# Hemoglobin SC disease and proliferative retinopathy

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Retinopathy is one of the clinical signs of hemoglobin SC disease.[1] It is often misunderstood and diagnosed late because it is initially asymptomatic due to its preferential localization in the periphery of the retina.[2,3]

A 25-year-old man from the Ivory Coast was admitted because of an abdominal pain crisis. The patient, who had sickle cell trait, had mild scleral jaundice and pronounced splenomegaly (17 cm at ultrasound examination).

Laboratory findings revealed mild microcytic anemia (Hb 12.4 g/dL, MCV 71 fL), reticulocytosis ($107\times10^9$/L), increased bilirubin (T 1.66 mg/dL, I 1.38 mg/dL), low haptoglobin level and high LDH (1145 U/L). Serum ferritin, iron and TIBC were in the normal range.

Hemoglobin study by high performance liquid chromatography (HPLC) revealed the presence of two abnormal bands corresponding to hemoglobin S and hemoglobin C. The sickling test was positive. The peripheral blood examination showed a large number of target cells and Hb C crystals in red cells. Due to the microcytosis with a normal iron status, globin chain synthesis and molecular studies of $\alpha$-globin gene clusters were performed and the presence of $\alpha$-thalassemia was excluded. The cause of microcytosis in our patient is still under evaluation. The fundus oculi of the patient, who had never reported any disturbances of vision, revealed ischemic areas and exudates, arterio-venous anastomoses and neovascular proliferation (*sea-fans*) in the four quadrants of the periphery of the retina (Figure 1). The patient underwent photocoagulative treatment with argon laser; sea-fans and ischemic areas disappeared completely and the retinal exudates were reabsorbed (Figure 2).

Since vitreous hemorrhages and retinal detachment can rapidly cause blindness,[3,4] all sickle cell patients, even in the absence of visual symptoms, should have careful ophthalmological examinations. This could prevent major complications and might identify patients who could benefit from more treatment of their sickling, possibly with non-conventional therapy (e.g. hydroxyurea).

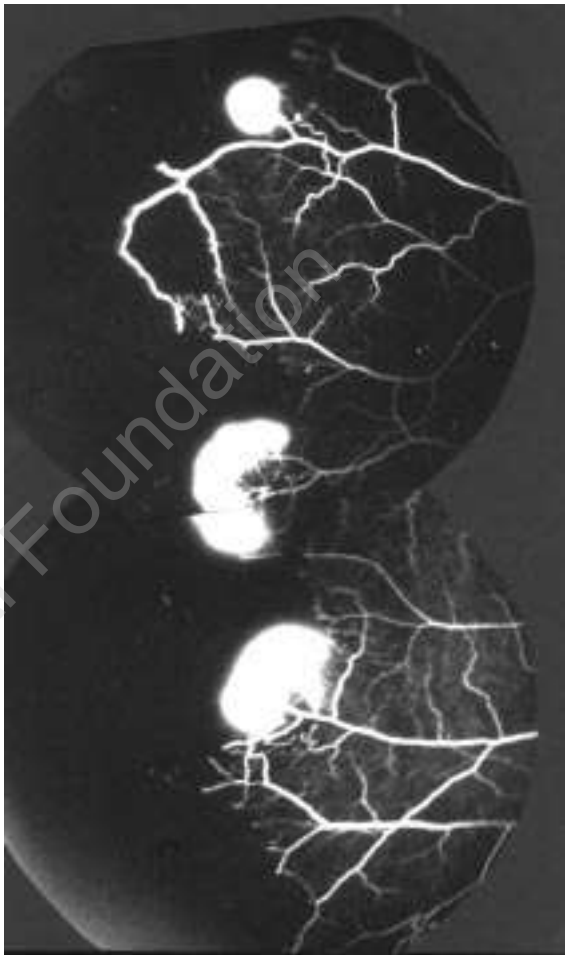

**Figure 1. Fluorescein angiogram in early phases shows closure of peripheral retina, arteriovenous anastomoses and *sea-fans*.**

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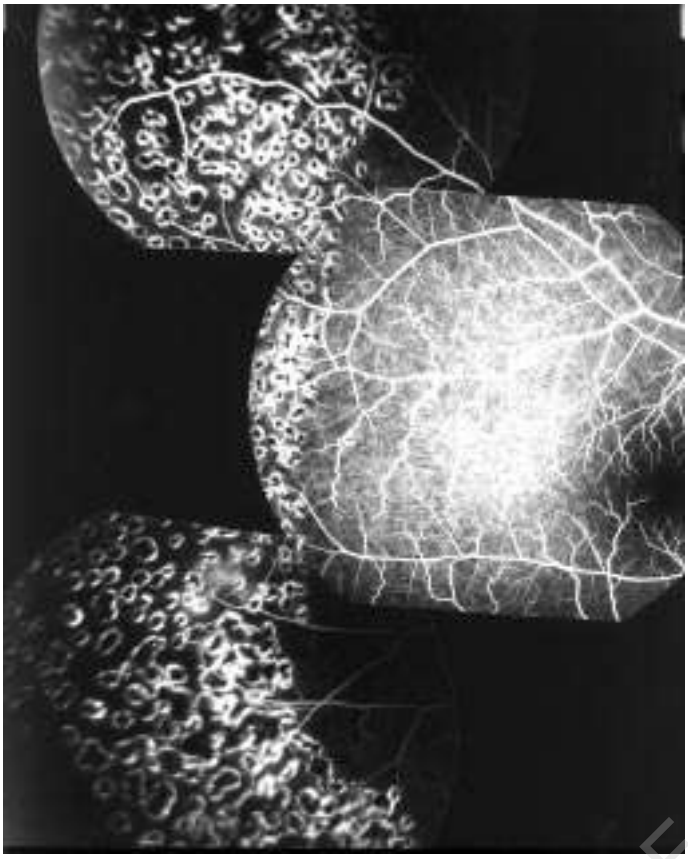

**Figure 2. Fluorescein angiogram of the same image shows the outcome of the photocoagulative treatment; the vascular neoformations have disappeared and the retina is no longer ischemic.**



## References


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