Chylothorax with Down Syndrome: Unusual Case Report

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Abstract

Trisomy 21 or Down syndrome (DS) is the most common chromosome aberration in the newborn. Its high incidence has led to better understanding of congenital anomalies and diseases in association with this syndrome. The presence of protein, triglycerides, and lymphocytes in the pleural effusion is diagnostic of chylothorax. Lymph vessel abnormalities or a leaky thoracic duct are thought to be origin of congenital chylothorax, which is reported many times in the literature. But, there is no information in the English literature about 1-year-old baby with DS who is complicated with chylothorax. Aplasia of the twelfth rib is rare but found to be more common in DS than in normals. We report on a 1-year-old girl with DS refered to our clinic with signs and symptoms of respiratory distress. Diagnosis and treatment of chylothorax, pleural emphyema, and pericardial effusion were obtained in algorithm.

Keywords: Chylothorax, Down Syndrome.

Introduction

Trisomy 21 may present with congenital chylothorax giving rise to symptoms of respiratory distress. To the best of our knowledge, there is no information in English literature about 1-year-old baby with DS who is complicated with chylothorax. We report on a case with trisomy21 and complicated cyhlothorax.

Case Report

One-year-old girl had admitted to a local hospital with fever and respiratory findings such as tachypnoea and dry cough for one week duration. She was refered to our emergency room after bilaterally chest tube insertion. She was first evaluated in the emergency room.

There was no history of trauma, surgery, and malignancy. On admission, her body temperature was 37.7°C, pulse rate 128/min and respiratory rate 36/min.

Poor breath sounds were noted in auscultation.

She had atypical facies with low-ser ears, epicanthic folds, high-arched palate and short fingers. Initial laboratory findings were as follows: Hb 10.1 g/dl, haematocrit 28.5%, WBC 26300/mm³, platelets 261000/mm³, total protein 4.4mg/dl, glucose 84mg/dl, LDH 1142 U/L, triglyceride 174 mg/dl, cholesterol 79 mg/dl.

She had bilaterally tube thoracostomy. Left chest tube drainage was 150cc and dirty-brown colored. Right sided chest tube drainage was 200cc and milky. Biochemical analysis of the left-sided pleural effusion was as follows: glucose 4mg/dl, protein 3.6mg/dl and LDH 5198 U/L. There was no bacteria on the gram stain.

Left-sided pleural effusion was diagnosed as pleural emphyema.

Biochemical analysis of the right-sided pleural effusion was as follows: glucose 40mg/dl, LDH 607 U/L, protein 2.3 mg/dl, triglyceride 266 mg/dl, cholesterol 32 mg/dl.

There was no bacteria on the gram stain. Right-sided pleural effusion was diagnosed as chylothorax.

Antero-posterior plain chest graphy showed aeration defect (pleural effusion) in the left hemithorax, mediastinal shift into right side, aplasia of the twelfth rib and bilaterally chest tubes. (figure 1).

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Chylothorax is the leakage of chyle from the thoracic duct or its branches into the pleural space secondary to obstruction or disruption of the thoracic duct. Neonatal spontaneous chylothorax may be idiopathic, but often accompanies congenital heart diseases. There was no congenital heart disease in our present case. Lymph vessel abnormalities or a leaky thoracic duct are thought to be origin of spontaneous chylothorax.

The prevalence is approximately 1 in 2000 infants admitted to neonatal intensive care units, and frequently associated with genetic anomalies such as trisomy 21, monosomy X, and Noonan syndrome, all with frequent lymph vessel abnormalities. This child was diagnosed as DS, and her spontaneous unilateral chylothorax might be related with genetic disease. Other causes of chylothorax include interruption of the thoracic duct by trauma or surgery and intrathoracic malignancies.

The ideal management of the patients with chylothorax is unknown. Modalities used in treatment of chylothorax are as follows: Conservative (nothing by mouth, medium-chain triglycerides, central hyperalimentation, drainage of pleural space (thoracentesis, closed chest tube thoracotomy), complete expansion of lung), Operative (direct ligation of thoracic duct, mass ligation of thoracic duct tissue, pleuroperitoneal shunting, pleurectomy, fibrin glue), and
radiotherapy. Closed chest tube drainage and central hyperalimentation provided successful management for present case.

Willich E. et al reported that abnormal ossification of the manubrium (33%), aplasia of the twelfth rib (18%), high lumbar vertebral bodies (50%), brachymesophalangia of the fifth ray (62%) are found to be more common in DS. Our present case had also aplasia of right-sided twelfth rib, but it is not required any management.

Conclusions

Spontaneous chylothorax may be associated with DS. Pleural effusion with DS must be promptly diagnosed and aspirated. Physicians should take into consideration DS patients with onset of shortness of breath, dry cough for pleural effusion. Physical, biochemical and radiological examinations have to be obtained for diagnosing.

References