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Main Article

Fluctuating hearing loss with otorrhoea in Hughes syndrome

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Abstract

We present an unusual case of bilateral aural symptoms in a 34-weeks pregnant woman with Hughes syndrome. This report underscores the unpredictable nature of this syndrome and its ENT complications. No previous case of fluctuating hearing loss and otorrhoea in Hughes syndrome has been described.

Key words: Antiphospholipid Antibody Syndrome; Deafness; Middle Ear

Introduction

The antiphospholipid syndrome was described by Hughes and Stovin in 1959, consisting of multiple pulmonary aneurysms and peripheral venous thrombosis.[1] Since then, several cases of Hughes–Stovin syndrome have been published, with bleeding successfully treated by embolisation.

Antiphospholipid antibodies (APLA) regroup a family of antibodies that recognise anionic and neutral phospholipids, which are the components of plasmatic cell membranes. These antibodies expose the patients to the risk of venous and/or arterial thromboembolic incidents (e.g. multiple aneurysms and massive haemoptysis), neurologic events (e.g. cerebrovascular accident) and several types of morbidity during pregnancy (fetal death, premature birth and multiple first trimester spontaneous abortion).[2–5]

Healthy individuals with such antibodies have an increased risk of suffering thrombotic events, particularly if concurrent prothrombotic factors are present.[6,7]

Antiphospholipid antibodies have also been noted in patients with bacterial and viral infections, and after the administration of certain drugs. These are usually immunoglobulin M aCL antibodies, which may occasionally result in thrombotic events.[8–10]

The effects of Hughes syndrome upon the ear have not previously been published.

Current therapy for the condition, in order to prevent recurrent thrombosis, is controversial. It seems that anticoagulant treatment is a better option than anti-aggregants alone.[2,11] Bleeding has been successfully treated by embolisation or aggressive surgical intervention.[3,12]

Patients with antiphospholipid syndrome who develop an initial thrombosis have an increased risk of subsequent thrombotic events in the same territory as the original thrombosis.

The risk of thrombotic recurrences, even during anticoagulation, is high.

No predictive factor for bleeding events has been identified (e.g. age, time receiving anticoagulant therapy, primary versus secondary antiphospholipid syndrome, positivity for anticardiolipin antibodies, positivity for lupus anticoagulant, previous arterial thrombosis, previous stroke, previous venous thrombosis and previous thrombocytopenia).[2,3]

Case report

A 34-weeks pregnant woman with Hughes syndrome presented with bilateral, fluctuating hearing loss, pressure sensations in the ears and severe otalgia, followed by otorrhoea and bleeding from the ears. She had been suffering from intermittent epistaxis during the pregnancy and had once required nasal cautery. The patient's international normalized ratio of prothrombin time was being monitored regularly. She was receiving clexane and a low dose of aspirin. A sore throat was present a few days prior to the ear symptoms.

On ENT examination, no abnormality was found regarding the right ear, nose, oral cavity, oropharynx and nasopharynx. The left drum was intact and red in colour, suspicious of a vascular lesion in the middle ear or a haemotympanum. An angioma was present on the distal posterior wall of the external auditory canal.

The audiogram showed normal hearing on the right to 2000 Hz, with a high frequency conductive loss at 4000 Hz. On the left, hearing was normal to 500 Hz, with a high frequency conductive loss

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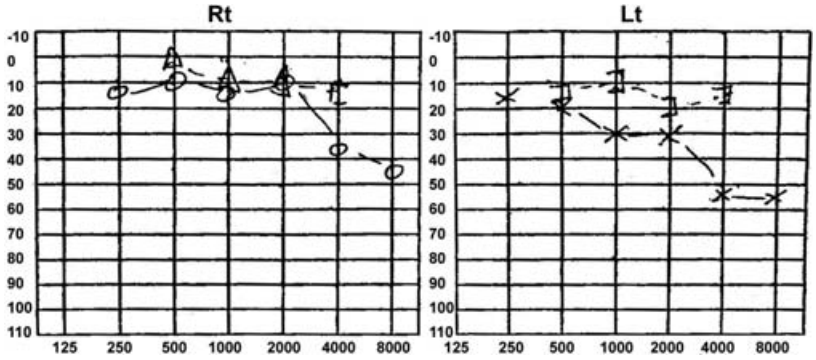

FIG. 1
Initial audiogram. Rt = right; Lt = left

above this frequency (Figure 1). A tympanogram was not performed because of the patient's pain.

The patient's aspirin was stopped and she was prescribed amoxicillin.

By the next day, there was a subjective improvement in the patient's hearing.

However, two days later, the patient's hearing loss increased and her otalgia became severe. In spite of this, examination the following morning showed normal ear drums. There was also a blood clot on the posterior wall of the left external auditory canal.

A further audiogram showed no change in right-sided hearing thresholds, compared with the earlier audiogram; however, on the left, there was a 10 dB deterioration in the 250–2000 Hz region (Figure 2). Tympanograms showed a type A pattern in the right ear, while the left ear showed a double peak, suggestive of some mass effect in the middle ear (Figure 2). The patient's pain had by this time resolved.

The patient was reviewed five weeks later (two weeks after her child's delivery). The ENT examination, audiogram and tympanogram (type A) were all normal (Figure 3).

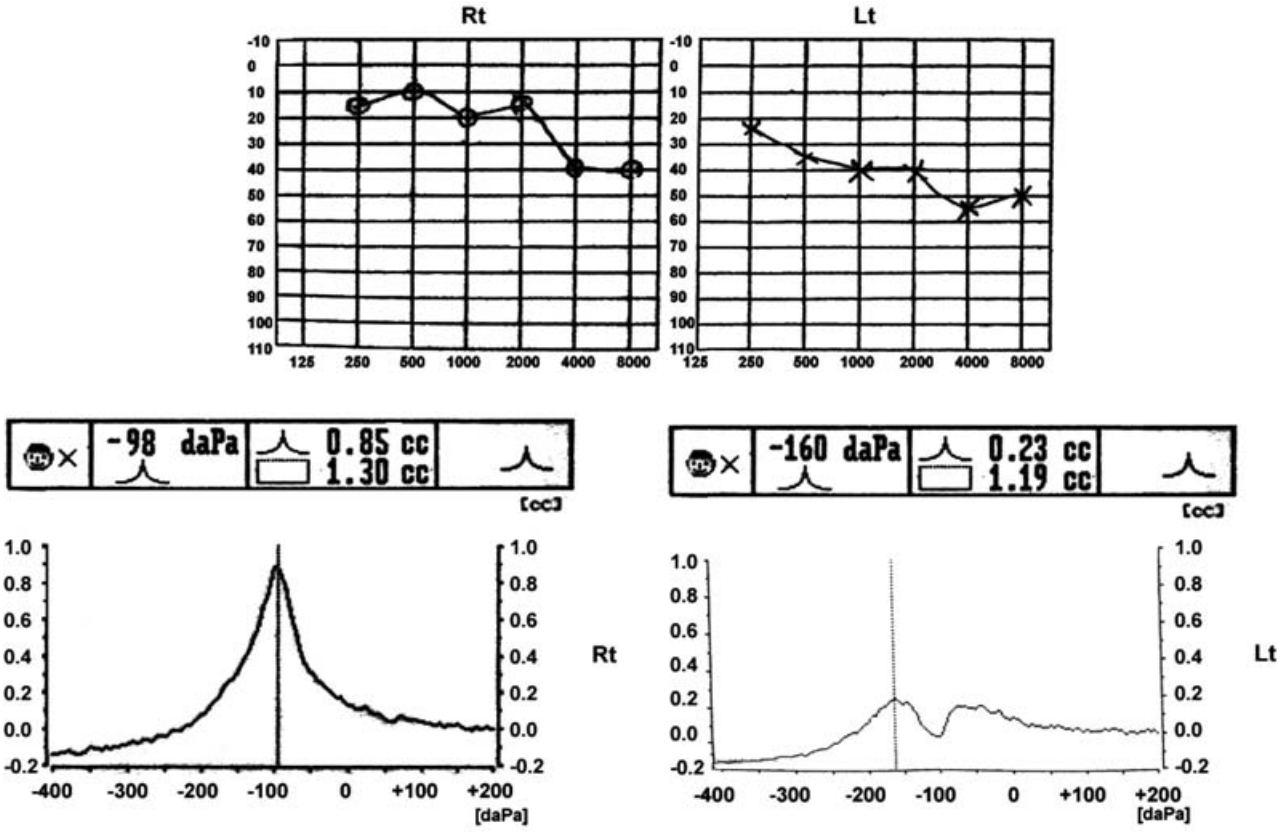

FIG. 2
Audiogram and tympanogram during treatment. Rt = right; Lt = left

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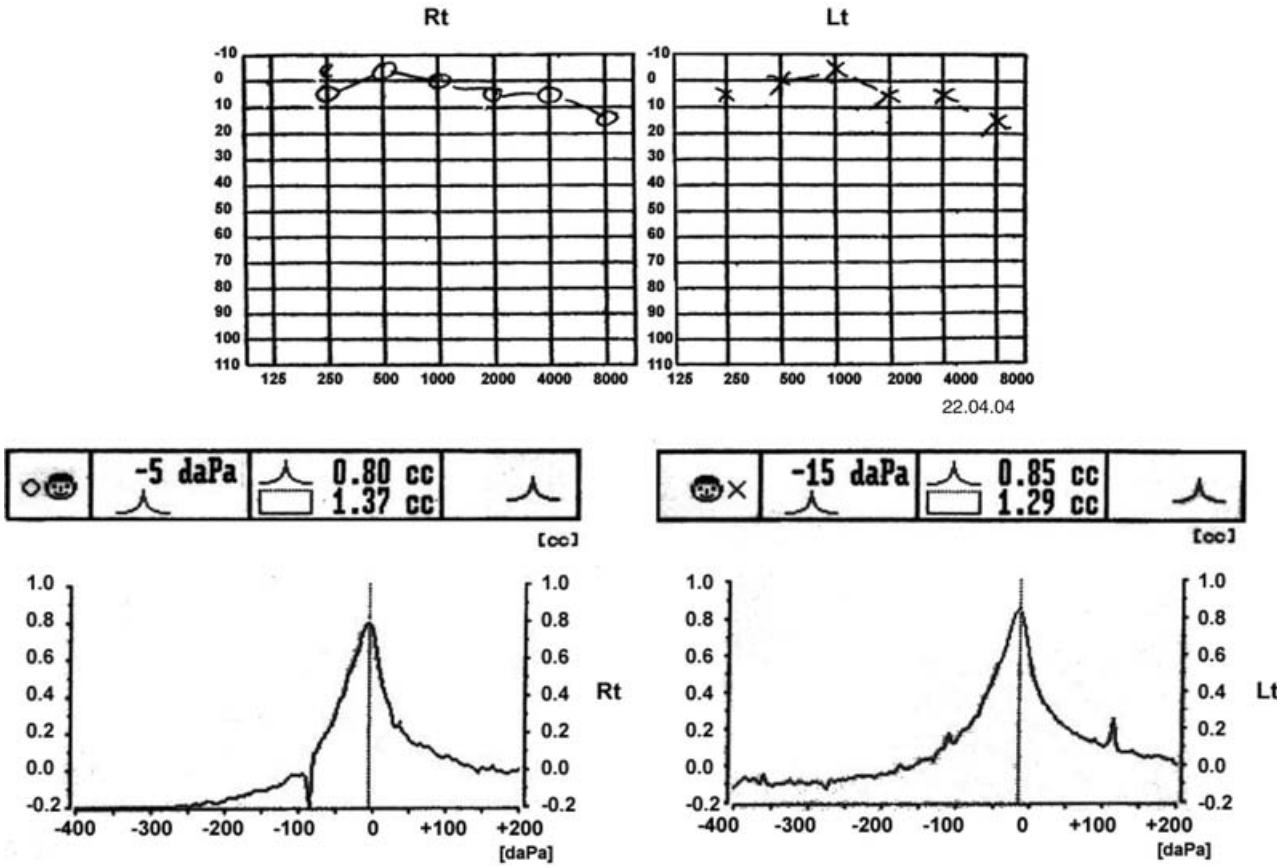

FIG. 3
Audiogram and tympanogram following treatment. Rt = right; Lt = left

Discussion

Fluctuating hearing loss is well known in ENT practice and may result from different causes (e.g. middle ear or cochlea, mechanical, or electrolyte or vascular). The vascularisation of the cochlea is terminal and very fragile. The circulatory networks of the external ear is supplied by the auriculotemporal branch of the superficial temporal artery and branches of the posterior auricular branches of the external carotid artery. The middle ear and mastoid are supplied by a different set of arterial branches mainly from the external carotid system (internal maxillary arthery, the middle meningeal arthery, the stylomastoid arthery, inferior tympanic arthery).[13]

Temporary hearing loss is one of a number of ENT conditions reported in pregnancy. Other such conditions include cutaneous, otologic, rhinologic, oral, pharyngeal and laryngeal manifestations, which are mostly benign and resolve after pregnancy.[14–17]

The possibility of a bleeding nasal polyp[14] was relevant to our patient. However, she had simple epistaxis requiring cautery, with no evidence of nasal polyp. Nasal pyogenic granuloma gravidarum is a similar lesion which does not resolve; it occurs as oral or nasal lesions in approximately 5 per cent of pregnant women and requires radical excision through an open rhinotomy after superselective embolisation.[18,19]

Otoneurological problems occurring during pregnancy include changes in signal processing of the hearing system (electrolyte balance and neurological alterations are attributed to hormone fluctuations)[20,21] and cranial nerve palsy.[22] Most are benign and reverse after parturition.

Permanent conductive hearing loss presenting in pregnancy can be due to otosclerosis which is aggravated in pregnancy.[23,24]

Temporary hearing loss in pregnancy can be neurologic, vascular or due to eustachian tube problems. Hearing loss due to neurological pathology would not be painful or accompanied by abnormalities of the tympanic membrane. In our case, there was no evidence of eustachian tube pathology and the tympanogram was not flat (as would be expected in the presence of an effusion); furthermore, on examination, the rhinopharynx was normal. Eustachian tube problems would have affected both ears, but our patient's middle-ear pressures were consistently normal on both sides.

Hughes syndrome is known to cause a variety of vascular abnormalities. Bleeding into the cochlea would have caused a more permanent sensorineural hearing loss, with discomfort rather than pain and a normal tympanic membrane appearance. It is therefore probable that our patient's problems were due to vascular pathology in the middle and external ear. This has not previously been described in the literature.

Our findings are contrary to expectations: the patient's hearing loss was entirely conductive.

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The pressure sensation in both ears, with fluctuating hearing loss and apparently normal ear drums, could be explained by the existence of an angioma or capillary haemangioma in the middle ear, similar to the one seen in the left external auditory canal but arising from different vessels (as the external ear is supplied by the auriculotemporal branch of the superficial temporal artery and by branches of the posterior auricular branches of the external carotid artery). The middle ear and mastoid are supplied by a different set of arterial branches, mainly from the external carotid system (i.e. the internal maxillary artery, middle meningeal artery, stylomastoid artery and inferior tympanic artery).

- **Hughes syndrome (antiphospholipid syndrome) consists of multiple pulmonary aneurysms and peripheral venous thrombosis**
- **Otorhinolaryngological manifestations of Hughes syndrome are uncommon**
- **This paper describes a patient with fluctuating hearing loss due to Hughes syndrome**
- **The authors discuss possible mechanisms of otological involvement in this condition**

An abnormal left tympanic membrane was seen on day one. It is possible that the patient's otalgia was caused by the pressure of an angioma or capillary haemangioma in the middle ear, followed by bilateral effusion as a reaction to micro-haemorrhages, as well as the outer ear otorrhoea.

The improvement of hearing after delivery was probably due to a change in the patient's antiphospholipid status.

Conclusion

Otorhinolaryngological complications appear to be relatively uncommon in Hughes syndrome. However, the risk of thrombotic events, even during anticoagulation, is high; these can be irreversible, and fatal if uncontrollable bleeding develops.

In such a very rare case, a multidisciplinary approach to diagnosis and treatment is essential. Medical treatment during pregnancy must be carefully and individually coordinated.

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