Congenital Isolated Pleural Effusion Associated with Obstructive Uropathy

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Fetal abnormalities are increasingly being discovered as routine ultrasound examinations become part of the normal pregnancy follow-up. Fetal pleural effusion may be one of these findings and is usually associated with hydrops fetalis. However, isolated pleural effusion is a rare anomaly with an estimated prevalence of 1 in 10,000–15,000 pregnancies. Most of the cases of IPE are due to chylothorax. Other etiologies for IPE are extremely rare [1]. We report a case of a newborn infant with congenital IPE caused by urinary tract obstruction.

Patient Description

A 2,910 g male was born by cesarean section at 39 weeks gestation. The 39 year old mother had a normal pregnancy follow-up, and routine ultrasound examination at 22 weeks of gestation was normal. Two days before delivery a repeat ultrasound demonstrated a massive right hydrothorax with mediastinal shift to the left, bilateral hydronephrosis (most prominent on the right side), an enlarged bladder, and oligohydramnios. An ultrasound performed in the operating room confirmed the diagnosis of right hydrothorax and a right pleural tap was immediately performed, draining 200 ml of clear fluid. The infant was transferred to the neonatal intensive care unit.

Physical examination on admission revealed a right-sided abdominal mass and a suprapubic mass. Complete blood count on admission was in the normal range. Blood tests at the age of 24 hours revealed abnormal renal function, with creatinine 1.58 mg/dl and urea 21 mg/dl. Both mother and infant's blood types were O+ and the direct Coombs test was negative. Immunoglobulin M antibodies for adenovirus, Epstein-Barr virus, cytomegalovirus and Parvovirus B-19 were all negative. Thyroid function tests were normal. Echocardiographic examination on admission demonstrated normal heart function without pericardial effusion. The chest drain was removed after 13 days with resolution of the pleural effusion.

Pleural fluid analysis revealed clear fluid containing 200 cells/mm^3 (neutrophils 8%, lymphocytes 12%, mesothelial cells 80%). The fluid was confirmed to be transudate (lactate dehydrogenase 199 U/L, protein 3 g/L). The cytologic examination was negative. The fluid was negative for chylomicrons in two separate examinations after the initiation of enteral feeds. Pleural fluid cultures were negative for bacteria, viruses

IPE = isolated pleural effusion

Table. Comparison of urine, pleural fluid and plasma electrolytes

	Urine	Pleural fluid	Plasma
Sodium (mmol/L)	32	120	124
Potassium (mmol/L)	8.0	5.3	5.0
Urea (mg/dl)	191.9	79.0	65.0
Creatinine (mg/dl)	15	4.9	3.2

and fungal organisms. Simultaneous electrolyte analysis of plasma, urine and pleural fluid were performed on the third day of life, showing similar plasma and pleural fluid sodium and potassium concentrations, but creatinine and urea levels in the pleural fluid were intermediate to those found in serum and urine [Table 1]. A urinary tract sonography was performed shortly after delivery and demonstrated massive right hydronephrosis, right hydroureter and mild left hydronephrosis. A cystic mass was noted near the right kidney. The bladder was distended with thickened walls. A urinary catheter was inserted as posterior urethral valves were suspected. The results of renal function tests, which were abnormal at birth, continued to deteriorate to creatinine 3.2 mg/dl and urea 65 mg/dl at the age of 4 days. The infant was oliguric and was treated with repeated doses of fresh frozen plasma, furosemide and low dose dopamine. A cystography performed after a few days of adequate urinary drainage demonstrated PUV without vesico-ureteric reflux. Abdominal computed tomography scan revealed a right retroperitoneal cystic mass suggesting the presence of a perirenal urinoma. Aspiration of the mass revealed a cloudy fluid. A temporary vesicostomy and subsequent ablation of the PUV were performed. Renal function gradually improved and at discharge serum creatinine and urea concentrations were 0.34 and 45 mg/dl respectively. Resolution of the urinoma was demonstrated on repeated ultrasound examination. One year later the infant was developing well and his renal functions tests were in the normal range.

Comment

Congenital pleural effusion is usually associated with hydrops fetalis. However, most cases of congenital IPE are due to chylous leak from the thoracic duct, causing chylothorax [1]. Non-chylous congenital IPE is a very rare condition. Etiologies for the anomaly are adenovirus infection, congenital goiter, and pulmonary anomalies like diaphragmatic hernia. Cardiac anomalies and trisomy 21 have been reported in association with congenital IPE [1].

Diagnosis of urinothorax is confirmed when chemical analysis of pleural fluid discloses a higher creatinine concentration than found in simultaneously obtained serum samples. This method for diagnosing urinothorax was described by Stark et al. [2], who reported three patients with obstructive uropathy and retroperitoneal urine collections and who developed pleural effusions. The diagnosis of urinothorax in these patients was confirmed when chemical analysis of pleural fluid showed higher creatinine concentrations than found in simultaneously obtained serum samples. Similar determinations made in 71 specimens from control patients with pleural effusions of non-urologic causes demonstrated that pleural creatinine concentration did not exceed the serum level [2].

In a literature search of Medline, we found only two cases of congenital urinothorax. Freidland and colleagues [3] described the first case in 1971. Another case was reported by Lee [4], who described a male newborn with a massive left congenital pleural effusion and a same-sided hydronephrosis caused by obstructing PUV. The effusion disappeared within a few days of adequate urinary drainage. The limitation of Lee's report is its purely descriptive nature of concomitant pleural effusion and PUV, without analysis of the pleural fluid and plasma creatinine ratio, which are the required laboratory tests for diagnosis.

We concluded that the pleural effusion in our patient was of urinary origin. This conclusion was based on: a) the combination of right pleural effusion and ipsilateral urinoma, b) pleural fluid/serum creatinine ratio greater than 1, and c) complete resolution of the pleural effusion after adequate drainage of the obstructed urinary system. It is possible, however, that the direct pleural drainage contributed to the disappearance of the pleural effusion

We hypothesize that urinoma was the result of high pressure obstruction of the urinary tract caused by the PUV. This sequel provides a pressure "pop-off" mechanism that resulted in preservation of better renal function [5]. Urine may reach the pleural cavity either by lymphatic drainage or by direct, rupture-mediated leakage into the pleural cavity. The collected urine equilibrated with the serum as a dialysate. The different rate of equilibration between small molecules (sodium, potassium) and large molecules (creatinine and urea) could explain the difference of the pleural to serum ratio of those molecules.

We conclude that urinothorax should be suspected in any case of congenital pleural effusion accompanied by obstructive uropathy and hydronephrosis. Recognition of this rare condition may avert the need for unnecessary evaluation, and prompt urinary drainage may well resolve the problem.

PUV = posterior urethral valves

References

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