AORTIC VALVE REPLACEMENT IN OSTEOGENESIS IMPERFECTA TARDA — A CASE REPORT

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ABSTRACT

A case of osteogenesis imperfecta tarda presenting with infective endocarditis and heart failure is discussed. Urgent aortic valve replacement was performed but the patient succumbed from pneumonia. The rarity of this disorder and the special problems encountered surgically in these patients are briefly discussed.

Key Words: Osteogenesis Imperfecta Tarda; Aortic Valve Replacement

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INTRODUCTION

Patients with osteogenesis imperfecta are often physically handicapped by their skeletal deformities. We, however, describe a patient who was incapacitated by his cardiovascular disease requiring valve replacement.

CASE REPORT

T.A.L., 19 years old, male, was admitted with progressive exertional dyspnoea and fever. As a child, he had often sustained multiple fractures following minimal trauma. Both his mother and sister had blue sclera and his sister, too, had brittle bones.

On admission, he was febrile (temperature: 38 degrees Centigrade) with sinus tachycardia (120 per minute) and a blood pressure of 180/0 mm Hg. He had blue sclera and multiple bony deformities. Hearing and dentition were normal. There was aortic regurgitation with cardiomegaly. He was in congestive cardiac failure. Investigations showed a normal haemoglobin (13.2 gm/ 100 ml) and a leucocytosis of 11,000/ul. The chest X-ray showed gross cardiomegaly and pulmonary edema. An electrocardiogram revealed left ventricular hypertrophy. A 2-dimensional echocardiogram confirmed left ventricular enlargement and fluttering of the anterior leaflet of the mitral valve was seen. The aortic annulus was dilated (30 mm in diameter) and the valve leaflets appeared long and redundant. The mitral and tricuspid valves appeared normal. Blood cultures grew Staphylococcus aureus.

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The patient was started on anti-failure therapy and parenteral antibiotics. Urgent aortic valve replacement was performed. At operation, the aortic valve leaflets were noted to be thin and deficient and the annulus was dilated. The anterior mitral valve leaflet was flail with a single chordal rupture. The aortic valve was replaced with a Bjork — Shiley mechanical prosthesis and buttressing with Dacron sutures was done. The patient made a good immediate post-operative recovery but died on the tenth post-operative day of pneumonia. Histopathological examination of the excised valve showed myxoid degeneration.

DISCUSSION:

Osteogenesis imperfecta is a heritable disorder of connective tissue but cardiovascular involvement is rare with less than 20 cases reported in the literature. Mc Kusick (1) in his analysis of more than 1000 patients with osteogenesis imperfecta reported only two patients with findings suggestive of aortic regurgitation while Hortop (2) et al found only 4 cases of valvular dysfunction in 109 patients with osteogenesis imperfecta.

The aortic valve, followed by the mitral valve, are the most commonly involved in osteogenesis imperfecta. The valve annulus may be dilated, as in our patient, and in one of two patients in Hortop's series with aortic incompetence. The valve leaflets may be thin, attenuated and prolapsed with ruptured chordae tendineae, all resulting in valvular incompetence. Occasionally, rarer anomalies may occur. Heppner (3) et al in 1975 described one patient who had an aneurysm of the sinus of valsalva and one of the three patients described by Criscitiello (4) et al had fenestrations in the aortic and pulmonary valve cusps and an aneurysmal deformity of the anterior leaflet of the mitral valve leaflet. The underlying pathology is extensive myxoid degeneration of the heart valves.

Heart valve surgery in patients with osteogenesis imperfecta poses certain special problems. There is often difficulty in placing sutures because of tissue friability. We used the method described by Weisinger (5) et al using Dacron sutures with buttressing of the suture line. Poor wound healing, wound dehiscence and a bleeding diathesis due to abnormal collagen synthesis may also occur. Despite this, valve replacement may be undertaken successfully in osteogenesis imperfecta.

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