Sudden proptosis of the left eye in a Saudi male

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A 15-year-old male was admitted with sudden left eye swelling accompanied by mild left eye pain and left side headache, no history of trauma, no decrease in vision and no change in the level of consciousness. Examination revealed a young male with no pallor or jaundice and with stable vital signs. General examination was unremarkable. Eye examination revealed left eye proptosis downward and outward, mild keratopathy, no visual defect, normal intraocular pressure by digital palpation, a clear lens, no papilledema, and a partially dilated and reactive pupil. There was limitation of left eye movement in all directions.

• What are the abnormal findings on the orbital CT scan (Figure 2)?
• What abnormalities can be seen in the peripheral blood smear (Figure 3)?
• What’s your diagnosis?

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Diagnosis: Orbital Compression Syndrome in Sickle Cell Disease

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Figure 1 shows the left eye proptosis. Figure 2 is a CT scan showing soft tissue opacity in the left orbit, with a density of 86 Hounsfield units, which is close to fresh blood density, and displacement of the left globe downward. Figure 3 is a peripheral blood film showing sickle cells, anisocytosis, and evidence of hyposplenism (target cells, Howell-Jolly bodies).

Discussion
Sickle cell disease is a genetic disorder of hemoglobin production characterized clinically by anemia, recurrent painful crisis, splenic and hepatic sequestration, acute chest syndrome, stroke and bacterial sepsis. Orbital involvement in sickle cell disease is rare.1,2,3 Only a few cases are reported from Saudi Arabia.4

The orbit is a concave-shaped space comprised of seven bones (frontal, greater and lesser wings of the sphenoid, zygoma, maxilla, lacrimal, palatine and ethmoid).5 Orbital swelling in sickle cell disease occurs with orbital bone infarction, cellulitis, orbital abscesses, and orbital compression syndrome. Orbital compression syndrome is characterized by frontal headache, eyelid edema, proptosis, and fever.3,6 The clinical presentation in our patient is compatible with the diagnosis of orbital compression syndrome. This syndrome is believed to be due to hematoma adjacent to the orbital bones and appears to result from bone marrow infarction. Bilateral involvement occurs in over half of the cases.6 Orbital compression syndrome ranges from mild, which can be resolved with conservative treatment to sight-threatening, which needs surgical exploration and evacuation of the hematoma to prevent loss of vision.7

References