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Clinical Records

Meatoplasty in Marfan syndrome

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Abstract
Marfan syndrome (MS) is a rare connective tissue disorder, uncommon in otological practice. Ear surgery in MS has not been previously reported. An identical complication of meatoplasty developed in two patients with MS after modified radical mastoidectomy and meatoplasty for chronic otitis media. Both cases presented post-operatively with an external auditory canal narrowing when the walls of the initial meatoplasty in both cases collapsed. It is proposed that this occurred because of the inherent structural abnormality of the cartilage in Marfan syndrome. A simple technique of revision meatoplasty is described that resulted in successful management of these cases.

Key words: Ear cartilages, surgery; Marfan syndrome

Introduction

There are a number of genetically controlled defects of collagen and glycosaminoglycan metabolism that result in diseases of connective tissue. These diseases include osteogenesis imperfecta, Ehler-Danlos syndrome, Marfan syndrome (MS) and epidermolysis bullosa (Prockup, 1991). The genetic defects in connective tissue metabolism result in weaker and more lax cartilaginous structures and a poor wound healing metabolism (Cohen *et al.* 1994). MS is defined on the basis of characteristic changes in three connective tissue systems: the skeleton, the eyes and the cardiovascular system (Prockup, 1991). The majority of published literature on surgery in MS involves cardiovascular problems and it was not possible to find any previous work on otological procedures in patients with MS. An identical complication of meatoplasty developed in two cases of MS after ear surgery had been performed. This is described and a simple technique to correct this complication is presented.

Case reports

Case 1

A 23-year-old male was referred to the senior author (PAF) in 1991 for assessment of a chronically discharging left mastoid cavity. He was a known case of MS. He had had a left modified radical mastoidectomy for cholesteatomatous disease in 1982 and a revision procedure with a meatoplasty in 1989. The meatoplasty had collapsed and access to the postero-superior aspect of the cavity was limited. He had a second revision procedure with amputation of the mastoid tip and a Fisch-type meatoplasty performed at our unit (Fagan, 1991). The meatoplasty again collapsed into a vertical slit after this operation (Figure 1) and he had further surgical intervention at which time an 0′ silk suture was used to hook the roof of the cartilaginous canal to the temporalis muscle (Figure 2). This supporting suture has been successful in achieving an adequate meatal opening and as a consequence a dry, self-cleansing ear. He has recently had an ossicular chain reconstruction with a good hearing result. At three-year follow-up the meatoplasty has remained patent and he has had no further problems with otorrhoea.

Case 2

A nine-year-old male was referred to the senior author for assessment of a post-operative meatal stenosis. He had had a right-sided intact-canal wall mastoidectomy performed four years previously for an acquired cholesteatoma. This required transformation to a modified radical procedure 10 months afterwards. He continued to retain epithelium and this was proving difficult to clean because of the associated development of a meatal stenosis. An exploration of his right ear was performed by the senior author and considerable recurrence of cholesteatoma was discovered in the middle ear and attic areas. The disease was cleared, all overhangs removed, mastoid tip amputated and a very wide meatoplasty carried out, after the method of Fisch (Fagan, 1991). He had a second stage ossiculoplasty, using a total ossicular replacement prosthesis (TORP) and a cartilage graft, performed 18 months later. He was lost to follow-up for the next two years but re-presented with a 12-month history of otorrhoea. He was found to have a meatal narrowing which was due to collapse of the canal walls (Figure 1) and could be held open quite easily with an aural speculum. A recent, tentative, diagnosis of MS had been made at another centre arising from cardiac and skeletal abnormalities. A revision procedure was performed at which the only abnormality found was the collapsed meatus. This was rectified by elevating the roof of the canal and keeping the meatus patent by using a heavy nylon suture support

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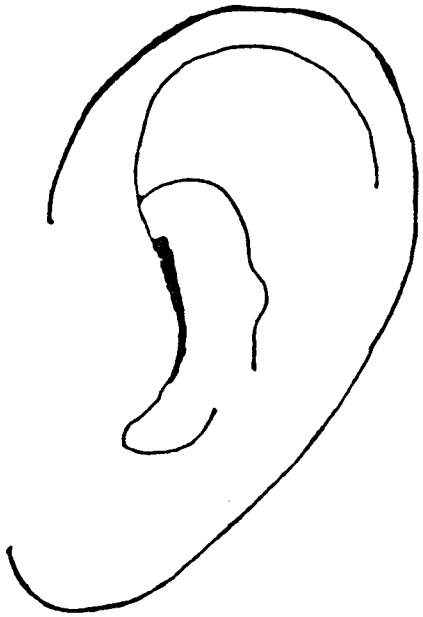

FIG. 1
Diagrammatic representation of the type of meatal narrowing.

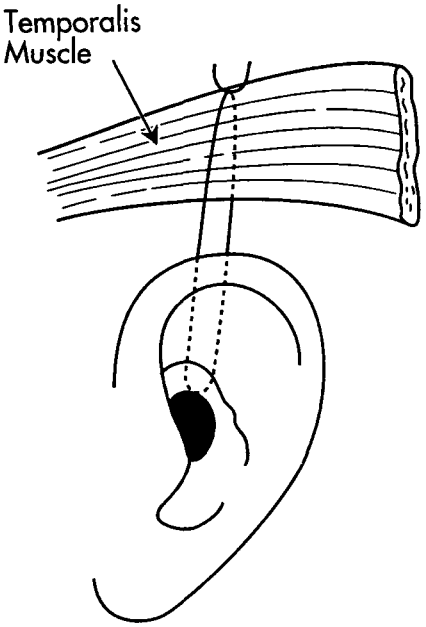

FIG. 2
Diagrammatic representation of the type of meatal repair.

anchored to the temporalis muscle (Figure 2). At three-year follow-up he has a dry, self-cleansing cavity, a wide meatal opening and is able to swim without special precautions.

Discussion

MS is inherited as an autosomal dominant trait and appears in each generation but occasionally the expression of the gene may be suppressed so that only some or perhaps none of the manifestations of the disease occur (Gardner, 1987).

The molecular defects in most variants of MS are unknown. Some of these patients have defects in their collagen structure and some have abnormal fibrillin in their elastin (Prockup, 1991). It is suggested that it is caused by mutations of the fibrillin gene located on chromosome 15 (Tsipouras and Devereux, 1993). There are defective cross-linkages of collagen fibres possibly due to a defective alteration in their primary structure (Walter and Israel, 1987). These abnormalities make surgery more difficult and wound healing more complicated (Prockup, 1991). The diagnosis is a clinical one and there is no established treatment (Cohen *et al.*, 1994).

An adequate meatoplasy is one of the important principles in achieving a dry, self-cleansing cavity in modified radical mastoidectomy (Sade *et al.*, 1982). The meatoplasty that was utilized in both these cases is the standard one performed by the senior author (PAF) for the past eight years in surgery for chronic ear disease and canal-widening procedures and is modelled on the method of Fisch (1980). A full-thickness incision extending backwards and upwards through the root of the helix is performed. The skin overlying the cartilage is mobilized and two small superior and inferior triangles of cartilage based on the incision are excised. Soft tissue deep to the skin flaps is then excised and the flaps are sutured to the deep surface.

Post-operative canal stenosis is associated with soft tissue fibrosis and not the cartilaginous collapse resulting in a vertical slit (Figure 1) seen in these two cases. The meatus could be held open quite easily in both cases by inserting an aural speculum and when the pinna was retracted upwards and backwards, the meatus was entirely adequate. These were the only two cases in the entire series of patients at our department, who had a Fisch-type meatoplasty performed, with this form of collapse.

It is our contention that by excising cartilage in an ear canal, already compromised by structurally abnormal cartilage, the cartilaginous framework was not strong enough to maintain a proper meatus. This should be taken into consideration when patients with connective tissue disorders are undergoing surgery and the suture support that we have described should be included in the initial surgery. It would not add much time or any morbidity to the procedure and would help to prevent the need for revision surgery.

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