

TABLE 62.1 Laboratory Results

Parameter	Patient initial visit	Patient 6 years later	Reference Range
WBC ($\times 10^9$/L)	17.6	9.2	4–10
Haemoglobin (g/dL)	6.9	8.3	13–15
Platelet ($\times 10^9$/L)	Adequate	364	150–400
Fasting blood glucose (mmol/L)	4.8	Not done	4.4–6.1



• **Fig. 62.2** Patient's blood film: irreversibly sickled cells (S), polychromasia (P), target cells (TC), nucleated red cells (NRC) – note similarity with lymphocyte (L). Adequate platelets.

The diagnosis made was sickle cell disease with a chronic ankle ulcer. The patient was managed with alternate daily wound dressing using normal saline irrigation and povidone-iodine. High white cell counts are commonly seen in sickle cell disease. They may be the result of bone marrow stimulation and do not necessarily indicate systemic infection. However, in view of the patient's generally poor condition and the *Pseudomonas* grown from his wound swab, it was decided to give him systemic antibiotic treatment.

Strict bed rest was difficult to enforce because the young man was self-employed and could not afford to take the required time off work. His recurrent bone pain was managed conservatively with good hydration, prompt treatment of infections and relief of other identifiable precipitants of crises. He received daily folic acid supplementation. Pain relief was achieved with paracetamol and tramadol.

However, on follow-up 3 years later, the ulcer was still not healed. Grafting was under consideration, when healthy granulation tissue would be achieved.

Six years later, the wound had still not healed and the patient could not afford a skin graft. He had 3 times weekly dressing and felt well in himself. His WBC was normal and his Hb had slightly increased (see Table 62.1).

SUMMARY BOX

Leg Ulcers in Sickle Cell Disease

Sickle cell disease (SCD) is a collection of autosomal-codominant genetic disorders characterized by the production of abnormal sickle haemoglobin S (HbS). Homozygous HbSS leads to sickle cell anaemia, the most severe form of SCD. Sickle cell disease is the commonest hereditary haematological disorder.

HbS has the tendency to polymerize during hypoxia. This leads to a reduction of the flexibility of the erythrocyte and to the typical sickle shape of the affected cell. Sickled red blood cells lead to haemolysis and vaso-occlusion.

The disease is characterized by episodes of acute illness against a background of progressive organ damage. Any organ can be affected by SCD at any age; however, certain features tend to predominate in certain age-groups. Leg ulcers tend to manifest in adulthood. Pathophysiology is complex and remains incompletely understood. Ulcers in SCD occur in areas with thin skin and little subcutaneous fat, most commonly on the ankles. They are notoriously difficult to treat. The ulcers are slow to heal and are characterized by unexplained relapses. They are commonly very painful and patients may occasionally require opioids for pain control. Colonization with pathogenic bacteria is common. Periosteal reaction is usually seen in the underlying bones but osteomyelitis is uncommon.

Of the many treatments, the most certain to aid healing is complete bed rest with leg elevation. Oral zinc sulphate tablets (200 mg tid) have been shown to be helpful. Systemic antibiotic therapy is given in acute sepsis. The use of hydroxyurea in leg ulcers is controversial. Chronic transfusion regimens to maintain the haemoglobin level above 10 g/dL may help when conservative therapy fails. Skin grafting for clean wounds is recommended, especially when the defect is large. The relapse rate is high, however. Poor nutrition which has not been given much attention may be a crucial factor and needs to be investigated on a wide scale in sickle cell disease.

Further Reading

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