affecting infants and young children (under 8 years of age). It consists of mature and immature adipose tissue in variable proportions. The more immature forms are made up of primitive mesenchymal cells, lipoblasts, and capillaries in a myxoid background. The more mature forms (more frequent in older children) are very much like lipoma. Lipoblastoma shows consistent abnormalities of chromosome 8q11-13, resulting in rearrangement of the PLAG-1 gene.

Liposarcoma in children is extremely rare (mostly in the 10 to 15 year age group) and most often myxoid in type. In the most differentiated lesions, a population of small spindle cells and small lipoblasts (predominantly univacuolated) is set in a myxoid matrix containing numerous thin-walled capillaries. Diagnostic multivacuolated lipoblasts are sometimes only found in the peripheral subcapsular parts of the tumor. Cytogenetically, myxoid liposarcoma is characterized by the recurrent translocation t(12;16)(q13;p11) or rarely, by the variant t(12;22)(q13;q12).

Intramuscular lipoma is a variant of lipoma, most often poorly circumscribed and infiltrative. It usually occurs in adult patients as a slow-growing mass. The most common cytogenetic abnormalities are aberrations involving 12q13-15, loss of material from 13q, and aberrations involving 6p21-23.

In our case, the typical clinical settings (young age, diffuseness of the process, and rapid growth) were all clues to the diagnosis of DL, which was consistent with the microscopic picture (poorly circumscribed, fully mature adipose tissue) and the absence of cytogenetic aberrations.

Strict follow-up was warranted because of the frequency of relapse reported in the literature. After 1 year, the patient is in good clinical condition, with no signs of recurrence. Moreover, the radical surgical approach achieved good aesthetic results while sparing the mammary gland (Fig 4).

Although rare, DL should be considered in the differential diagnosis when a rapidly growing lipomatous mass is found in a neonate as early surgical treatment can be crucial to obtaining complete removal, thus reducing the risk of recurrence and its associated morbidity.

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References


Congenital Tracheobiliary Fistula in an Adolescent Patient

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Congenital tracheobiliary fistula is a rare malformation that allows communication between the respiratory system and hepatobiliary tract. We describe a male adolescent patient who was admitted with a destroyed lung caused by repetitive bile pneumonitis with a congenital tracheobiliary fistula. Left pneumonectomy was performed, and the fistula tract was successfully divided.

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Congenital tracheobiliary fistula (TBF) is a rare anomaly in which there is an abnormal communication between the trachea and the biliary tract. The presence of respiratory symptoms from early infancy means that congenital TBF is usually diagnosed during the neonatal period. Herein we describe an 18-year-old man with a destroyed left lung caused by recurrent pneumonia. The cause of the recurrent infection was bile pneumonitis originating from congenital TBF.

An 18-year-old man with no history of trauma or abdominal operation presented with cough and greenish sputum. When he was 6 months old he had been admitted to an intensive care unit for bronchiolitis. He had experienced repeated admission because of bronchiectasis with recurrent pneumonia up to the current presentation. His height was 155 cm, and his body weight was 38.7 kg; the recurrent infection had kept him from reaching the standard national average weight and height. Laboratory tests showed no elevation in serum total and directed bilirubin levels (0.3 and 0.1 mg/dL, respectively). His levels of γ-glutamyltransferase, alkaline phosphatase, aspartate transaminase, and alanine transaminase were within normal limits. A chest roentgenogram showed a totally collapsed left lung and an air column between the bifurcated bronchi (Fig 1). Coronal and volume-rendered chest computed tomography images identified an accessory bronchus from the carina to the liver and the total destruction of the left lung, with cystic bronchiectasis on the right middle lobe (Fig 2). Bronchoscopy demonstrated bilious secretion from an anomalous orifice between the right and left main bronchi, and stenosis in the middle fistula tract (Fig 3). Endoscopic retrograde cholangiopancreatography revealed no evidence of biliary tract obstruction or other anomalies. A technetium-99m diisopropyliminodiacetic acid hepatobiliary scan showed increased multifocal radioactivities in the right lung.

A double-lumen endotracheal tube was inserted, and a bronchial balloon was positioned in the right main bronchus, with the accessory bronchus and the left main...
bronchus being obstructed during the operation. A left thoracotomy was performed from the fourth intercostal space and the fistula tract originating from the carina. The fistula extended downward, burrowing between the aorta and the left atrium. Respiratory acidosis developed during dissection of the left main bronchus and the accessory bronchus as a result of the dislocation of the bronchial balloon of the endotracheal tube. This was relieved by clamping the accessory bronchus and the left main bronchus, which allowed continuation of the left pneumonectomy. The proximal TBF was doubly ligated and divided. Histopathologic examination of the left lung revealed diffuse interstitial fibrosis and a cavitary lesion with bile pigmented material. A postoperative bronchoscopy demonstrated a completely obstructed fistula, and the patient had an uncomplicated postoperative course.

Comment

Congenital TBF is a rare developmental malformation. Most cases have been detected in the neonatal period because of early-onset respiratory distress symptoms or intractable pulmonary infection [1–3], with adolescent and adult patients rarely being reported [4]. In contrast to previous reports [1–3], our patient had survived until adolescence without severe respiratory distress; only one case of congenital TBF involving an adult patient has been reported [4]. Gugenheim and colleagues [5] reported 16 cases of nontraumatic bronchobiliary fistula in adult patients, but they were caused either by biliary obstruction after surgical procedures or by infectious disease, and our patient could have a diagnosis of congenital TBF because he had no history of trauma or hepatobiliary operation.

The reason why our patient was able to survive until adolescence is unclear, but this could have been a result of the stenosis in the middle fistula—as revealed by chest computed tomography and bronchoscopic examinations—preventing bile regurgitation into the lung. However, his recurrent pneumonia had progressively destroyed the lung parenchyma and increased the incidence of coughing with sputum production. Elevated abdominal pressure during coughing could aggravate bile regurgitation.

Two embryologic theories of TBF development have been suggested [1]: (1) union of an anomalous bronchial bud with an anomalous bile duct and (2) duplication of the upper gastrointestinal tract. Several authors [2, 6] have supported the first theory because they found a ciliated pseudostratified epithelium and cartilage surrounded by mucinous glands in their surgical specimens of congenital TBF. Our bronchoscopic findings of weblike stenosis in the midportion of the fistula also supported the first theory of an anomalous union of the bronchial bud with the bile duct.

Surgical repair is the only appropriate treatment, and a good surgical outcome can be expected if patients have no associated biliary malformation. The reported incidence of a coexisting anomaly of the common bile duct is 36.8% [3]. The fistula is the only outlet for bile in patients with accompanying common bile duct anomaly, which is associated with earlier presentation, typically during the neonatal period [2]. Therefore, associated biliary malformation should be suspected in every case of TBF. The usual method of surgical treatment in congenital TBF is excision of the fistula through a right thoracotomy. We performed a left pneumonectomy with the approach from the left side because the left lung had been totally destroyed by the longstanding recurrent infection.

In conclusion, we observed and treated a rare congenital anomaly—TBF in an adolescent patient—that caused total destruction of the lung. Our case indicated that a long-standing congenital TBF can result in the lung being destroyed. Therefore, early diagnosis is important for saving the lung. In cases of recurrent pulmonary infection, the physician should consider congenital anomalies of the bronchial tree, even in older patients.

References

Thoracoscopic Esophageal Repair of a Spontaneous Barrett’s Ulcer Perforation

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Spontaneous esophageal perforation of a Barrett’s ulcer is a rare condition that is associated with high morbidity and mortality. It occurs as a result of a missed diagnosis of underlying Barrett’s esophagus or because of unresponsiveness to medical management. Owing to the life-threatening nature of this disease, emergency surgical intervention is indicated. Here, we report the first case of spontaneous Barrett’s esophageal perforation that has been managed using a thoracoscopic approach. This case highlights the feasibility of minimally invasive methodology for emergent esophageal disorders. It also demonstrates the altered esophageal wall properties of Barrett’s esophagus through musculofibrous thickening and muscularis mucosae duplication that may contribute to the technical outcomes of esophageal surgical management.


Spontaneous esophageal perforation of a Barrett’s ulcer is a rare condition associated with high morbidity and mortality [1]. It can result from resistance to medical therapy or a missed diagnosis of a Barrett’s ulcer, and is a surgical emergency that requires immediate intervention. Here, we describe the first case of spontaneous Barrett’s esophageal perforation that was managed using a thoracoscopic approach.

A 67-year-old man presented to our institution 2 hours after sudden onset of severe dyspnea and abdominal pain. He was referred from a regional psychiatric institute because of schizophrenia. There was no hemothysis, dysphagia, or history of vomiting. Abdominal examination was unremarkable, although chest auscultation and palpation revealed a dullness and lack of air entry to the right thoracic base. There was no emphysema or Hamman’s crunch. Computed tomography scan (Fig 1) demonstrated periesophageal debris within the right hemithorax, which led to the diagnosis of esophageal perforation.

Urgent esophagogastroscoopy confirmed the diagnosis of esophageal perforation and the presence of red mucosa “tonguelike” lower esophageal projections that raised the suspicion of Barrett’s disease. The decision was made for emergency surgical management using a minimally invasive thoracoscopic approach. The patient was intubated with a double-lumen endotracheal tube and placed in the prone position. The right side of the chest was insufflated with low-pressure CO$_2$ (9 mm Hg). The operation was performed on the right side of the patient in a plane parallel to the view of the camera. The mediastinal pleura overlying the esophagus was opened from the level of the ayzyous vein to the crus. The presence of slough and pus was noted and washed out. The pleura was thickened, and the perforation site at the lower esophagus was confirmed after mobilization to reveal a 3-cm longitudinal defect. In view of perforation dimension, a decision for surgical repair was made.

A nasogastric tube was utilized to verify the true afferent and efferent esophageal lumens, which were subsequently both sited with long limbs of a T-tube. Thoracoscopic closure of the perforation (Fig 2) was performed over the T-tube with interrupted 3-0 polyglactin (Vicryl; Ethicon, Somerville, NJ) sutures. It was noted that the suspected Barrett’s tissue was thicker and demonstrated more tensile strength than the typical friable tissue observed in Boerhaave’s syndrome esophageal perforations. The procedure was concluded with thoracic washout and the placement of two drains, followed by the laparoscopic insertion of a feeding jejunostomy. Postoperatively, the diagnosis of Barrett’s ulcer perforation was confirmed on histology (Fig 3). The patient’s esophageal defect had successfully healed by 8 weeks, with full recovery (after insertion and removal of T-tube). His psychiatric status was, however, associated...