

LETTER TO THE EDITOR

Plasma exchange in a child with systemic lupus erythematosus antiphospholipid antibodies and profound deafness

We have previously reported sudden onset of profound deafness associated with antiphospholipid antibodies in a 12-year-old child with systemic lupus erythematosus (SLE).¹ On admission there was a 3-month history of deafness, intermittent fever, oral ulcers, a photosensitive malar rash, and a discoid rash on her back and scalp leading to scaly areas of alopecia. Pure tone audiometry showed severe bilateral sensorineural deafness with no response up to 120 decibels. Antinuclear antibody was 1.8 (normal < 1). Anticardiolipin antibody IgM was 8.9 MPL U/ml (normal 0–6.0) and IgG was 13.10 GPL U/ml (normal 0–12). She was started on oral prednisolone 2 mg/kg/day for 10 days, then 1 mg/kg/day for 2 weeks, then 1 mg/kg/ on alternate days for the next month. During the following month the total dose was reduced to 5 mg. The fever and rash responded promptly but the deafness did not improve. A report of the use of plasma exchange in an adult with SLE deafness² prompted us to offer this modality of treatment to this child.

On re-admission 6 months after the onset of her symptoms, pure tone audiometry was repeated and showed that there was no improvement. There was still no response at 120 decibels. In the absence of a plasmapheresis machine, a 1-volume manual plasma exchange was done over 2 days, as follows: 10% of the child's blood volume (200 ml) was withdrawn into a single bag containing citrate-phosphate-dextrose-adenine I (CPDA I) and centrifuged at $1200 \times g$ for 10 minutes in a Beckman J6-MC centrifuge

at 4°C and the supernatant plasma was discarded. Her packed cells were returned to her, infused with the help of a syringe pump. The reduced plasma volume was corrected with fresh frozen plasma which was infused separately after the packed cells.

One litre of her blood was withdrawn in five such cycles. The procedure was repeated after 4 days and by the end of the process it was calculated that her total blood volume had been exchanged. After the first five cycles she could hear sounds and responded to them but she could not discern speech. Following her plasma exchange, her audiogram showed that she could hear with her left ear. Her audiogram showed response to 105 db at 500 Hz and at 95 db at 2K. She was started on aspirin, one tablet daily. Repeat audiogram done a month later showed that the hearing in the right ear was very marginally better with some hearing at 110 db. The left ear showed no deterioration. She was able to understand some words but her hearing was not sufficient for sustained conversation without the help of a hearing aid.

Although neurological features occur in 60% of patients with SLE, deafness is not an established characteristic of the disease.² The mechanism of hearing loss in SLE might be thrombosis related to antiphospholipid antibody,³ which Carreras and Vermeylen suggest might cause endothelial damage which results in decreased prostacyclin production and leads to platelet aggregation and thrombosis.⁴ The antiphospholipid antibodies were slightly but not dramatically

elevated in our patient. Plasma exchange is thought to work in SLE by clearing immune complexes. Plasma exchange for SLE was introduced in 1974 but its use is not yet completely established. Hamilton described the use of plasma exchange to combat deafness in a patient with SLE.⁵ Complete resolution was noted but recurrence requiring repeat exchanges every 3 to 6 months was noted.²

Sentaro Kobayashi *et al.* also reported the use of plasmapheresis in SLE for deafness of gradual onset that had evolved to a 110 db loss over a 4-year period.⁶ After two series of plasmapheresis, hearing had improved by 40–60 db. This improvement was less dramatic than the complete resolution of deafness noted in Hamilton's patient who was plasma-exchanged 6 weeks after the onset of deafness. Our patient was plasma-exchanged 6 months after the onset of deafness and had an improvement of 10–20 db in one ear. It seems reasonable to conjecture that plasma exchange done early in the disease is associated with better recovery.

The manual technique used for plasma exchange in this case was not associated with any complications and could be used in centres without plasmapheresis machines.

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