Bronchopulmonary Sequestration

22

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22.1 Introduction

Bronchopulmonary sequestration (BPS) is a rare developmental disorder in which part of the lung is disconnected from the main airway and receives aberrant arterial inflow from the aorta instead of from the pulmonary arteries. BPS belongs to the group of foregut malformations and is often accompanied by other defects. The incidence of BPS has increased considerably in the last few years due to the widespread routine use of fetal ultrasound screening. Several patterns of clinical expression are possible and therapeutic attitudes ranging from expectant observation to early surgical removal or embolization remain controversial.

22.2 Pathology and Classification

The sequestered lung tissue has bronchial and bronchiolar ducts, areas of collapsed airspaces and occasionally cysts lined by pseudostratified cilliary epithelium with mucus-secreting components similar to those found in congenital cystic adenomatoid malformations (CCAMs). The coexistence of BPS with CCAMs was highlighted after observation of a relatively large number of cases diagnosed prenatally as CCAMs that had both features. Such "hybrid" tissues [1] were also described by other authors [2–4] and confirmed by pathological microdissections of surgical specimens [5]. These studies showed also that bronchial atresia is a relatively common finding in BPS along with other foregut malformations such as CCAMs and lobar emphysema [6]. The aberrant arteries of the sequestered tissue are rich in elastic fibers that are consistent with their systemic origin (Fig. 22.1). Atherosclerosis, even in young individuals, can be occasionally observed in the wall [7].

The recent finding of abnormal expression of mucinogenic factors such as MUC5AC, cytokeratin 20 and human v-erb-b2 erithroblastic leukemia viral oncogene homologue 2 (HER)2 as well as of K-RAS mutations within secretory areas of CCAMs and BPS called particular attention to the risk of cancer in these tissues [8].

Pryce et al. [9, 10] distinguished two types of BPS:

- intralobar bronchopulmonary sequestration (IBPS), in which the malformed tissue is a part on one lobe of the lung surrounded by lobar pleura;
- extralobar bronchopulmonary sequestration (EBPS), in which there is a clear sepa-

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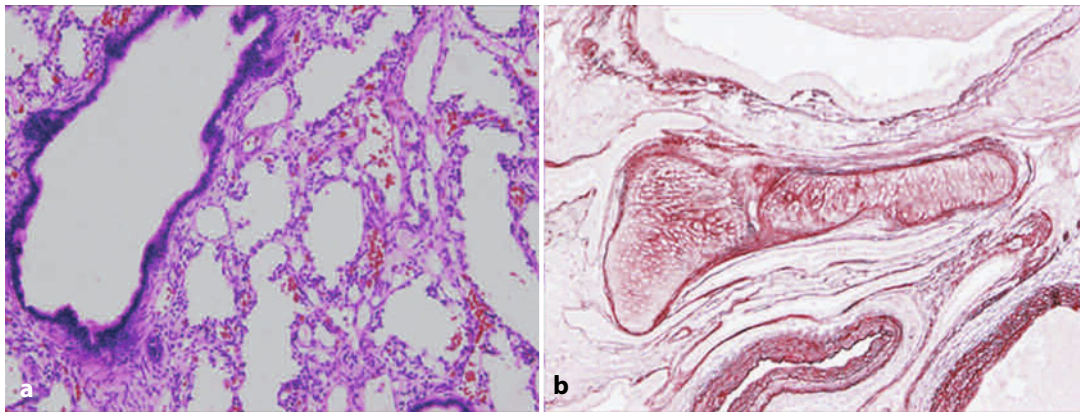

Fig. 22.1 a H&E staining of a section of an IBPS specimen shows disorganized airspaces with secretory bronchial-like cysts lined by a ciliated mucus-secreting epithelium. In **b**, stained for elastic fibres, systemic arteries with abundant elastic elements are seen below a cartilaginous island

ration between the normal lung and the sequestered tissue, including often a complete fissure and individual pleural covering.

Very seldom, as proof of its foregut origin, the malformed lung parenchyme (intralobar or extralobar) is connected with the esophagus or the stomach through a more or less well-formed bronchus.

BPSs are more often located in the lower lobes or below them. In these cases, arterial inflow originates from the thoracic or abdominal aorta and reaches the malformed tissue directly or by traversing the diaphragm. The pattern of venous return may be normal into the left atrium (as in most cases of IBPS) or into a systemic vein, the inferior vena cava or the azygos in EBPS. When venous return is anomalous and runs through the diaphragm to reach the inferior vena cava or portal vein, the malformation is known as the "Scimitar syndrome" because of the particular vascular silhouette delineated by the venous collector upon imaging [11]. Lung hypoplasia, bronchial malformations and dextrocardia are common in these cases and, although arterial inflow from the aorta occurs occasionally, the nosologic and therapeutic problems of the Scimitar syndrome are different from those of BPS, and are out of the scope of the present review.

EBPSs are more often located underneath the lower lobes of the lung, particularly in the left side. They can be incorporated into the sac of a congenital diaphragmatic hernia and can even be infradiaphragmatic, a location in which they can be misdiagnosed as adrenal masses. EBPSs have no connection with the airway and are therefore often compact. However, upon detailed histological study, a number of them [4, 5] that may amount to one-half [12] or more [13] have areas of CCAMs. EBPSs are relatively often associated with other malformations, particularly of the lung, heart, great vessels and gastrointestinal (GI) tract [14].

IBPSs are 3–4-times more frequent than EBPSs. They are also located predominantly in the lower lobes of the lungs, and the proportion of patients with associated malformations is much lower than in EBPS. Bilateral or whole-lung involvement is rare, but may occur [14]. IBPSs often have areas of associated CCAMs [15, 16].

22.3 Pathogenesis

Although in the past an acquired post-inflamatory origin was attributed to BPSs [17] and particularly to IBPSs [18], frequent prenatal diagnosis, pathology close to that of CCAMs,

and an association with other defects point to an embryonic, maldevelopmental origin that is further supported by some anomalies of molecular signalling. A few familial observations further reinforce this interpretation [2]. In cases with connection with the foregut, the simplest topographic explanation is the late and ectopic emergence of an abnormal lung bud from the distal foregut that becomes vascularized by systemic arteries. This would explain why EBPSs (but not IBPSs) seem to derive rather from "orthotopic" lung buds. Nevertheless, they probably share similar mechanisms because they occasionally have connections with the foregut.

Molecular and genetic actors leading to BPSs are not well known, but there is evidence of a wide degree of overlap with those of CCAMs and other foregut malformations [19]. Hox gene Hoxb5 protein is abnormally increased in BPS and CCAM tissues. This gene is normally expressed in the subepithelial mesenchyme of the airway branching points during the pseudoglandular stage of lung development and should not be so active at term [20]. Conversely, the cell adhesion molecules alpha (2)-integrin and E-cadherin, which are related to Hoxb5, are also overexpressed in them, attesting their likely participation in the pathogenesis of such conditions [21].

22.4 Clinical Features

Fetal disease: A large proportion of BPSs are diagnosed before birth when a solid or cystic portion of the lung receiving systemic arterial inflow is detected upon fetal ultrasonography. The vast majority of fetuses with BPS do well and only occasionally does the mass cause massive pleural effusion and life-threatening hydrops. In a series of 14 BPSs selected from 192 fetuses with lung lesions, only 2 BPSs required some form of prenatal intervention for drainage of ipsilateral hydrothorax, whereas in 4 BPSs, the mass regressed during gestation, and in the remaining 8 post-natal surgery was successful [22]. Some cases with BPS, pleural effusion and hydrops have abnormal venous anatomy and obstructed venous outflow that explain these disturbances [23]. Regression of BPS during fetal life is not uncommon [24], particularly in hybrid cases with adenomatoid components [1]. Some intraabdominal BPSs detected during pregnancy progressively involute and disappear [25].

Neonatal disease: Except in cases in which the malformation is huge and accompanied by pulmonary hypoplasia, neonatal symptoms are often absent in BPS. These infants, particularly those with EBPS and therefore without airway connection with the mass, have mild or no symptoms [26] and are often diagnosed by serendipity. Patients with massive hydrothorax or pulmonary hypoplasia have severe respiratory insufficiency (eventually pulmonary hypertension) and may require refined neonatal intensive care, including extracorporeal membrane oxygenation (ECMO) [27] prior to surgery (28). Infradiaphragmatic or intra-abdominal BPS, often hybrid in nature, tend to remain silent and are also diagnosed during imaging for other purposes [29–33].

Late symptoms: BPSs without CCAM components have no secretory tissue. Therefore, they do not tend to increase in size after birth. However, their marginal connection with the airway may insuflate the sequestered part of the lung. Infection is possible (particularly in IBPS) and "repeated pneumonia" in the same area of the lung (particularly in the lower lobes) should be investigated looking for BPS. Recurrent respiratory infection is the leading picture of BPS, but occurs predominantly in IBPS in which there are communications with the airway [13, 26]. Increased pulmonary blood flow from the systemic arterial supply can result in hemorrhage, hemoptysis, or high-output cardiac failure, although these are rare events [34]. Most BPSs remain asymptomatic during infancy and childhood, and repeated infection, shortness of breath and hemoptysis may manifest later or even in adult life [35].

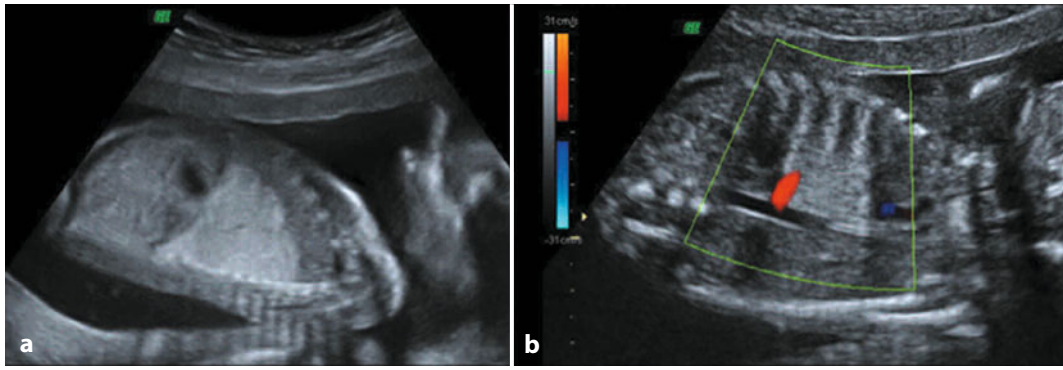

Fig. 22.2 Fetal ultrasound in which a condensation is visible in the lung (**a**). Upon color-Doppler ultrasonography, arterial inflow from the aorta is obvious (**b**)

22.5 Diagnosis

Routine prenatal ultrasound screening allows the diagnosis of most cases of fetal BPS from the twentieth week onwards. Areas of the lung with solid or complex ecogenicity and systemic arterial inflow upon color Doppler ultrasonography are indicative of BPS [36], especially if they are located in the lower lobes (Fig. 22.2). Pleural effusion and hydrops are rare but, in these particular cases, assessment of pulmonary development is necessary. Infradiaphragmatic BPSs located in the adrenal areas require diagnosis from congenital neuroblastoma (although these are more often cystic). Fast fetal magnetic resonance imaging (MRI) may occasionally help to define prenatally the nature and features of the sequestration and its vascular supply [37, 38].

Later in life, the diagnosis of BPS is based on the same methods used for imaging CCAMs plus the demonstration of the aberrant systemic arterial supply. Plain radiographs of the chest may show areas of condensation (eventually with cysts) in the lower lobes of the lung (Fig. 22.3). These may often be so discrete that they are easily missed. MRI [39] and computed tomography (CT) [40] can show the features of the sequestered tissue, but CT fares better for this purpose. In prenatally diagnosed, clinically silent cases, it is better to undertake an elective CT between the first and sixth months of life in order to define the features of the malformed tissue. Color Doppler ultrasonography is the preferred method for demonstrating the aberrant arterial supply at any stage of the diagnosis. However, detailed imaging is necessary whenever treatment is contemplated. Arteriograpy used to be the method for this purpose, but it has been progressively replaced by multidetector three-dimensional (3D) angio-CT [41-44], which allows complete mapping of the arterial inflow and venous outflow in amazing detail (Fig. 22.4).

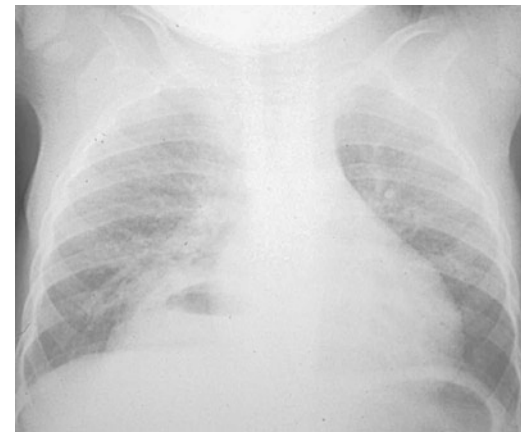

Fig. 22.3 Plain radiograph of the chest in a 18-month-old female with repeated lung infections. A right basal condensation with a cystic image is visible. She had an IBPS that was resected successfully

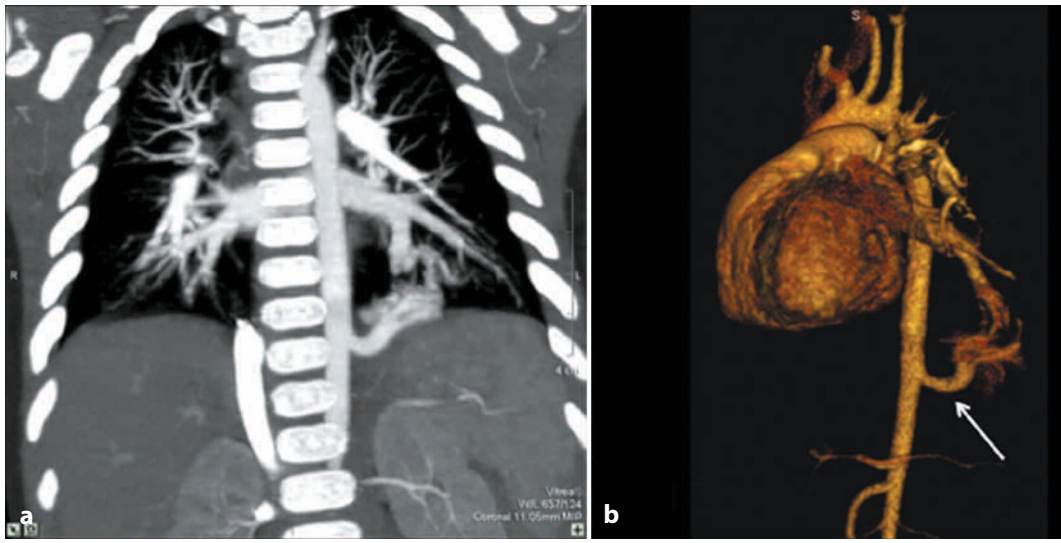

Fig. 22.4 Angio CT in a patient with a left basal IBPS. An ectopic arterial supply from the aorta and venous drainage to the pulmonary veins is readily visible (**a**). Reconstruction of the arterial supply (*arrow*) greatly facilitates the surgical strategy (**b**)

Other diagnostic procedures such as bronchoscopy are probably unnecessary. If communication with the foregut is suspected, esophagoscopy and barium meals are indicated.

22.6 Treatment

During fetal life: BPSs do not require prenatal treatment except in cases with massive pleural effusion and hydrops. Fetal thoracentesis and thoraco–amniotic shunts may be indicated if this life-threatening condition is detected, and have been applied successfully in several cases [22, 45-47]. Very seldom, large BPSs causing pulmonary hypoplasia have been treated by open surgery on the fetus [48]. Most BPSs detected before birth remain stable or undergo variable degrees of regression.

After birth: Symptomatic cases, those with neonatal respiratory distress, repeated pneumonia during infancy or childhood, or rarer events such as hemoptisis or high-output cardiac failure require active treatment. However, most BPSs are asymptomatic and whether they require treatment is debatable, particularly because of their well-known potential for regression. Some authors advise observation only for BPS, particularly for small, asymptomatic EBPSs [46, 49, 50]. Conversely, removal of the aberrant tissue is advised by other authors [35, 51, 52] on the basis of two arguments. The firstly is the presence of mucus-producing, cystic elements in one-half to two-thirds of cases. The tendency to accumulate mucus and the risks of infection that constitute the indications for removal of CCAMs also apply for BPSs. Secondly, the risk of cancer in the aberrant tissue later in life is rare but real (see below).

After elective imaging during the first semester of life, active treatment should be planned. There are two possible approaches: embolization or surgical removal.

Embolization was introduced in the stream of the rapid progress of percutaneous interventional radiology and cardiology. It was originally applied in the Scimitar syndrome to re-route the venous outflow by plugging the infradiaphragmatic collector while occluding the systemic arterial inflow with coils [53].

Coils [54, 55], plugs [53, 56] or ethylene vinyl alcohol copolymer [57] introduced through arterial catheters in the lumen of the feeding vessel can selectively destroy the sequestered tissue, and it is certainly tempting to rely on these methods as an alternative to major surgery. In cases with high-output cardiac failure, this procedure can save life and may be followed by surgical removal if necessary [34]. Preoperative embolization has also been recommended [58]. However, embolization for the primary treatment of BPS had only limited development in pediatric surgery [59] because it has its own complications (e.g., pleural effusions, limb ischemia) [54, 60] and because the sequestered tissues are not fully destroyed [61] and require further embolization or surgery [54, 60, 62, 63] in a sizeable proportion of cases. Most publications on the embolization of BPS involve case reports or small series that rarely contribute information to the long-term results of the procedure.

Surgical removal after ligation of the vascular pedicles is the preferred alternative in EBPS in which the sequestered tissue is separated from the lung and has its own pleural covering [26, 35, 46, 64]. The same applies to infradiaphragmatic EBPSs and to those associated with diaphragmatic hernia, which can be removed with the hernial sac during repair.

When dealing with IBPS, lobectomy is the first-line treatment [26, 27, 64, 65]. Parts of the affected lobe may be healthy and have normal arterial inflow, but complete resection is the only way to achieve destruction of the entire sequestered tissue. It has been shown for CCAMs (and most BPSs contain such tissue) that macroscopic appreciation of the extent of the diseased tissue is rarely precise, and that lobectomy remains the best way of staying on the safe side [66]. The procedure should be started by carefully mobilizing the affected lower lobe upwards to identify first the aberrant systemic artery (Fig. 22.5). This may be very large, but with thin elastic walls, and careful ligation and division should be done taking into account that retraction of the artery underneath the diaphragm could be cat-

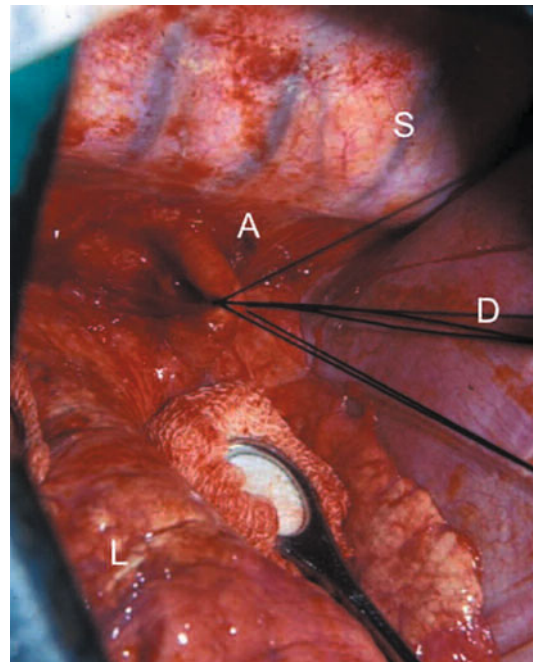

Fig. 22.5 First step for a right basal lobectomy for IBPS. The ectopic artery (*A*) is being ligated. The spine (*S*), lung (*L*) and diaphragm (*D*) are visible

astrophic. Once the artery is secured, lobectomy proceeds in the usual way (although dense adhesions due to previous repeated infections may make this part of the procedure difficult). In expert hands, lobectomy in an infant has few complications and minimal functional consequences provided that most of the resected lung is isolated from the airway.

Thoracoscopic lobectomy for BPSs has developed rapidly. This trend will probably continue because the proportions of conversions and complications are low [67–73].

22.7 BPS and Cancer

The risk of cancer in BPS is probably related to the hybrid nature of many of these lesions [74] and to the presence of CCAM elements and mucus-secreting structures prone to becoming malignant [8]. This practically never happens during childhood, but several cases that developed in adults with undiag-

nosed sequestrations were reported. The nature of these tumors was variable: sclerosing hemangiomas [75], pleuropulmonary blastomas (PPBs) [76, 77], adenocarcinomas or bronchoalveolar carcinomas [78, 79], carcinoid tumors [80], multiple neuroendocrine tumorlets [81, 82], fibrous mesothelioma [83] or lymphoepithelioma-like carcinoma [84]. It is obvious that the risk of malignancy (even if very remote) is an argument for the removal of the sequestered tissue.

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