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Sudden sensorineural hearing loss in haemoglobin SC disease

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Abstract
Presented herein is a case report of sudden sensorineural hearing loss in the setting of haemoglobin SC disease. The relationship of the two is rare; the authors have found that this is only the second report in the literature. In this instance, partial exchange transfusions were performed in an attempt to decrease viscosity and improve blood flow. Thereafter, hearing stabilized and then slowly improved. The evidence for the beneficial role of these transfusions in this setting is, at best, circumstantial, but it is theoretically sound and worthy of further study.

Key words: Hearing loss, sensorineural, sudden; Haemoglobin SC disease

Introduction

Haemoglobin SC disease is a heterozygous state in which individuals inherit both the haemoglobin S and haemoglobin C genes. The homozygous condition SS produces sickle cell anaemia. The abnormal haemoglobins S and C result from different substitutions at the sixth position of the beta chain of the haemoglobin molecule Table I. Patients with haemoglobin SC disease may manifest the same symptoms as patients with sickle cell anaemia, although these symptoms occur less frequently in SC disease. The pathophysiology of sickle cell disease is primarily that of occlusions of the microvasculature by sickled red blood cells.

Sickling represents the external manifestation of a three-stage intracellular process involving abnormal haemoglobin S molecules (Babior and Stossel, 1984). In a deoxygenated environment, the haemoglobin S molecules randomly aggregate. The substitution of valine at the sixth position facilitates the binding of neighbouring haemoglobin S molecules. The valine interacts with the phenylalanine and leucine amino acids located at the 85 and 88 positions of the alpha chains of adjacent haemoglobin molecules. When 15 haemoglobin molecules have accumulated, polymerization begins. The deoxygenated haemoglobin S molecules precipitate rapidly to form thousands of 14-stranded fibres. These fibres are composed of five couplets surrounding a pair of couplets. The fibres align into long bundles causing the cell to deform and assume its sickled shape. The sickled cells are unable to change their shape to squeeze through arterioles and capillaries, resulting in micro-occlusions and further deoxygenation. The lesser severity of haemoglobin SC disease is a result of the weaker binding of haemoglobin C molecules into the polymers of haemoglobin S. The SC cells are, therefore, less likely to sickle.

Sudden sensorineural hearing loss has only been described in sickle cell anaemia. Transient vertigo has been described in patients with haemoglobin SC disease, however these patients had normal audiograms (Serjeant *et al.*, 1973). The following case describes a woman with haemoglobin SC disease who developed sudden unilateral sensorineural hearing loss that slowly improved following partial exchange transfusion. This is only the second report in the literature of sudden sensorineural hearing loss in haemoglobin SC disease (Morrison and Booth, 1970).

Case report

A 43-year-old woman with haemoglobin SC disease presented in March 1989 complaining of the sudden onset of decreased hearing in her right ear that had occurred two weeks earlier. She reported occasional tinnitus in the right ear and unsteadiness, but no true vertigo. She denied any antecedent upper respiratory infections. She complained of back pain, which was characteristic of her painful crises. She had no prior otological history. Her past medical history was significant for painful crises every five to six months, aseptic necrosis of the femoral heads, hypersplenic crisis, bilateral lower leg thrombophlebitis, and left subclavian vein thrombosis after attempts at venous access. Her head and neck examination was normal, without nystagmus or ataxia.

An audiogram showed normal hearing in her left ear (except at 8000 Hz) and a mild to severe sloping sensorineural hearing loss in her right ear (Figure 1). Speech discrimination was 100 per cent in the left ear and 48 per cent in the right. Ten days later, hearing in the right ear had diminished further by an average 15 decibels at each frequency, with speech discrimination reaching zero per cent (Figure 2). An electronystagmogram was normal. A fistula test was negative. A magnetic resonance image with gadolinium enhancement of the temporal bones and posterior

TABLE I

Haemoglobin A	Val	His	Leu	Thr	Pro	Glu	Glu
Haemoglobin S	Val	His	Leu	Thr	Pro	Val	Glu
Haemoglobin C	Val	His	Leu	Thr	Pro	Lys	Glu

Initial sequence of the beta chain of the haemoglobin molecule. (Val = valine; His = histidine; Leu = leucine; Thr = threonine; Pro = proline, Glu = glutamic acid; Lys = lysine).

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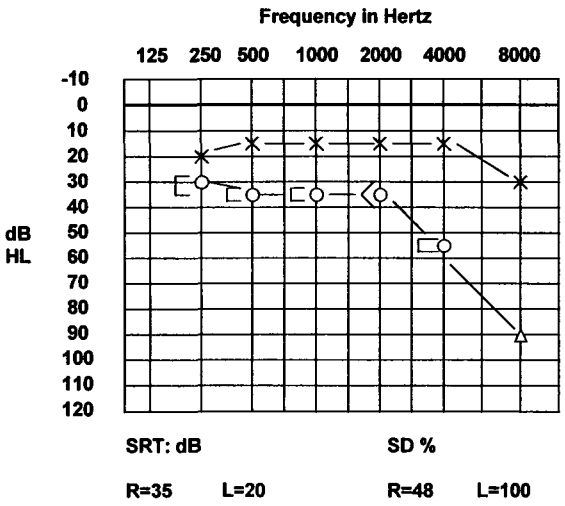

FIG. 1

Initial audiogram. (O = Air conduction right ear; X = air conduction left ear).

fossa was normal. The patient was hospitalized and a lumbar puncture performed. Cerebrospinal fluid culture and VDRL were negative. She received two exchange transfusions of two units each. Post-transfusion haemoglobin electrophoresis showed a haemoglobin S level of 18 per cent and haemoglobin C level of 22 per cent.

The hearing in the right ear did not immediately normalize, but rather stabilized and then improved slowly. An audiogram performed six weeks after partial exchange transfusion (18 May 1989) demonstrated slight improvement in the hearing thresholds in the lower frequencies and speech thresholds (speech threshold 35 dB) but no improvement in the higher frequencies, and a speech discrimination of 40 per cent. Clinically she had decreased tinnitus and was no longer dizzy. An audiogram performed one month later (19 June 1989) revealed some improvement in the lower frequencies and speech thresholds (speech threshold 30 dB), no improvement in the high frequencies, but speech discrimination of 76 per cent. She was clinically not dizzy at this time. An audiogram performed on 29 August 1989, five months post partial exchange transfusion, revealed continued improvement in the low frequencies and speech frequencies (speech threshold 20 dB) no change in the higher frequencies, speech discrimination 72 per cent. At that time she was still not dizzy but noted clicking in the ear. Two subsequent audiograms, one performed at seven months and one performed at one year post partial exchange transfusion revealed the hearing to have stabilized with speech thresholds of 25 and 20 dB respectively, but with speech discriminations of 92 and 96 per cent (Figure 3). She was no longer dizzy, although at her last examination she was hospitalized for a sickle-related crisis and was nauseated with a headache.

An auditory brain stem response performed seven months after initial presentation showed normal neural conduction.

The patient subsequently became HIV positive and died of complications of that disease. Presumably she developed the HIV infection from HIV-infected blood either prior to the initiation of HIV blood screening or from a donor with such recent acquisition of the virus that antibodies were not identifiable in the blood screening.

Discussion

Sensorineural hearing loss (SNHL) was first noted in sickle cell anaemia by Morgenstein and Manace (1969). They examined the temporal bone from a 10-year-old boy who previously had been found to have a mild bilateral SNHL. Many inner and outer hair cells were absent or abnormal; the stria vascularis was disrupted. In 1970, Morrison and Booth mentioned a patient with haemoglobin SC disease among their 94 patients with sudden SNHL. No other information was presented regarding the hearing loss other than that it was bilateral.

Todd *et al.* (1973) studied 83 Jamaicans with sickle cell anaemia and found a 22 per cent prevalence of SNHL *versus* four per cent in the control population. Subsequently, three patients with sickle cell anaemia who experienced a sudden SNHL were described (Urban, 1973; Orchik and Dunn, 1977; Hotaling *et al.*, 1989). All three patients experienced some improvement in their hearing with the resolution of their crises. The patient described by Orchik and Dunn did not experience any improvement in the ear with the profound loss.

The rationale for partial exchange transfusion is found in the work of Anderson *et al.* (1963). They noted that, when normal red blood cells constitute at least 30 per cent of the total red blood cell volume, the viscosity of sickle cell blood under conditions of maximum deoxygenation is decreased, thereby improving blood flow. Compared to standard transfusion, partial exchange transfusion decreases the risk of iron overload, prevents fluid overload, and minimizes the amount of blood needed to achieve the desired percentage of normal cells. Partial exchange transfusion has been

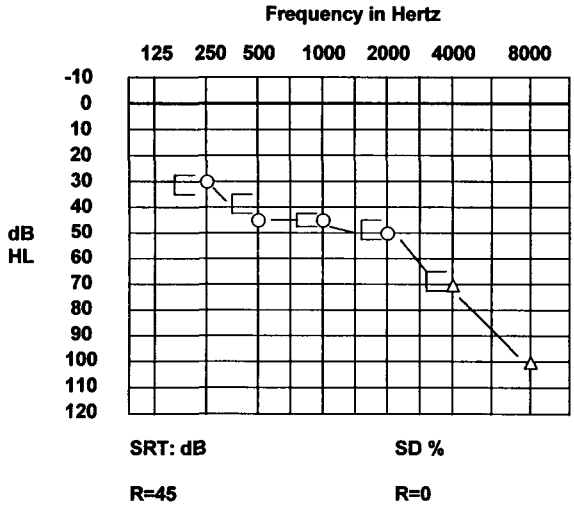

FIG. 2

Right ear ten days later than initial audiogram.

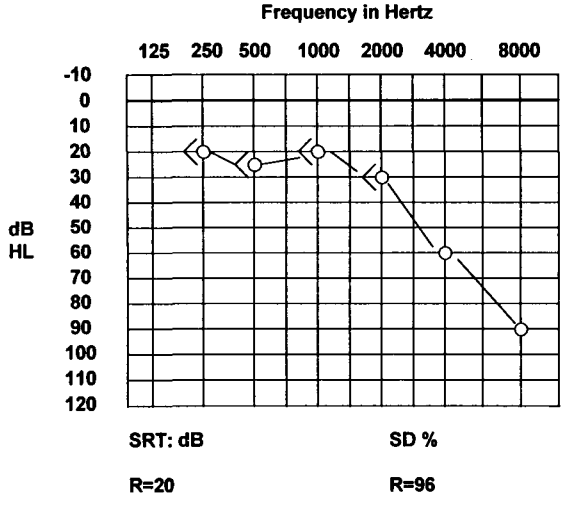

FIG. 3

One year follow up of right ear.

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used successfully to treat painful crises, pulmonary infarction secondary to marrow embolism during pregnancy, and macular infarction (Brody *et al.*, 1970; Davey *et al.*, 1978; Khwarg *et al.*, 1985). Fluorescein angiography performed in a patient treated with partial exchange transfusion documented restoration of blood flow in a previously occluded macular vessel.

A role for increased blood viscosity in the pathophysiology of SNHL was demonstrated by Davis and Nilo (1965) in a patient with polycythaemia vera who was treated with periodic phlebotomy. When the patient in our report did not respond to standard treatment for painful crisis, it was decided to perform a partial-exchange transfusion. Haemoglobin S and haemoglobin C levels were reduced to 40 per cent of blood volume. The patient's pure tone hearing and speech discrimination began a slow improvement with the lowering of the blood viscosity.

We cannot prove that the partial exchange transfusion was responsible for the improvement in hearing, or even that the sudden sensorineural hearing loss is definitively sickle-crisis related; however there is a temporal relationship. It is also possible that the patient suffered from a perilymph fistula accounting for her hearing loss. However, she did undergo electronystagmography with fistula testing, which was negative for fistula. It is conceivable that the hearing loss could have been due to an endolymphatic-type hydrops. The clinical picture does not correlate. In our opinion the most likely etiology of this sudden sensorineural hearing loss is sludging related to the sickle cell disease.

Conclusion

The second case of sudden sensorineural hearing loss in a patient with haemoglobin SC disease is presented. The patient's hearing improved after a partial exchange transfusion. Although the evidence for a role of the transfusion in the hearing improvement is circumstantial, there is a sound theoretical and empirical basis for the use of partial exchange transfusions in treating the complications of sickle cell disease. Further studies are needed to determine if partial exchange transfusions may play a role in the treatment of SNHL in sickle cell disease.

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