A Complicated Case of Cor Triatriatum Dexter

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The patient, a married woman aged 26 years, had remained well and apparently quite normal until 1962; she had had 4 uncomplicated pregnancies. In September of that year, she developed symptoms of a left basal pneumonia which resolved under standard therapy. No other abnormality was noted at that time. The patient remained well for several months thereafter, but was readmitted to hospital in early February 1963 with an influenza-like illness of two weeks' duration. At this time, she complained of increasing tiredness, generalized headache, sweating, and left-sided pleuritic pain. Physical examination on this occasion was again unremarkable, except that tenderness was noted in the right hypochondrium, though the liver was not clinically enlarged. She then complained of aching discomfort over the right chest wall, not obviously associated with any clinical abnormality. Ten days after admission, oedema of the face became apparent. Examination revealed a swollen right arm, a much enlarged and tender right breast, associated with dilated veins in the chest wall, and non-pulsatile distension of the neck veins. A diagnosis of superior vena caval obstruction was made. The liver was enlarged 2 cm. below the right costal margin, and was tender. The cardiac findings were unremarkable. The heart was in sinus rhythm; the apex beat was diffuse and poorly localized. There was no abnormal praecordial pulsation, nor was any murmur heard. In particular, there was no mid-diastolic murmur of tricuspid stenosis. Serial electrocardiograms showed non-specific, low voltage complexes, and flattening of T waves. X-ray examination of the lungs and mediastinum showed minimal broadening of the right upper mediastinum, the lung fields being clear. In view of the altered clinical picture, particularly in association with previous respiratory infections, exploratory thoracotomy was decided upon and performed in Mearns Kirk Hospital, Glasgow, in March 1963. Granulomatous tissue of indeterminate origin was found in the vicinity of the superior vena cava. A finger inserted via the vena cava to the right atrium revealed induration of the posterior atrial wall; this was considered to be a grossly disorganized and stenosed tricuspid valve. It was felt unjustifiable to explore this further, because of the danger of producing tricuspid incompetence.

At this time, gastric washings were reported as being positive for acid and alcohol-fast bacilli; antituberculous

therapy with para-aminosalicylic acid and isoniazid with streptomycin was instituted. This was continued for 12 months.

Between June 1963 and the terminal illness in February 1966, the patient was admitted to the medical wards on 8 further occasions. The complaints were similar each time, namely, increasing dyspnoea and increased swelling of the extremities and/or the abdomen. Ascites was now a constant finding and it was obvious that the patient had obstruction of both the superior and inferior vena cavae. There was slow but inexorable deterioration in the patient's condition. Despite, ultimately, a regimen of 80 mg. frusemide three times a day, spironolactone 50 mg. four times a day, and the use of Southey's tubes on two occasions, oedema of the extremities, ascites, and recurring pleural transudate became intractable. The patient died 34 months after her initial presentation.

During the period of medical care, intensive efforts were directed towards defining the cause of the obscure lesion encountered at thoracotomy. It was clearly important to determine whether remedial surgery would be possible. To this end, Professor George Smith of the Department of Surgery at Aberdeen University was consulted and he offered to perform venous angiography. The patient was admitted to the Aberdeen Royal Infirmary, where an attempt was made to outline the superior vena cava via the axillary vein. This procedure failed to outline the vessel, but revealed a plexiform system of collateral veins. Further attempts via the right and left femoral veins showed occlusion at the level of the common iliac segments. They demonstrated a well-developed prevertebral plexus and, on the left side, the hemiazygos system. Mr. W. P. Cleland of the Hammersmith Hospital was then consulted regarding the feasibility of inserting replacement grafts of the superior and inferior vena cavae. This resulted in the patient's admission to his unit in the Hammersmith Hospital. Further investigations, including venography, failed to demonstrate a vessel of sufficient size reaching the heart which would be suitable for an anastomotic or bypass procedure. At this time a finding of excessive platelet adhesion was noted. The significance of this observation remains obscure.

The possible value of fibrinolytic therapy was discussed. She was seen by Professor A. S. Douglas of the Department of Medicine, Glasgow Royal

FIG. 1.—Section of heart showing thickened pericardium.

Infirmary, who considered that this treatment was not indicated.

Necropsy Findings

The lungs were firmly adherent to the parietal pleurae and the diaphragm and, on section, they showed bronchiectatic dilatation in both left lobes. Extensive bronchopneumonic changes were present throughout the left lung and the right lower lobe. There was no evidence of pulmonary tuberculosis. On examining the heart, the pericardial sac was found to be completely adherent and, at points, attained a thickness of 1 cm. (Fig. 1). The heart was below average size, weighing 200 g., and, on opening the right atrium, a transverse septum was found dividing the chamber into roughly equal upper and lower halves, communicating through a small orifice of approximately one-eighth of an inch (3–4 mm.) in diameter (Fig. 2) and separating the caval orifices from the tricuspid valve. The tricuspid valve and the right ventricle were normal, as was the pulmonary artery. There was no abnormality on the left side of the heart. There was no patent foramen ovale. The inferior vena cava was obliterated about 2.5 cm. below the insertion of the renal veins and was replaced distally by fibrous tissue. The superior vena cava was patent, though surrounded by organized fibrous tissue. The liver was enlarged and showed fatty change, rather more than that associated with chronic venous congestion, and a fine early cirrhosis was considered to be present. The kidneys were normal.

FIG. 2.—Membrane in right atrium with probe through communicating orifice.

Discussion

Cor triatriatum dexter is a rare, congenital, cardiac anomaly. No similar case has yet been recorded in the records of the Hammersmith Hospital (C. M. Oakley, 1966, personal communication).

In retrospect, it is difficult to see what more could have been done for this patient, despite the presence of a lesion that was eminently suitable for surgical correction. The clinical picture was dominated by evidence of occlusive disease of the great veins, presumably thrombotic, the aetiology of which was unknown. This, in association with a small heart, the absence of right atrial dilatation radiologically, and of a diastolic murmur compatible with tricuspid stenosis pointed to an extra-cardiac, obstructive lesion. The diagnostic difficulty encountered at thoracotomy was due to the presence of organized exudate in the vicinity of the superior vena cava, the significance of which was not then appreciated (J. Welsh, 1966, personal communication). The presence of the intra-atrial membrane obscured the normal state of the tricuspid valve. The nature of the lesion could only have

been ascertained by direct, transthoracic atriotomy, which could not be undertaken unless there was a high probability of a remediable lesion being present, conditions that could not be satisfied in this case.

Summary

A case is presented of cor triatriatum dexter,

complicated by obstruction of the inferior vena cava, by apparent obstruction of the superior vena cava, and, terminally, by constrictive pericarditis.

I wish to thank Dr. T. J. Thomson for permission to study this patient under his care, Dr. W. M. Davidson for the necropsy report, Mr. P. Waldie for the photographs, and Professor G. Smith and Mr. W. P. Cleland for their help and interest in this difficult case.