Calcific pulmonary stenosis

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Two cases of pure calcific pulmonary stenosis are reported. The diagnosis was suspected because of an immediate diastolic murmur suggesting pulmonary regurgitation in association with auscultatory features of pulmonary stenosis. Valvar calcification was confirmed at operation in one patient and was demonstrated radiologically in the other.

It is proposed that a diastolic murmur of pulmonary regurgitation in patients with auscultatory signs of pulmonary stenosis suggests the possibility of calcific pulmonary stenosis.

Calcific pulmonary stenosis is rarely diagnosed in life. To date, only 20 cases with radiological evidence of calcific pulmonary valve stenosis have been reported, and in most circumstances they have been associated with other cardiac malformations (Gabriele and Scatliff, 1970; Rodriguez, Bennett, and Lehan, 1971). This communication reports two new cases of pure calcific pulmonary stenosis and indicates how the diagnosis may be made clinically.

Case reports

Case 1

A 33-year-old man was referred because of a 6-month history of easy fatigability, dyspnoea on effort, palpitations, and occasional lightheadedness. A heart murmur was first detected at 8 years.

Physical examination showed a praecordial bulge, right ventricular heave, and a systolic thrill along the left sternal border. A loud fourth heart sound was present. The second heart sound was widely split and had a decreased pulmonary component. A loud, 5/6, ejection systolic murmur preceded by a click was present in the pulmonary area and along the left sternal border. A short, immediate diastolic murmur, better heard in the upright position (2/6), was found in the same area (Fig. 1). The auscultatory findings were considered compatible with severe pulmonary valve stenosis and mild pulmonary regurgitation.

The electrocardiogram revealed severe right atrial and ventricular hypertrophy. The chest x-ray showed mild cardiac enlargement, decreased pulmonary vascature, and a prominent pulmonary trunk. No intracardiac calcification could be detected.

A haemodynamic study was not performed, and the patient was submitted to open heart surgery. Right atrial enlargement, severe right ventricular hypertrophy, and marked dilatation of the pulmonary artery were present. The pulmonary valve was severely narrowed, bicuspid, and heavily calcified. Severe infundibular

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FIG. 1 Phonocardiogram of Case 1. A loud fourth heart sound (S_4) precedes the first heart sound (S_1). A very loud ejection systolic murmur and a short early diastolic murmur (DM) can be seen.

stenosis was also present. No evidence of previous bacterial endocarditis was found. A pulmonary valvotomy, with removal of the calcium deposits and infundibular resection, was performed.

The postoperative course was uneventful, and the patient was discharged from hospital 10 days after operation. Both the systolic and diastolic murmurs were still present but the ejection systolic murmur was much softer (3/6). The fourth heart sound and the ejection click were no longer audible.

Case 2

A 19-year-old man was referred for evaluation of a murmur discovered in early infancy. His only complaints

were atypical chest pain and shortness of breath on severe exertion. There was a history of poor somatic growth, peptic ulcer, and hypercalcaemia, which was probably secondary to hyperparathyroidism. Determinations of serum calcium on three consecutive days were 9, 11, and 10.4 mg/100 ml, respectively. Estimations of urinary calcium excretion in 24-hour periods and under a balanced diet were 368, 219, and 140 mg. Tubular reabsorption of phosphate was 78 per cent. The clinical and laboratory findings were not considered to warrant exploration of the neck.

On physical examination he was obviously underdeveloped for his age. The face was somewhat unusual with a prominent nose and low set ears. The right arm was hypotrophic with reduced motility of the shoulder and elbow joints. X-rays of the right upper limb disclosed epiphyseal dysplasia of the head of the humerus. The feet were also malformed showing pronated deviation. Auscultation of the heart revealed a widely split second heart sound with a pulmonary component that was diminished in intensity. A grade 3/6 ejection systolic murmur and a grade 2/6, short, immediate diastolic murmur were present in the pulmonary area (Fig. 2). Based on the auscultatory findings, mild pulmonary stenosis and insufficiency with calcification of the pulmonary valve was diagnosed.

The electrocardiogram showed an S₁, S₂, S₃ pattern, indeterminate QRS axis in the frontal plane, and was suggestive of right atrial enlargement. Chest x-rays revealed a normal sized heart, a slightly prominent main pulmonary artery, and normal pulmonary vasculature. The lateral view suggested the presence of calcium in the pulmonary valve area. Fluoroscopy using an image intensifier at the time of cardiac catheterization clearly showed the presence of calcium in the pulmonary valve area. The right ventricular and pulmonary artery pres-

ures were 35/2 and 12/4 mmHg, respectively, with a gradient of 23 mmHg across the valve. Left-sided pressures were normal and no shunts were present. Selective pulmonary artery cineangiography revealed mild to moderate pulmonary regurgitation not considered to be factitious.

No treatment was advised and the patient was discharged from hospital for continued medical observation at the outpatient clinic.

Discussion

The diagnosis of calcific pulmonary stenosis in the published cases was either made directly at operation or at necropsy (Mirowski, Mehrizi, and Shah, 1964; Roberts *et al.*, 1968), or by radiological demonstration of calcium deposits in the pulmonary valve area (Dinsmore *et al.*, 1966; Roberts *et al.*, 1968). In most reports, little reference has been made to the clinical data of these patients.

The two cases reported here presented auscultatory findings of severe and mild pulmonary stenosis respectively. A brief, soft, immediate diastolic murmur was also heard in both patients. Since the murmur of pulmonary regurgitation occurs somewhat infrequently in patients with pure pulmonary stenosis (Vogelpoel and Schrire, 1960), it was assumed that an additional factor, such as calcification of the valve, could interfere with valve closure and account for the regurgitant murmur. This assumption was corroborated at operation and fluoroscopy in our two cases.

Calcification of a stenotic pulmonary valve usually occurs in middle-aged or elderly patients, and is often associated with cardiac malformations

FIG. 2 Phonocardiogram of Case 2. There is wide splitting of the second heart sound with a decreased pulmonary component (P₂). Both the ejection systolic murmur (SM) and the short early diastolic murmur (DM) can be seen.

(Dinsmore *et al.*, 1966; Duke, 1967; Roberts *et al.*, 1968; Hardy *et al.*, 1969; Gabriele and Scatliff, 1970; Rodriguez *et al.*, 1971). Survival into adult life (Dinsmore *et al.*, 1966; Roberts *et al.*, 1968), sustained rise in the right ventricular systolic pressure secondary to severe pulmonary stenosis (Roberts *et al.*, 1968; Rodriguez *et al.*, 1971), bacterial endocarditis (Dinsmore *et al.*, 1966; Duke, 1967; Hardy *et al.*, 1969), and a previous pulmonary valvotomy (Duke, 1967) have been mentioned as possible predisposing causes for calcium deposition in the pulmonary valve. Our elder patient was 33 years of age, and severe pulmonary stenosis was seen at operation. He had no history of bacterial endocarditis and the surgical findings confirmed this. The other patient was 19 years and had mild pulmonary valve stenosis. No previous case of calcific mild pulmonary stenosis has been reported in a recent review by Rodriguez *et al.* (1971). Though there was no history of bacterial endocarditis, this possibility cannot be excluded. However, this patient had had hypercalcaemia and probable hyperparathyroidism. The metabolic abnormality in the presence of a congenitally deformed valve might then account for the calcium deposition.

It is not clear why calcific pulmonary stenosis does not occur more frequently. Roberts *et al.* (1968) have pointed out the contrast between the frequent occurrence of calcification of the aortic valve and the rarity of calcific pulmonary stenosis. Calcification of the aortic valve has been attributed to a chronic increase in ejection pressure to which a malformed valve is submitted. These factors are found in patients with pulmonary stenosis; however, the occurrence of calcium deposition is extremely unusual. As a possible explanation it has been sug-

gested that these patients seldom survive to the 5th or 6th decade, and in addition, frequently, the venous return to the right heart bypasses the obstruction through atrial or ventricular septal defects (Roberts *et al.*, 1968).

The diagnosis of calcific pulmonary stenosis, especially in cases of severe pulmonary valve stenosis, is of prognostic importance since in these patients the operative mortality is unusually high (Rodriguez *et al.*, 1971).

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