Combined perilymphatic fistulas of the round window and lateral semicircular canal
A report of 2 cases

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Summary

Two patients with combined perilymphatic fistulas of both the round window and the lateral semicircular canal are presented. They became asymptomatic only when both fistulas were closed. In both cases the hearing improved concurrently. It is recommended that when a traumatic perilymphatic fistula is not cured by closing window fistulas, the lateral semicircular canal be explored.

During the past 15 years otologists have learned that sensorineural hearing loss and vestibular symptoms after different forms of trauma may be associated with the operative finding of perilymph leaking into the middle ear through a fistula in the stapediovestibular ligament of the oval window, or in the round-window membrane.

In 1968 Fee1 described the finding of three oval window fistulas, two after head injury and one with no definite preceding trauma. In 1971 Goodhill2 gave three more accounts of surgically confirmed fistulas, and in 1976 he described his experience with 47 examples.3 The preceding trauma included closed head injury, which accounted for two of the first cases described by Fee,1 and for about half of the cases Healy et al.4 described in 1974. This cause has been implicated in many other cases.

As far as we know, all reported cases had fistulas in the round or oval windows.5-11 We report 2 cases of combined fistulas in both the round windows and lateral semicircular canals.

Case reports

Case 1

A 23-year-old man presented 1 week after suffering blunt traumatic assault to his head, involving mainly the right side. The patient complained of inability to move the right side of his face and severe vertigo. On examination we found complete right facial paralysis and nystagmus grade III. Otoscopically the ears were normal. On mastoid radiography a linear fracture 3 cm long was noted in the right temporal bone. Audiometry showed mixed hearing loss of 45/40 dB in the right ear. The left ear was normal. Exploration of the right ear was undertaken under general anaesthesia, and a rupture of the round window was found. In addition a fracture was found over the facial nerve from the horizontal part to the lateral semicircular canal and along the canal. A large fistula of 1 x 2 mm (with perilymph leakage) was found at the end of this fracture line. The facial nerve was decompressed, and both fistulas (round window and semicircular canal) were closed with fascia from the temporal muscle. One day later the patient was still dizzy with vertigo but the nystagmus had improved to grade I; 3 days later the nystagmus had abated, and the patient was free from vertigo and dizziness.

Three months later, his facial movements were symmetrical and normal, and hearing in the right ear had improved to 20/20 dB. Fourteen months after surgical repair the patient is completely well.

Case 2

A 34-year-old man was seen 10 days after receiving stab wounds and blunt trauma to his head. He complained of dizziness and walked with a very wide base. Nystagmus grade I was present. Otoscopically the ears were normal. Audiometry revealed mixed hearing loss in the right ear of 35/30 dB, and a normal left ear. Mastoid radiography did not show any fracture. At operation under general anaesthesia a small rupture of the round window of the right ear was found with perilymph leakage, and this was closed with temporal fascia.

Postoperatively the patient was less dizzy and the nystagmus disappeared, but he still walked on a wide base, and his hearing deteriorated to 55/55 dB. These persistent symptoms prompted exploratory surgery 6 weeks after the initial operation. The round and oval windows were found to be intact. The lateral semicircular canal was then inspected by antrotomy, and a linear fracture (0,5 cm long) was found which incorporated the semicircular canal and which was leaking perilymph. This was sealed with fascia temporalis. The day after operation, the patient had no subjective dizziness, and after 3 more days he walked with a normal gait. Ten months postoperatively he was asymptomatic, and his hearing had improved to 30/30 dB.

Discussion

Post-traumatic perilymphatic fistula does not pose a major diagnostic problem,12,13 but although a fistula can be demonstrated and repaired, some of the patients have residual and persistent dizziness, vertigo, nystagmus and hearing loss. Halvey and Sade14 reported that dizziness persisted in 22% of patients and Althaus15 reported 24% with persistent vertigo. Goodhill,16 Healy et al.14 and Singleton et al.7 also reported that not all of their patients recovered from dizziness and vertigo. Persistent hearing loss is an even commoner problem: Halvey and Sade14 reported that none of their cases improved; Althaus15 reported hearing improvement in only 34%, and Goodhill16 in
41%. Other authors report a significant degree of hearing deterioration in some of their patients.

In the patients we described, combined fistulas of both the round window and lateral semicircular canal were found. In the first patient both fistulas were closed at the initial operation. Although both of these were large fistulas, the patient regained his sense of balance and hearing very soon after the operation. In the second case, slight improvement of dizziness was found after closure of the round window, but his hearing deteriorated from 30 dB to 55 dB. After closure of the fistula in the semicircular canal his dizziness abated and his hearing improved to 30 dB. Two cases are not statistically significant, but on the other hand both of them improved dramatically after closure of both fistulas, and in our second case we could see that closure of only one fistula improved the patient’s condition only slightly. We now believe that some of the traumatic fistulas encountered are combined fistulas; if a patient shows no or only minimal improvement postoperatively, especially if dizziness or vertigo does not abate, an exploration of the lateral semicircular canal is mandatory.

Case report

A 24-year-old white man presented with symptoms of markedly reduced effort tolerance and intermittent atypical chest pain over a period of 6 months. He had previously played competitive tennis 5 times a week, but at the time of admission he could hardly manage a few minutes. The patient denied any family history of cardiac disease. Physical examination revealed a healthy-looking young man. The pulse rate was 70/min, regular, and the blood pressure 120/80 mmHg. The apical impulse was not displaced but was forceful in character. No gallop sounds or murmurs were heard. Chest radiography was normal with a cardiothoracic ratio of 42%. The left ventricular ejection fraction was 74%. A left ventriculogram was obtained in the right anteroposterior and lateral projections. The characteristic spade-on-a-playing-card configuration of apical hypertrophic cardiomyopathy (AHM) at end-diastole was clearly demonstrated (Fig. 2). The ratio between apical and mid anterior free-wall thickness was 1.77 (normal value 1.05 ± 0.24). Clinical examination and ECGs in the patient’s parents and two younger siblings were normal.

Summary

The clinical, ECG and angiographic features of apical hypertrophic cardiomyopathy in a 24-year-old white man are reported. Only 4 non-Oriental cases have so far been described and the present case is the second from South Africa. Attention is drawn to the possibility that there are two distinct types of apical hypertrophic cardiomyopathy.

REFERENCES


Discussion

The syndrome of non-obstructive hypertrophic cardiomyopathy with marked concentric apical hypertrophy appears to be common in Japan. In fact, Yamaguchi et al. reported an incidence of 3% in patients who had left heart catheterisation and angiography for evaluation of either ischaemic heart disease

Apical hypertrophic cardiomyopathy

A case report

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