The Journal of Laryngology & Otology (2008), ***122***, *204–206*.

doi:10.1017/S0022215107007736
Printed in the United Kingdom
First published online 10 April 2007

Clinical Record

Sudden sensorineural hearing loss in a patient with primary antiphospholipid syndrome

K-T Kang, Y-H Young

Abstract

Objective: Despite multiple systemic manifestations, sudden sensorineural hearing loss in a patient with antiphospholipid syndrome is rarely reported.

Patient: A 46-year-old man with primary antiphospholipid syndrome had a sudden onset of hearing loss and tinnitus in the right ear in December 2005, because he discontinued use of warfarin and acetylsalicylic acid for a few days.

Results: Audiometry revealed saucer-type sensorineural hearing loss with a pure tone average of 73 dB in the right ear, and flat-type hearing loss with a pure tone average of 25 dB in the left ear. Electronystagmography displayed multiple central signs and bilateral canal paresis, while a vestibular evoked myogenic potential test revealed bilateral delayed responses. After admission, the patient was re-treated with warfarin and acetylsalicylic acid. Follow-up audiometry showed recovery of right-sided hearing, with a pure tone average of 12 dB, three days after presentation.

Conclusion: Consensus exists on the effectiveness of anticoagulant agents in aiding a favourable outcome of sudden sensorineural hearing loss in patients with antiphospholipid syndrome.

Key words: Antiphospholipid Syndrome; Sensorineural Deafness; Warfarin

Introduction

Autoimmune disorders correlated with sudden sensorineural hearing loss have been widely investigated.[1] Of these, antiphospholipid syndrome is a systemic autoimmune disorder characterised by arterial and/or venous thrombi and recurrent pregnancy loss, accompanied by persistently positive anticardiolipin antibodies or lupus anticoagulant tests.[2] The pre-eminent clinical feature is recurrent episodes of arterial, venous and small vessel thrombi, particularly of renal and coeliac origins.[3] A variety of effects in multiple organ systems have been described; for example, ophthalmological manifestations include amaurosis fugax, ischaemic optic neuropathy, visual field loss and diplopia,[4] while cerebrovascular events include ischaemic stroke and transient ischaemic attack.[5] Despite multiple systemic manifestations,[6] sudden sensorineural hearing loss in a patient with antiphospholipid syndrome is rarely reported.[7,8]

Recently, we managed a 46-year-old man with primary antiphospholipid syndrome who had suffered recurrent episodes of vascular thrombosis, followed by sudden onset sensorineural hearing loss. Herein, we present this case.

Case report

In 1994, a 35-year-old man suffered fever and lower abdominal pain, which brought him to his local hospital. Acute peritonitis was suspected, and an emergency laparotomy revealed ischaemic change with impending rupture of the small intestine. Segmental resection of the bowel and appendectomy were performed.

However, in 1995, the patient suffered left foot pain with gangrenous change, and he was referred to our hospital. Angiography demonstrated narrowing of the left posterior tibial artery; a below knee amputation was thus performed. Laboratory tests showed a positive lupus anticoagulant test, and antiphospholipid syndrome was diagnosed. During this hospitalisation, audiometry was performed due to the patient's tinnitus; this showed normal hearing with a pure tone average of 5 dB in the right ear, and a high tone, sloping-type hearing loss in the left ear with a pure tone average of 12 dB (Figure 1).

Subsequently, the patient was regularly treated with warfarin sodium 5 mg and acetylsalicylic acid 100 mg, daily.

Unfortunately, on 1 December 2005 the patient suffered a sudden onset of tinnitus and hearing impairment in the right ear and was admitted to our ward. He denied recent upper airway infection, head trauma or noise exposure, but had discontinued use of warfarin and acetylsalicylic acid for a few days.

Audiometry showed a saucer-type sensorineural hearing loss with a pure tone average of 73 dB in the right ear, and a flat-type hearing loss with a pure tone average of 27 dB in the left ear (Figure 1). Electronystagmographic examination revealed abnormal eye tracking and optokinetic nystagmus tests. Caloric testing indicated canal paresis in both ears. Vestibular evoked myogenic potential testing elicited

From the Department of Otolaryngology, National Taiwan University Hospital and National Taiwan University College of Medicine, Taipei, Taiwan.
Accepted for publication: 10 January 2007.

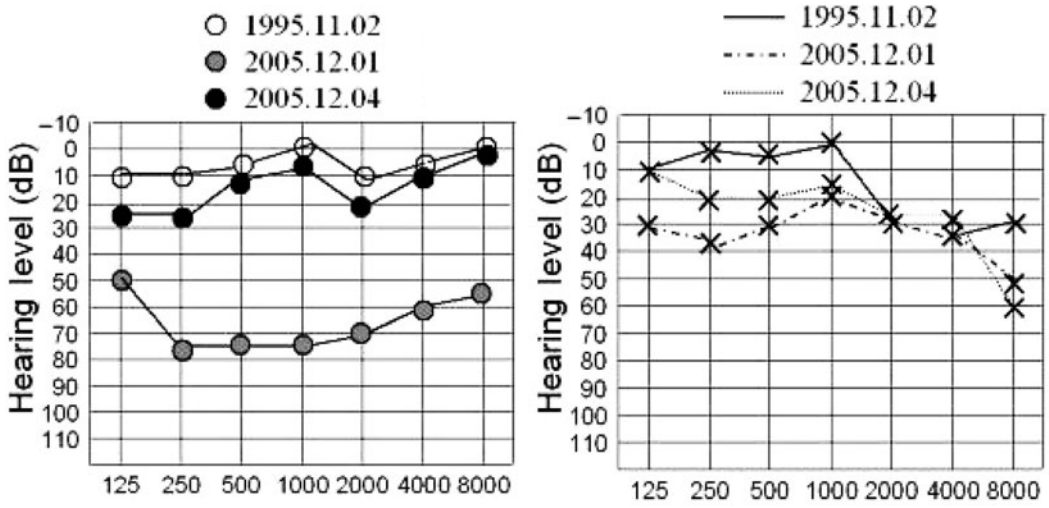

FIG. 1

Chronological hearing changes in a patient with primary antiphospholipid syndrome. Audiogram performed on 1 December 2005 shows saucer-type sensorineural hearing loss in the right ear and flat-type hearing loss in the left ear. Recovery of right ear hearing was observed on 4 December 2005. o = right ear; x = left ear

bilateral delayed responses, with the latencies of p13 as 19.6 and 18.2 ms (normal value <17.1 ms) on the right and left ears, respectively. However, magnetic resonance imaging (MRI) and magnetic resonance angiography scanning demonstrated no particular cranial fossa abnormalities.

Laboratory tests were all within normal limits, including complete blood cell counts, biochemistry profile, plasma protamine paracoagulation test, fibrin degradation products, D-dimer and C-reactive protein. An antiphospholipid profile was also within normal limits, including antiphospholipid immunoglobulin (IgG) (3.193 GPLU (1gG phospholipid units)/ml), anticardiolipin IgG (1.358 MPLU (1gM phospholipid units)/ml) and IgM (1.304 MPLU (1gM phospholipid units)/ml). Antinuclear antibody was 1:320 (+) homogeneous. However, a coagulation profile showed prolongation of the prothrombin time (21.7 s; normal value 9.5–13.0 s) and the partial thromboplastin time (36.4 s; normal value 26.9–36.3 s).

After admission, the patient was re-treated with warfarin and acetylsalicylic acid, as before. Three days after presentation, follow-up audiometry revealed recovery of right-sided hearing, with a pure tone average of 12 dB, while the left ear had a pure tone average of 20 dB (Figure 1). Six months later, the patient's hearing remained unchanged.

Discussion

Antiphospholipid syndrome is a common cause of venous and arterial thrombi. A wide variety of neurological disorders (e.g. stroke and mononeuritis) have been reported in antiphospholipid patients, with the pathogenesis related to the formation of microthrombi.[9] In our patient, a saucer-type audiometry pattern was observed, which is often present with acoustic neuroma; proposed mechanisms include compression of the cochlear nerve, nerve conduction block, vascular compression, or toxic or metabolic effects of the tumour.[10] However, MRI scanning failed to demonstrate any space-occupying lesion in the cranial fossa. Furthermore, multiple central signs on electronystagmography and bilateral delayed vestibular evoked myogenic potential responses indicated possible retrolabyrinthine pathology, with a brainstem lesion especially likely.[11]

Patients with antiphospholipid syndrome with or without other autoimmune disease are classified as having secondary or primary antiphospholipid syndrome, respectively.[12] Although sudden sensorineural hearing loss has been reported in an antiphospholipid patient with systemic lupus erythematosus (SLE),[13] sudden sensorineural hearing loss in a primary antiphospholipid syndrome patient has rarely been mentioned.

The diagnosis of antiphospholipid syndrome requires clinical thrombotic events together with positive laboratory testing (i.e. Sapporo criteria).[14] In our patient, two episodes of gangrenous change in the bowel and extremities were consistent with clinical thrombotic features. Positive laboratory tests must be repeated six weeks later to confirm the diagnosis. Recent research also shows an elevated antiphospholipid antibodies level in patients with sensorineural hearing loss, which supports the hypothesis that antiphospholipid antibodies are involved in the pathogenesis of some inner-ear diseases.[15,16] Conversely, the antiphospholipid profile in our patient was not elevated, which may indicate that the sudden hearing loss was due to transient ischaemic attack rather than immune complex deposition.

- **Antiphospholipid syndrome is a systemic autoimmune disorder characterised by vascular thrombi and recurrent pregnancy loss**
- **Despite multiple systemic manifestations, sudden sensorineural hearing loss in a patient with antiphospholipid syndrome has rarely been reported**
- **This paper describes the case of a 46-year-old man with primary antiphospholipid syndrome who suffered sudden onset hearing loss and tinnitus after discontinuing use of warfarin and acetylsalicylic acid for a few days**

The mainstay of treatment in antiphospholipid syndrome is anticoagulation. Patients with recurrent thrombotic events are recommended to use warfarin and low dose

https://doi.org/10.1017/S0022215107007736 Published online by Cambridge University Press

acetylsalicylic acid.[3] In contrast, most SLE patients suffering from sudden sensorineural hearing loss, with positive antiphospholipid antibodies, are treated with steroids. However, spontaneous recovery of sudden sensorineural hearing loss in SLE patients has been reported.[17] Hence, the standard treatment for antiphospholipid patients with sudden deafness remains to be established. In the current case, rapid recovery of hearing could be attributed to the use of anticoagulant agents.

Our understanding of antiphospholipid syndrome has progressed, although the mechanism by which the antiphospholipid antibodies mediate the inner-ear deficit remains to be clarified. Consensus exists on the effectiveness of anticoagulant agents in aiding a favourable outcome of sudden sensorineural hearing loss. Further, similar research on clinical antiphospholipid patients with sudden hearing loss may help us to elucidate the pathogenesis of this syndrome and to formulate a treatment modality.

Acknowledgement

This report was supported by grant number NSC 95-2314-B002-170 from the National Science Council, Taipei, Taiwan.

References

1 Toubi E, Ben-David J, Kessel A, Halas K, Sabo E, Luntz M. Immune-mediated disorders associated with idiopathic sudden sensorineural hearing loss. *Ann Otol Rhinol Laryngol* 2004;**113**:445–9
2 Khamashta MA, Bertolaccini ML, Hughes GRV. Antiphospholipid (Hughes) syndrome. *Autoimmunity* 2004;**37**: 309–12
3 Sammaritano LR. Antiphospholipid syndrome: review. *South Med J* 2005;**98**:617–25
4 Behbehani R, Sergott RC, Savino PJ. The antiphospholipid antibody syndrome: diagnostic aspects. *Curr Opin Ophthalmol* 2004;**15**:483–5
5 Wong M, Sangle S, Jan W, Hughes GR, D'Cruz DP. Intracerebral arterial stenosis with neurological events associated with antiphospholipid syndrome. *Rheumatol* 2005;**44**:948–9
6 Marai I, Zandman-Goddard G, Shoenfeld Y. The systemic nature of the antiphospholipid syndrome. *Scand J Rheumatol* 2004;**33**:365–72
7 Compadretti GC, Brandolini C, Tasca I. Sudden sensorineural hearing loss in lupus erythematosus associated with antiphospholipid syndrome: case report and review. *Ann Otol Rhinol Laryngol* 2005;**114**:214–18
8 Bir LS, Esmeli FO, Ardic FN. Sudden sensorineural hearing loss in a patient with primary antiphospholipid syndrome. *J Thromb Thrombolysis* 2006;**21**:277
9 de Groot PG, Derksen RHMW. The antiphospholipid syndrome: clinical characteristics, laboratory features and pathogenesis. *Curr Opin Infect Dis* 2005;**18**:205–10
10 Noguchi Y, Komatsuzaki A, Nishida H. Electrocochleographic study in patients with vestibular schwannomas and U-shaped audiograms. *Audiology* 2000;**39**:19–23
11 Chen CH, Young Y-H. Vestibular evoked myogenic potentials in brainstem stroke. *Laryngoscope* 2003;**113**:990–3
12 Galli M, Barbui T. Antiphospholipid syndrome: definition and treatment. *Semin Thromb Hemost* 2003;**29**:195–204
13 Green L, Miller EB. Sudden sensorineural hearing loss as a first manifestation of systemic lupus erythematosus: association with anticardiolipin antibodies. *Clin Rheumatol* 2001; **20**:220–2
14 Wilson A, Gharavi AE, Koike T, Lockshin MD, Branch DW, Piette JC *et al*. International consensus statement on preliminary classification criteria for definite antiphospholipid syndrome. *Arthritis Rheum* 1999;**42**:1309–11
15 Mouadeb DA, Ruckenstein MJ. Antiphospholipid inner ear syndrome. *Laryngoscope* 2005;**115**:879–83
16 Bachor E, Kremmer S, Kreuzfelder E, Jahnke K, Seidahmadi S. Antiphospholipid antibodies in patients with sensorineural hearing loss. *Eur Arch Otorhinolaryngol* 2005;**262**:622–6
17 Digiovanni JJ, Nair P. Spontaneous recovery of sudden sensorineural hearing loss: possible association with autoimmune disorder. *J Am Acad Audiol* 2006;**17**:498–505

Address for correspondence:
Dr Yi-Ho Young,
Department of Otolaryngology,
National Taiwan University Hospital,
1 Chang-Te St,
Taipei, Taiwan.

Fax: +886 2 23946674
E-mail: youngyh@ha.mc.ntu.edu.tw

Dr Yi-Ho Young takes responsibility for the integrity of the content of the paper.
Competing interests: None declared