Atrial Septal Defect Coexisting With Constrictive Pericarditis - A Diagnostic Conundrum

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Abstract
The combination atrial septal defect and chronic constrictive pericarditis is very rare and poses diagnostic challenges because one tends to mask the clinical features of the other. We report such a case in which the initial diagnosis was suspected to be chronic constrictive pericarditis. This diagnosis was later changed to secundum atrial septal defect after transthoracic echocardiography. The complete diagnosis was made intra-operatively and the patient was relieved of both pathologies surgically.

CASE REPORT
A 15 year-old schoolboy was referred to our institution for specialist evaluation on account of anasarca and cough of 6 weeks’ duration. The main symptoms at presentation were progressive dyspnea on effort, a dry cough and generalised body swelling. Physical examination revealed anasarca with engorged neck veins. The pulse was 110/minute, regular with fair volume with a blood pressure of 110/60mmHg. The jugular venous pressure was estimated to be 4cm above the sternal angle. The apex beat was neither visible nor palpable. The second heart sound was split (judged to be physiologic) and there was a grade II/VI ejection systolic murmur loudest at the upper left sternal edge. No pericardial rub was heard. The lung fields were clear to auscultation. Ascites was present and the liver was ballotable. Lower extremity edema was present up to mid-leg level. An abdominal ultrasound examination showed congestive hepatosplenomegaly and ascites. The liver function tests were essentially normal. Chest x-ray showed a cardiothoracic ratio of 0.55. No pericardial calcification was noted. Chronic constrictive pericarditis was suspected to be the cause of the patient’s symptoms.

Decongestive therapy was begun with intravenous frusemide and spirinolactone with good effect.

However, a transthoracic echocardiogram (TTE) showed dilated right-sided chambers and a secundum atrial septal defect (1.8cm) with a left-to-right shunt. The initial working diagnosis was therefore changed accordingly and the patient was subsequently referred to the surgeon as a case of congestive cardiac failure secondary to atrial septal defect (ASD).

The patient remained on oral medication while preparing to undergo ASD closure. There was a relapse of congestive symptoms after a year in spite of oral decongestive treatment. A repeat TTE was performed but no ASD was seen. A transesophageal echocardiogram (TEE) was later performed which demonstrated a secundum ASD (Figure 1) but no colour flow could be detected across the defect.
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A right heart catheterization was done and showed a mean central venous pressure of 15mmHg, mean right ventricular pressure of 22mmHg (33/7mmHg), mean pulmonary artery pressure of 23mmHg (32/16mmHg) and pulmonary capillary wedge pressure of 17mmHg. The mixed venous oxygen tension was 45.6mmHg with a mixed venous oxygen saturation of 82.4% on room air. There was no change in the oxygen saturation after inhalation of 100% oxygen. The features did not clearly confirm the presence of an ASD. Nevertheless, a decision was taken to proceed with surgery with the diagnosis of secundum ASD based on clinical grounds and the TEE findings.

After performing a median sternotomy, the pericardium was found to be grossly thickened and densely adherent to the myocardium with areas of calcification. A formal pericardiectomy was performed without cardiopulmonary bypass. After pericardial decortication of both left- and right-sided chambers, progressive right-sided chamber distension was noted. An intra-operative TEE was performed to evaluate the interatrial septum. This clearly demonstrated a secundum ASD with colour flow indicating a left-to-right shunt at atrial level. Aorto-bicaval cannulation was performed and cold crystalloid antegrade cardioplegia was administered through the root of the cross-clamped ascending aorta to achieve cardiac arrest. Total cardiopulmonary bypass with a membrane oxygenator was established. The ASD (2.5cm x 1.5cm) was closed primarily through a right atriotomy. Reperfusion and weaning from bypass was accomplished uneventfully. The post-operative course proceeded smoothly with marked hemodynamic improvement. The patient was discharged home on the 10th post-operative day and remains asymptomatic after 23 months’ follow-up. Histology of the excised pericardium showed a thickened pericardium with extensive fibrosis and infiltration by chronic inflammatory cells extending into the adjacent fatty tissue. No evidence of tuberculosis or specific inflammation was found.

DISCUSSION

The tortuous route to complete diagnosis in this patient highlights William Osler’s observation in 1892 - “Probably no serious disease is so frequently overlooked by the practitioner. Post-mortem experience shows how often pericarditis is not recognized or goes on to resolution and adhesion without attracting notice”.

The combination of ASD and chronic constrictive pericarditis (CP) is very rare. Diagnosis of this clinical entity is compounded by the fact that the features of the ASD may be masked by those of CP. Tanaka et al report a case of recurrent CP and ASD in which the ASD was missed at the time of the first pericardiectomy. In our patient, although the initial working diagnosis was CP, the possibility of coexistence with ASD was not considered even after the index echocardiography.

It is important to make the diagnosis of CP because surgical treatment is often curative. Conversely, without treatment, there is progressive hemodynamic and physical deterioration with a reduced life expectancy.

The presence of pericardial thickening on imaging studies does not necessarily indicate constrictive physiology. Conversely, patients with CP may not have pericardial thickening on imaging studies or even histologically following pericardiectomy.

Non-invasive imaging modalities such as chest x-ray examination, echocardiography, computerized tomography (CT) scans and magnetic resonance imaging (MRI) are often used alone or in combination to demonstrate pericardial thickening in CP. Cardiac catheterization may confirm a suspicion of constrictive physiology but it is invasive and not often used unless the diagnosis remains in doubt after non-invasive methods are exhausted.

As in the case reported, echocardiography does not reliably detect pericardial thickening but it has gained increased usefulness in the evaluation of CP based on information provided by Hatle and colleagues who reported dynamic

Figure 1

Figure 1. Pre-operative TEE showing the ASD.
changes with respiration occurring in patients with CP. These changes - the dissociation of intrathoracic and intracardiac pressures as well as enhanced ventricular interaction, can be demonstrated echocardiographically and increases the diagnostic yield in cases of CP.

As challenging as the diagnosis of CP is when it exists alone, coexistence with an ASD can prove clinically misleading; sometimes, a complete diagnosis is made only at autopsy. In our patient, the clinical features were arguably dominated by those of CP but this was overlooked when echocardiography failed to demonstrate pericardial thickening and suggested a diagnosis that was thought to account for the clinical features of right-sided heart failure. There was no credible explanation for a supposedly large ASD without Doppler evidence shunting across the defect. A complete catheterization was not done in this patient due to equipment problems but the right heart catheterization was not suggestive of a left-to-right atrial level shunt. In retrospect, we postulated that this was so because of possible equalization of left and right atrial pressures imposed by the constrictive physiology; intra-operative TEE showed a large left-to-right shunt across the ASD after surgical relief of the constriction.

The cause of this unusual combination of lesions, if indeed there is a common etiological factor, remains unknown. It is probable that the coexistence of these lesions is purely coincidental.

CONCLUSION

It is extremely important to recognize this association of lesions during life because both are amenable to surgical correction. The diagnosis of CP requires a high index of suspicion and a multi-modality approach to clinical investigation. Echocardiographic evaluation of CP must not be limited to assessment of pericardial thickening but should include assessment of dynamic changes in cardiac physiology with respiration. Clinicians should bear in mind that the coexistence of CP and ASD may give rise to no demonstrable shunt echocardiographically across the ASD.

References

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