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Clinical Records

Sudden total deafness in sickle cell disease

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Abstract
Sickle cell disease is a world-wide problem which has been noted to cause high tone sensorineural hearing loss. We report a case with sudden onset bilateral hearing loss which progressed to total sensorineural deafness. To our knowledge there has been no report of such a case occurring previously.

Introduction

Sickle cell disease is a haemolytic anaemia which is most prevalent in tropical and subtropical regions but which is encountered world-wide due to population migration. The disorder is inherited along mendelian lines and may be associated with other haemoglobinopathies, most commonly with the thalassaemia trait. Symptoms of anaemia are usually mild because haemoglobin S [HbS] readily releases oxygen to the tissues but the disease is punctuated by painful crises induced by episodes of infection, dehydration or deoxygenation. At such times, sickle cells obstruct small vessels by sludging and thus cause ischaemia and tissue necrosis.

A review of the literature reveals an association between sickle cell disease and deafness but we have not found any record of it causing sudden, irreversible, total bilateral hearing loss. We report such a case occurring in a patient with sickle cell thalassaemia.

Case report

A 25-year-old West Indian female with sickle cell thalassaemia developed bilateral otalgia whilst on a holiday flight. This pain began while the aircraft was cruising at high altitude and not during ascent or descent. We were unable to obtain a history of barotrauma. The otalgia persisted for several days but was not sufficiently severe to cause the patient to seek medical attention during her holiday. On the morning following flight, the patient awoke with bilateral partial deafness, worse on the left

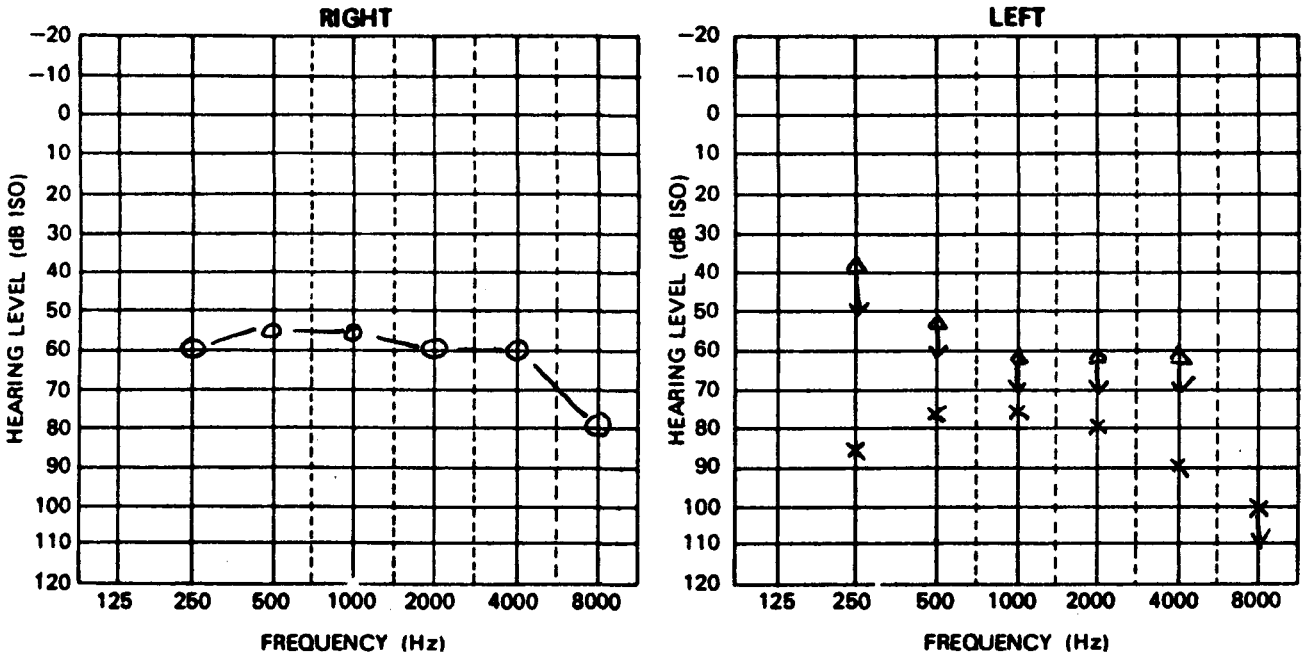

Fig. 1
Patient's audiogram when first seen on 20th November 1989.

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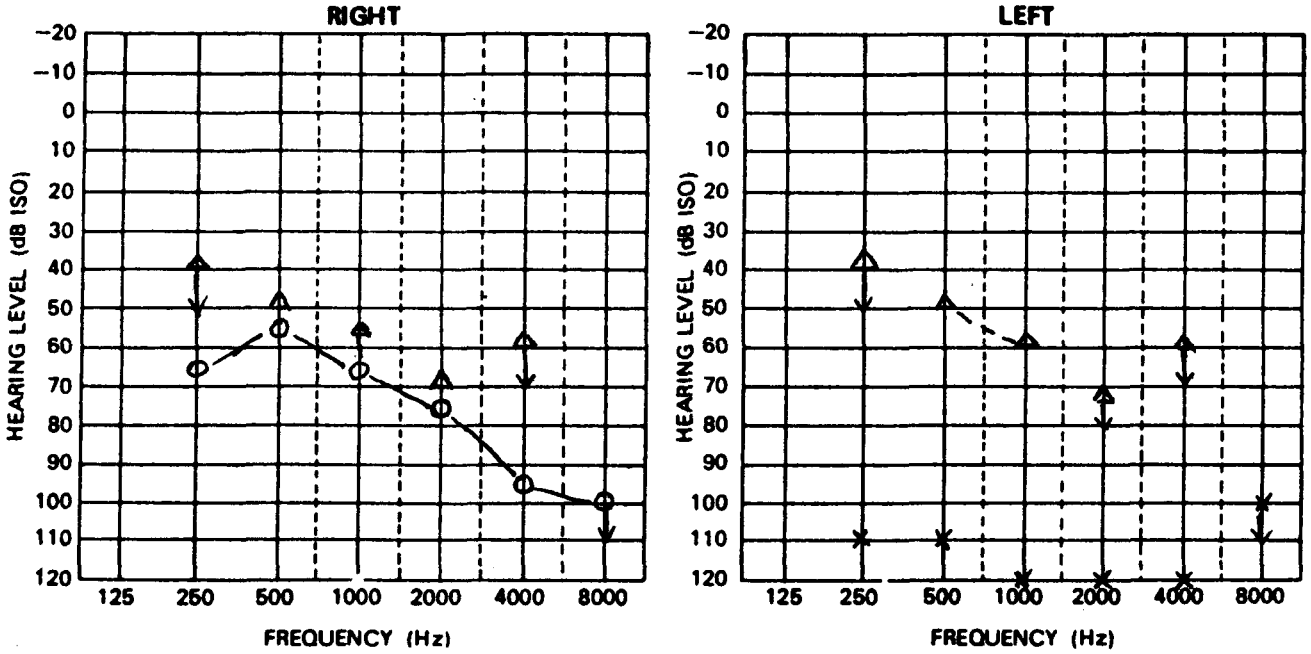

FIG. 2
Patient's audiogram on 4th December 1989 showing progression of sensorineural deafness.

side. There was no history of any imbalance or vertigo and she was otherwise well. Her hearing had previously been thought to be normal.

This patient had suffered two sickle cell crises in the ten years preceding this event but none in the immediate past. There was no family history of deafness and she was not taking any medication.

She was referred for an ENT opinion four weeks after her holiday by which time she had a marked sensorineural hearing loss with mean air conduction thresholds of 75 dB on the left side and 55 dB on the right, across the frequency range from 250 Hz to 4000 Hz (Fig. 1). Acoustic reflexes were normal and serology tests were unhelpful. The haemoglobin level was 7.9 g/dl, a drop of approximately 2 g/dl from the patient's normal level. Within two weeks there was almost no recordable hearing in the left ear except for two peaks at 110 dB [250 Hz and 500 Hz], with a falling curve on the right (Fig. 2). Three weeks later there was no hearing whatsover in either ear (Fig. 3). This patient remains completely deaf more than a year after her problem began.

Discussion

We believe that this patient's deafness was caused by sickle cell changes within the microcirculation of her inner ears which occurred despite flying in a modern pressurized jet aircraft. A possible differential diagnosis is simultaneous bilateral perilymph leaks but we have never seen any such cases reported. Perilymph leaks usually cause tinnitus, vertigo, imbalance and a hearing loss which tends to be fluctuant. The patient reported

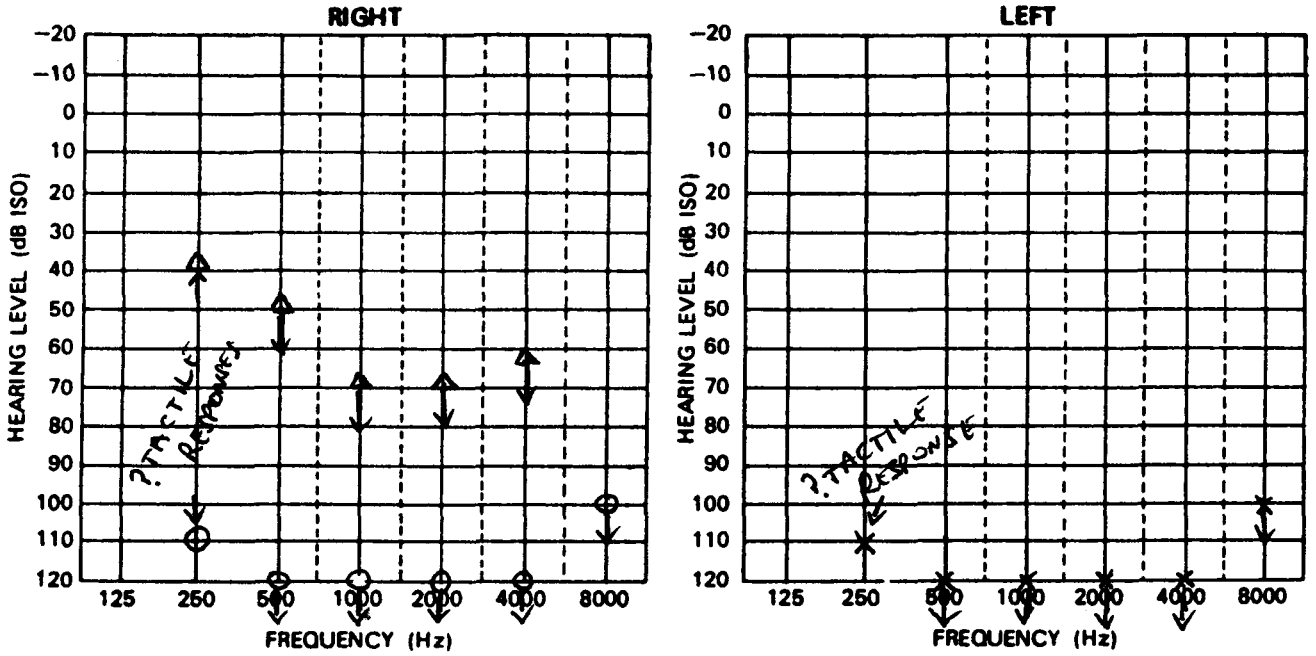

FIG. 3
Patient's audiogram of 15th January 1990 showing total bilateral sensorineural deafness. Subsequent audiograms have shown no change.

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here complained of otalgia followed by progressive deafness and was unaware of any barotrauma. Similarly, there was no evidence to support a lesion of the central neurological auditory pathways. This patient has successfully passed assessment for cochlear implantation which adds support to our theory that the haemoglobinopathy affected the inner ear alone. Previously reported cases of profound hearing loss in sickle cell disease have been part of major crises affecting many other organs and have also made at least a partial recovery (Urban, 1973, Orchik and Dunn, 1977).

Marcus *et al.*, (1976) reported a family of four sisters, two of whom had sickle cell thalassaemia. These two girls complained of vertigo in isolation or vertigo with hearing loss after strenuous exercise. One of these girls woke with sudden partial unilateral sensorineural deafness but her eventual outcome is unknown because she did not attend for follow-up.

Reported patterns of hearing loss in sickle cell disease vary. Sudden loss as described above probably represents infarctive episodes. Larger series (Todd *et al.* 1973; Friedman, *et al.*, 1980) report a 12 per cent to 22 per cent incidence of high tone sensorineural deafness in sickle cell patients when compared to controls. Todd *et al.*, (1973) have suggested that this type of hearing loss is due to a continuous low grade venous thrombotic process affecting the cochlear, without clinically recognized episodes. Berry (1975) found that sickle cell patients who experienced more frequent crises showed poorer threshold responses. This finding was not supported by the study of Friedman *et al.* (1980) of 43 homozygous sickle cell patients. It has been suggested that marrow space expansion in the petrous temporal bone could cause compression of the eighth cranial nerve in the auditory canal with ensuing deafness. Serjeant *et al.* (1975) examined this theory and found no correlation between abnormal audiograms and internal auditory meatal dimensions in patients with sickle cell disease.

Morgenstein and Manace (1969) provided evidence that hypoxia may be responsible for deafness in these patients when they examined the temporal bones of a 10-year-old boy who died during a crisis. He was known to have had a moderate bilateral sensorineural hearing loss during life. The pathological findings included absent and abnormal hair cells with sickle cell clumping throughout the venous and capillary vessels of the temporal bones. There were ischaemic changes evident in the organ of Corti and signs of hypoxia in the strial vessels.

It is evident that sickle cell diseae may affect hearing, sometimes with disastrous results. Given the pathological findings reported by Morgen and Manace (1969), it is surprising that hearing is not affected more frequently than has been reported, particularly during sickle cell crises.

Key words: Deafness, sudden; Sickle cell disease

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