

Chylous Ascites caused by Lutembacher's Syndrome: A Case Report and Review of the Literature

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ABSTRACT

Chylous ascites is a well-documented sequelae of traumatic rupture of the thoracic duct and mechanical obstruction of the lymphatic system due to neoplastic, inflammatory, or congenital anomalies. Less commonly, chylous ascites results from altered hemodynamics and lymphatic flow, as seen in constrictive pericarditis and heart failure. We report a case of chylous ascites due to severe mitral stenosis plus atrial septal defect known as Lutembacher's syndrome. From our knowledge, this has never been reported before. The chylous ascites disappeared after repairing the heart. The pathophysiology of chylous ascites formation in this clinical syndrome will be discussed, with a review of the literature.

Keywords: Atrial septal defect and mitral stenosis; chyloperitoneum; chylous ascites; Lutembacher's syndrome

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Chylous ascites may be the result of many pathological conditions, including congenital defects of the lymphatic system, nonspecific bacterial, parasitic and tuberculous peritoneal infection, liver cirrhosis, malignant neoplasm, blunt abdominal trauma and surgical injury. The most common etiological factors are abdominal malignancy and congenital lymphatic abnormalities in adults and children, respectively.¹ Constrictive pericarditis, dilated cardiomyopathy, and ischaemic heart disease have sometimes been associated with chylous ascites.²⁻⁴

The association of congenital atrial septal defect and rheumatic mitral stenosis, customarily referred to as Lutembacher's syndrome, is a recognized pathophysiologic entity.⁵⁻⁶ To our knowledge, this is the first reported case of Lutembacher's syndrome presented with heart failure and chylous ascites.

CASE REPORT

A 17 year old woman was admitted to our unit with the symptoms of heart failure for three years and progressive abdominal distension for three months. The physical examination showed a cachectic young lady, marked ascites. Her blood pressure was 100/60 mmHg, pulse 84 beats/min and regular, mild dyspnea. She got very high jugular

venous pulse with apex beat at 6th intercostal space on the anterior axillary line. On auscultation, there was grade 4/6 diastolic rumbling murmur, increased first heart sound at apex, and grade 3/6 systolic murmur along the left lower parasternal border.

Blood laboratory studies were normal. Serum total protein was 6.5 g/dl with an albumin concentration of 3.8 g/dl. Chest radiography showed moderate cardiomegaly and pulmonary congestion. (Fig 1) Echocardiogram revealed atrial septal defect (2x2.5 cm²), severe mitral stenosis (0.52 cm²), and severe tricuspid regurgitation. The hemodynamic data from cardiac catheterization showed severe pulmonary hypertension (74/35 mmHg), high left and right atrial pressure (28 and 23 mmHg respectively) and 3.87:1 left to right shunt. Computerized tomography showed massive intraperitoneal effusion, no abnormal masses or enlarged lymph nodes. Paracentesis yielded a milky fluid with the following biochemical composition: triglyceride 634 mg/dl, cholesterol 75 mg/dl, total protein 5.3 g/dl, albumin 3 g/dl. The cell count of the fluid was 1,500 (95% lymphocyte). Cytology and cultures, including mycobacterial, from the peritoneal fluid were negative.

The patient underwent successful open mitral valvulotomy, atrial septal defect closure and tricuspid valve annuloplasty. She recovered uneventfully on a medium chain triglyceride diet for 6 months. Her ascites gradually resolved over a month. (Fig 2,3) She is still in functional class 1 on her last follow up, (6 year postoperatively).

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Fig 1. Chest radiography showed moderate cardiomegaly and pulmonary congestion.

DISCUSSION

Since 1813, the association of atrial septal defect with mitral stenosis has been described many times. Lutembacher^{5,6} drew special attention to the condition in



Fig 2. Day 3 post-operative.

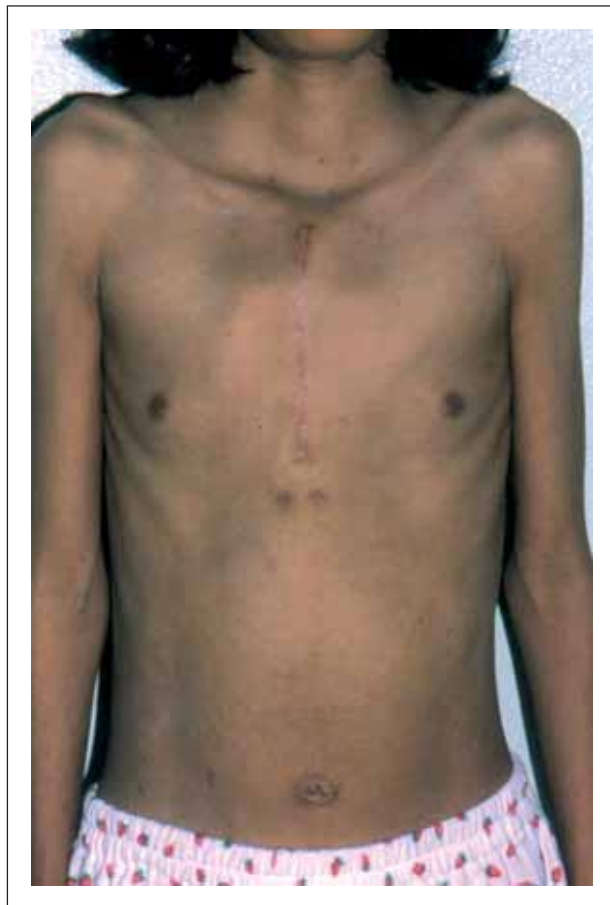


Fig 3. Week 6 post-operative.

1916 and described it as an anatomic and pathophysiologic entity. Because of the misunderstanding of the mid-diastolic murmur found in atrial septal defect, Lutembacher's syndrome was previously diagnosed with great frequency. Nadas and Alimurung⁷ were the first to point out the rarity of this syndrome and to emphasize the tricuspid origin of the diastolic flow murmur in the atrial septal defect. This syndrome is the combination of two unrelated diseases, the congenital secundum atrial septal defect and the acquired rheumatic mitral stenosis. The clinical features⁵ suggestive of this syndrome are: (1) atypical auscultatory findings of rheumatic mitral stenosis: no presystolic accentuation of the diastolic rumble; (2) the presence, quite commonly, of a systolic murmur accompanied by a palpable thrill in the pulmonary area; and (3) cyanosis of slight to moderate degree (not seen in the absence of heart failure). Radiologically, supportive evidences of the diagnosis are: (1) the large size of the pulmonary main trunk and its branches, with (2) vigorous pulsations; in association with (3) a mitral or mitral-tricuspid cardiac contour, and (4) slight or absent left atrial enlargement. Finally, electrocardiographic datas suggestive of this diagnosis are: (1) the presence of RBBB and (2) commonly, qR type complexes in Lead V1 with (3) concomitant left atrial enlargement ("mitral" P waves). Outstanding manifestation in this clinical syndrome is early severe pulmonary hypertension, (accentuated effects from increased pulmonary venous pressure, so increased left to right shunt).

Chylous ascites caused by heart disease has been reported sporadically²⁻⁴. This is the first reported case of chylous ascites caused by Lutembacher's syndrome. The postulated mechanism of chylous ascites produced by this

syndrome may be from: (1) High venous pressure increases the abdominal lymph production secondary to an augmented capillary filtration. The lymph flow of the thoracic duct can increase by up to 12 fold the normal rate, but the stiffness of the veno-lymphatic junction in the neck limits lymphatic flow⁸. (2) High pressure in the left subclavian vein reduces lymphatic drainage. As a result of the restricted lymphatic drainage, lymphatic venous collaterals form but cannot handle the normal lymph flow. The chylous fluid leaks into the peritoneal cavity as a result of the rupture of dilated intestinal lacteals.

This form of chylous ascites (caused by heart disease) is easily treated by correction of the heart problem.²⁻⁴ As in our patient, when the central venous pressure decreased by repairing the defect in the heart, the ascites gradually improved spontaneously without any direct surgical ligation on the leakage point.

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