A sickle cell disease (SCD) patient with vaso-occlusive crisis (VOC) developed sudden sensorineural hearing loss (SNHL), which responded well to a rapid course of corticosteroids along with exchange transfusions. The otolaryngologist should be aware of the otologic manifestations of SCD such as SNHL. A good response to steroids, which averted a permanent hearing loss, warrants further studies to define the augmentive benefits of such therapy along with exchange transfusions in patients with SCD in VOC who develop sudden SNHL.

Sickle cell disease (SCD) is a hemolytic anemia quite prevalent in Oman, with the most recent epidemiological data suggesting a sickle cell gene carrier rate of about 6% and SCD prevalence of 0.2%. The disease is characterized by a change in the shape of red cells to the classical sickled shape leading to a process of vessel occlusion involving the small and medium-sized vessels in almost all parts of the body. The process of vaso-occlusion is made worse by episodes of infections, dehydration or deoxygenation.

It is also known that patients with SCD are at an increased risk of thrombosis and this process is considered to contribute to several clinical features of the disease including leg ulcers, priapism, avascular necrosis of bone, pulmonary hypertension, acute chest syndrome, stroke, and apparently any random infarctive lesions that can occur anywhere in the body. The cochlea, the organ of hearing, is not immune from this process and audiometric assessment is a sensitive indicator of any sensorineural (perceptive) hearing loss resulting from any occlusive vascular involvement.

The prevalence of involvement of the ears in SCD patients is variable but it is reported to occur between 11% to 41% in black patients with this disease and can lead to sensorineural hearing loss (SNHL). The pattern of involvement is also variable and it can be either unilateral, bilateral, temporary or permanent. We report on a young female with a complicated history of SCD who presented with a sudden onset of sensorineural hearing loss (SNHL) that completely resolved following antecedent medical therapy. We wish to highlight here the need for appropriate and rapid audometric evaluation and follow-up treatment so that a reversible and treatable cause for rapid hearing loss in patients with SCD with VOC such as ours can be quickly achieved.

CASE
A 26-year-old Omani female with SCD (Hb S/S genotype) was admitted in January 2010 with acute painful crisis involving the chest, legs and back. Her medical history was significant and included a stroke in childhood with complete neurological recovery, protein C deficiency, acute chest syndrome, and iron overload due to repeated blood transfusions/exchange. She was taking hydroxyurea, desferoxamine, penicillin V, folic acid, co-codamol, aspirin and enoxaparin. On admission, she had stable vital signs with pallor and icterus. She was treated with hydration, pain management with morphine infusion and non-steroidal anti-inflammatory drugs. Two days after admission, the patient awoke with acute complete hearing loss in the left ear. There was no history of imbalance or vertigo and she was otherwise well, apart from her ongoing painful crisis. Her hearing had previously been considered normal.
Furthermore, clinical examination found no neurological deficit. Investigations showed hemoglobin 7.5 g/dL, white blood cells 12.1×10⁹/L, platelets 344×10⁹/L, reticulocytes 538×10⁹/L (25%), hemoglobin S 70.8%, hemoglobin F 1.0%, hemoglobin A 24.6% (transfused blood), hemoglobin A₂ 3.4%, lactate dehydrogenase 640 μ/L (normal range, 135-214), haptoglobin <0.06 g/L, total bilirubin 88 μmol/L, with otherwise normal renal and liver functions.

She was evaluated in the ENT department, and audiometry, middle ear compliance, acoustic reflex, and tone decay were ordered. Audiometry studies showed a profound hearing loss in the left ear and mild hearing loss in the right (Figure 1 top). Radiological evaluation showed a normal chest x-ray, ultrasound of the abdomen showed gallstones, magnetic resonance imaging with angiogram of the brain did not reveal any new neurological lesions except for a large frontal cystic encephalomalacia due to an old infarction. She had extensive investigations to look for an underlying pathology, including autoimmune work such as antinuclear antibodies, autoantibody screen and antineutrophilic antibodies and they were all negative. Polymerase chain reaction (PCR) for CMV, hepatitis B, C and HIV were all negative, and rapid plasma reagin as well as Treponema pallidum hemagglutination were also negative. She was treated with exchange transfusions and started on oral steroids (prednisone 60 mg daily for a week), which was gradually reduced over two weeks. Additionally she received beta histidine 16 mg three times daily, as well as exchange transfusions. Audiometry was done every three days and the patient discharged with normal hearing in both ears in 2 weeks (Figure 1 bottom).

DISCUSSION

Normal erythrocytes are compressible, disc-shaped cells that flow easily through the smallest blood vessels, navigating blood vessels that are 1/3 of their diameter; these cells live about 120 days. In stark contrast, in SCD patients the abnormally sickle-shaped red cells are rigid, and get stuck in small blood vessels, leading to the most frequent manifestations of this disease, namely vasocclusion. Moreover these red cells live for only 20 or fewer days, perpetuating the vicious cycle of increased red cell production followed by increased red cell destruction. The sickle cells interrupt blood flow by blocking small blood vessels especially in the capillary-venous bed, where the partial pressure of oxygen is reduced, promoting and accelerating the sickle shape change. This morphological change, and its associated physiological change, drastically reduces the ability of red blood cell to navigate and deliver oxygen throughout the body.

SNHL is a multifactorial process with idiopathic sudden SNHL being the most common diagnosis. The cochlea is highly sensitive to disruption of blood flow, and occlusion of labyrinthine vessels were shown in an animal model by Perlman to induce irreversible hearing loss. Ischemia of the stria vascularis, increased blood viscosity by crystallization of red blood cells, with development of stasis, ischemia or hypoxia appears to be the outstanding pathophysiological phenomenon during crisis. Given its size, this tiny organ receives a disproportionate percentage of the body’s blood and the extensive cochlear vascular anatomy, the high metabolic activity required to maintain both ionic and electrical characteristics of endolymph, and the highly dependent nature of evoked responses from the inner ear on brief episodes of anoxia, suggest that a secure and extensive cochlea blood flow is critical.

Our patient presented with crisis due to SCD and

Figure 1. Pure tone audiogram before (top) and after (bottom) treatment.
developed sudden hearing loss in all the frequencies of more than 30 dB of the left ear, strengthening the evidence that this process was related to SCD and she was treated with rapid exchange transfusions and steroids. Interestingly, she presented with a number of confounding factors. She had a history of stroke, and although she had a normal hearing following the stroke, with no neurological deficit, it is conceivable that she was at a high risk of developing a vascular CNS event. Furthermore, she was intermittently on desferoxamine, an agent that is linked with auditory complications, although usually this is only thought to occur at a lower ferritin level, and her current ferritin level was 2930 ng/mL (reference range, 20-300 ng/mL). This patient also had a complicated clinical course and presented with painful crisis, had a low hemoglobin level, with a very high reticulocyte count (known for their adhesive properties) and high hemoglobin S level, all features supportive of aggravating a vaso-occlusive process due to SCD. However, we had simultaneously investigated her extensively to exclude other causes of sudden SNHL such as infection with negative viral studies, or no serological evidence of syphilis, and no evidence of autoimmune disease as documented by negative autoimmune markers. Furthermore this patient responded extremely well to steroids with rapid resolution of her symptoms and the hence the rare possibility of an autoimmune process cannot be completely ruled out. A rapid short course of steroid can be recommended in such cases, unless there is an absolute contraindication to their use, as the consequences of permanent hearing loss are profound.

REFERENCES