Congenital bronchobiliary fistula

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Introduction

Congenital Broncho biliary fistula is a rare developmental abnormality with an abnormal fistula between the respiratory system and biliary tract. Patients with this anomaly may develop symptoms at any time from neonate age to adulthood, but most of them present with signs of bilious saliva, and choking, within a few days after birth. Treatment is by surgical resection of the fistula. Here we discuss a child with this rare anomaly who became symptomatic immediately after birth.

Case

A full term, 8-day old male baby was referred to our hospital with a history of respiratory distress since birth. The child had required ventilation shortly after birth and had failed efforts for extubation twice.

On arrival to our hospital, we noticed that the endotracheal tube aspirates appeared bilious.

We went ahead with a bronchoscopy which showed a fistulous opening in the right bronchus. Bile was seen coming out of it.

With a possibility of a bronchobiliary fistula, we went ahead and did an MRI, which demonstrated a fistulous tract between the right bronchus and extra hepatic biliary duct thus confirming the diagnosis.

The Child was taken up for surgery the next day. On lateral thoracotomy a large fistulous tract opening into the right bronchus was seen which was confirmed with intra – operative bronchoscopy. The tract was proximaly ligated and contrast injected into its distal part which demonstrated a fistulous opening into the left hepatic duct. (Figure 1) The complete tract was resected.

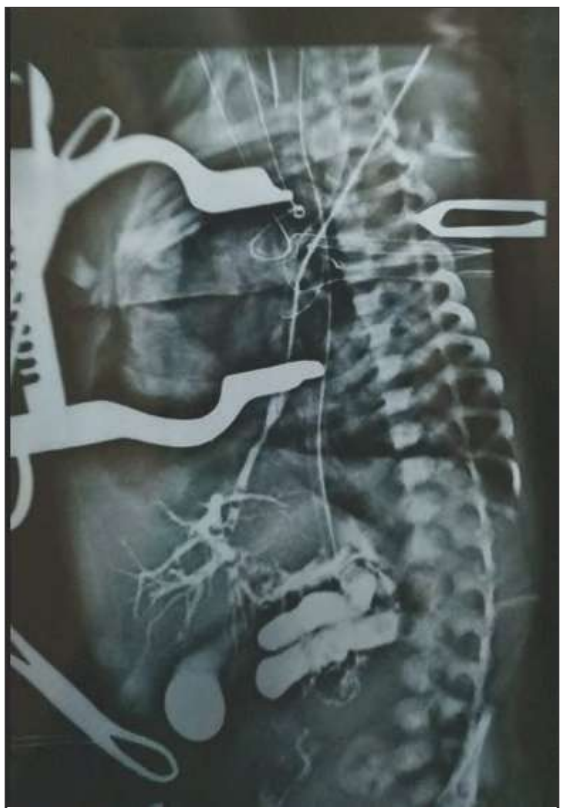

Figure 1. Intra – operative fistulogram demonstrating bronchobiliary communication.

Post – operative period was uneventful. Child was discharged after 7 days and is doing well on follow – up.

Discussion

Congenital broncho – biliary fistula is rare and less than 50 cases have been reported in

literature. [1] The onset time and severity of symptoms are related to the diameter of the fistula; therefore, the symptoms can appear at any age from newborn to adulthood. However, most of them occur in newborns and infants. The main clinical features of CBBF are recurrent coughing, bilious sputum, or bile-stained sputum in tracheal intubation. Patients generally develop progressive dyspnea, cyanosis, severe pneumonia, respiratory distress syndrome, and even apnea; shortly after birth and mechanical ventilation is often needed as in our child. It has been often misdiagnosed as an esophagotracheal fistula, gastroesophageal reflux, aspiration pneumonia, or high intestinal obstruction [2,3] Bile-stained expectoration is the most typical sign among the aforementioned symptoms and helps in differentiating from the other causes.

The pathogenesis of this disease is not clear. Abnormal development of the bronchial bud, which fuses with the bile duct, is regarded as the cause of the anomaly

In literature it has often been associated with biliary atresia, diaphragmatic hernia, esophagus atresia, or tracheoesophageal fistula. Around 30% have been found to have a co-existing biliary malformation [1,4] however our child did not have any of these associated anomalies.

Bronchoscopy is often used for making a diagnosis, as it can be used to find the abnormal opening of the fistula with bile flowing via the opening. Magnetic resonance imaging and 3D-CT reconstruction can provide valuable clues to the diagnosis. [5]

Surgical resection is the ultimate treatment of choice and in an uncomplicated case like ours, once the fistula is separated, it is ligated and sutured at the upper end and at the hiatus and then removed.

To conclude, congenital bronchobiliary fistula is rare, but its clinical symptom i.e the presence of bile – stained sputum (biliptysis) is typical of this condition and points towards the diagnosis. Once it is considered as a possibility, its diagnosis and treatment are not difficult, and the prognosis is excellent.

References

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