INFECTIVE ENDOCARDITIS IN MARFAN'S SYNDROME — A CASE REPORT

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SYNOPSIS

A case of Marfan's Syndrome with mitral valve prolapse and infective endocarditis is reported. The cardiovascular complications of Marfan's Syndrome is briefly reviewed.

INTRODUCTION

Dolichostenomelia which translates to mean 'long, thin limbs' was first described by Marfan in 1896 (1). It has subsequently been referred to as Marfan's Syndrome and several other features of disease including ocular and cardiovascular disease have been described. Among the cardiovascular complications are aortic aneurysms, dissection of the aorta and mitral and tricuspid valve prolapse. As with other valve deformities or anomalies, reports of infective endocarditis have been described (2). We report below such a case of infective endocarditis on a prolapsed mitral valve in Marfan's Syndrome.

CASE REPORT

HSC is a 23-year-old Chinese woman who presented to us with a three-week history of high fever with chills and rigors. She did not give a history of any obvious focal source of infection and she was unaware of any existing cardiac valvular disease.

Physical examination revealed a thin, tall woman with unmistakable features of Marfan's Syndrome (Fig. 1). She had long fingers, an arm span of 185 cm compared to a height of 165 cm and high-arched palate (Fig. 2). No ocular abnormalities were detected. Incidental examination of her father revealed similar features. She was also febrile and ill. She was pale and had subconjunctival haemorrhages (Fig. 3), a splinter haemorrhage (Fig. 4) and numerous Roth spots in both fundi. She was noted to have several carious teeth but did not have any dental procedures carrried out recently. Cardiac examination revealed a pan-systolic mururmur in the mitral area, radiating to the axilla, consistent with mitral incompetence. A clinical diagnosis of Marfan's Syndrome) with mitral valve prolapse and superimposed infective endocarditis was made.

The cardiac diagnosis of mitral valve prolapse was confirmed by a 2D-echocardiogram examination. No vegetations were seen on the valve leaflets and the aortic root and ascending aorta were normal. Streptococcus viridans was grown from blood cultures and the patient commenced on appropriate intra-venous antibiotics with good response to treatment.

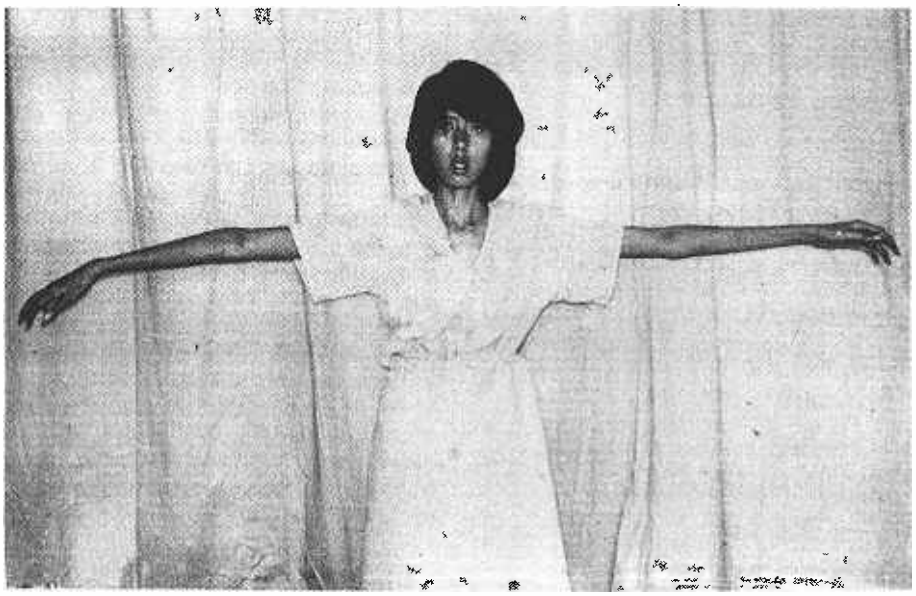

Figure 1 — Long arm span and arachnodactyly

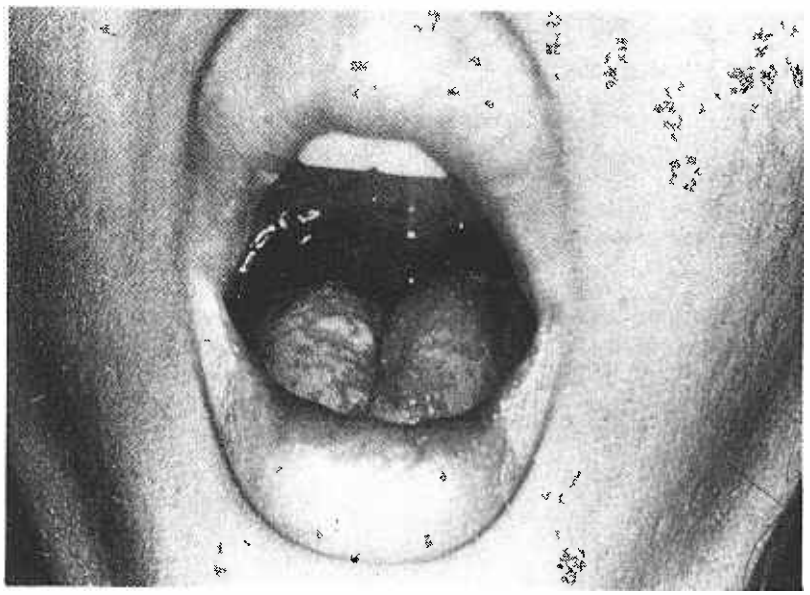

Figure 2 — High arched palate

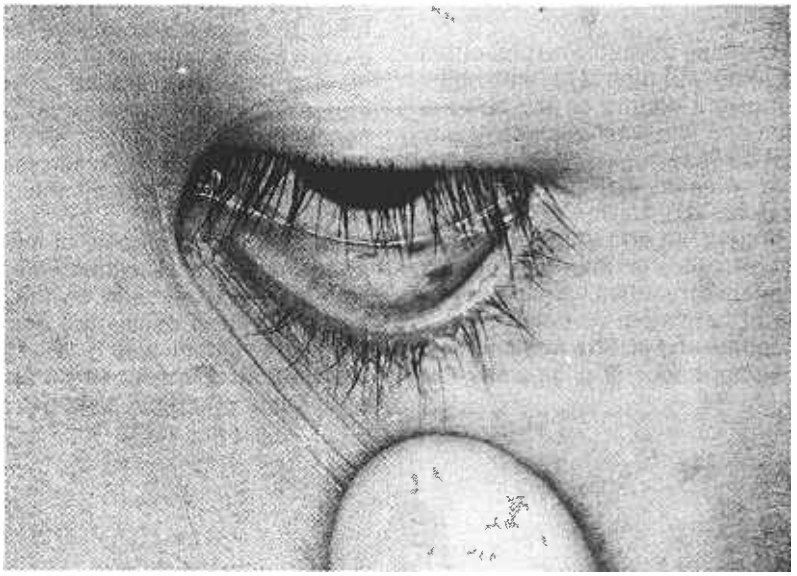

Figure 3 — Subconjunctival haemorrhage

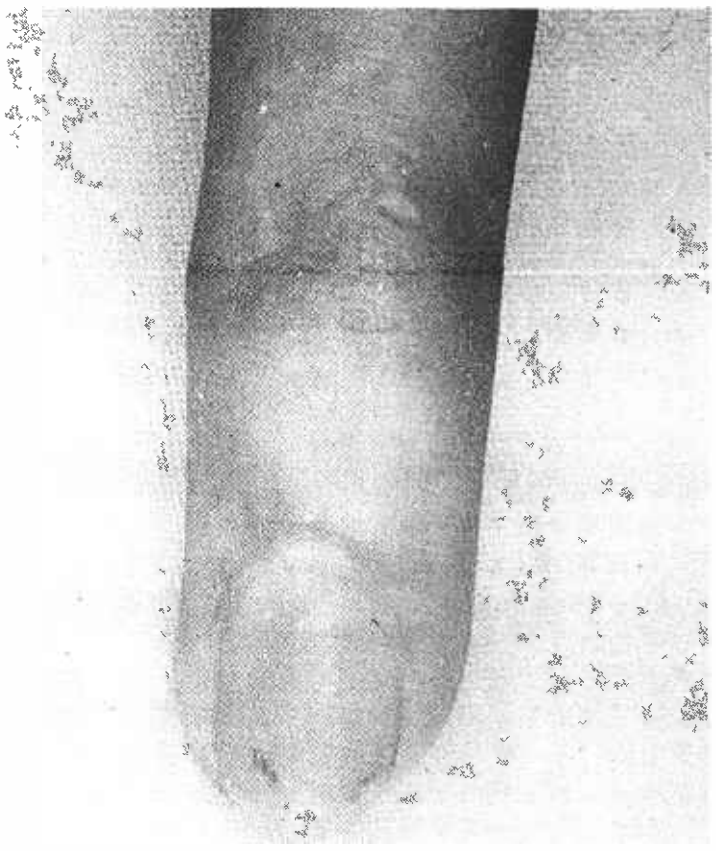

Figure 4 — Splinter haemorrhage

DISCUSSION

Marfan's Syndrome is classified as a heritable disorder of connective tissue because clinical and pathological alterations involve supporting elements. It has been assumed that an inborn error of protein metabolism, particularly in collagen or elastin accounts for Marfan's Syndrome but conclusive evidence is not yet available. The prevalence of Marfan's Syndrome is estaimated to be about 4 to 6 per 100,000 without any discernible racial or ethnic predilection (3).

Cardiovascular complications of Marfan's Syndrome is well known and abnormalities like aneurysm of the ascending aorta, dissecting aneurysms and valve prolapses stem from the laxity of the supporting tissues. In Pyeritz and McKusick's report (3), 39 of 50 consecutive patients studied had some cardiovascular complication, and of interest, 15 had mid-systolic clicks alone and 9 had mid-systolic clicks and a late-systolic murmur; these findings being considered as evidence for a mitral valve prolapse. In other studies (4, 5), up to 60% of persons believed to be affected by Marfan's Syndrome have auscultatory evidence of either mitral or aortic regurgitation or systolic clicks. Roberts and Honig (6) studied 18 patients with Marfan's Syndrome at necropsy and were able to subdivide them into three distinct groups on the basis of different cardiovascular lesions. In Group 1 were patients with fusiform ascending aorta aneurysms, Group 2, dissecting aneurysms and Group 3 isolated mitral regurgitation. Of the 9 patients who had mitral regurgitation, 6 were found to have floppy mitral valves.

Cases of infective endocarditis in Marfan's Syndrome have not been frequently reported. Wunsch et al (2) noted that up to their report in 1965 only 5 cases have been described. In his review, all 6 cases including his own involved the mitral valve. However, some of the patients described had evidence that the valve abnormalities were rheumatic in origin rather than as a complication of Marfan's Syndrome.

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