

Pulmonary sequestration: a review of 26 cases

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Received 1 September 1997; revised version received 15 April 1998; accepted 12 May 1998

Abstract

Objectives: Pulmonary sequestration is a continuum of lung anomalies for which no single embryonic hypothesis is yet available. The aim of this study was to assess the diagnostic tools and treatment for the rare condition, pulmonary sequestration, in an unspecialised centre. **Methods:** We performed an analysis of 26 cases of pulmonary sequestration (paediatric and adult) operated at the Centre Hospitalier Universitaire Vaudois between May 1959 and May 1997. A review of the extralobar and intralobar types of sequestrations is discussed. Angiography is compared to other diagnostic tools in this condition, and treatment is discussed. **Results:** Twenty-six cases of pulmonary sequestrations, a rare congenital pulmonary malformation, were operated on in the defined time period. Seventy-three percent (19) of the cases were intralobar and 27% (seven) extralobar. Extralobar localisation was basal in 71% and situated between the upper and the lower lobe in 29%. In six cases, the diagnosis was made by exploratory thoracotomy. In the other 20 cases, diagnosis was evoked on chest X-ray and confirmed by angiography. Lobectomy (46%) was the most common treatment procedure. Segmental resection was performed in 30% of the cases and bilobectomy in 4%. Post-operative morbidity was low. The most significant complications were pleural empyema, haemothorax and haemopneumoperitoneum in case of extralobar sequestration. There was no evidence of metaplasia or pre-neoplastic changes. **Conclusions:** Despite its rarity, some radiological features are sufficiently suggestive of diagnosis of pulmonary sequestration. Investigations are necessary in order to avoid unexpected pathology at the time of operation. Resection of the involved lung leads to excellent results and the long-term outcome is highly favourable. © 1998 Elsevier Science B.V. All rights reserved

Keywords: Pulmonary sequestration; Malformation; Extralobar; Intralobar

1. Introduction

Pulmonary sequestration is a rare congenital malformation characterised by a mass of non-functioning lung tissue separated from the normal bronchopulmonary tree and vascularised by an aberrant systemic artery [6,16]. It represents between 0.15 and 6.45% of all pulmonary malformations. No chromosomal abnormality could be identified in any of the patients presenting with such a malformation. Rokitsansky and Rektorzic described the first case over 100 years ago. These authors proposed that the sequestration was due to a separated but normally developed lung fraction. Since then, several theories were put forward to explain the genesis of this anomaly [1–4]. Pryce's traction theory is generally felt to be the most accurate explanation of this pathology. He and his group were the first to use the

term sequestration. They furthermore identified the association between congenital bronchial isolation and vascularisation by an aberrant artery of the systemic circulation. The latter arise commonly from the thoracic aorta, from the coeliac trunk or from intercostal arteries.

Two types of pulmonary sequestration are recognised, depending on whether or not the malformation possesses its own pleural covering. Intralobar sequestration is an abnormal region within the normal pulmonary parenchyma without its own pleural covering. Extralobar sequestration corresponds to a true accessory lung, with its own pleural envelope [3,5,8–13]. There are numerous differences between these two types.

1.1. Localisation

Intralobar sequestration is in general found in the posterolateral segment of the left lung while extralobar sequestration is more often found in the lower lobes, especially in the

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left costodiaphragmatic sinus. In the case of extralobar disease, infradiaphragmatic, apical and bilateral lesions are also found. Extralobar masses are often clinically silent and only found by chance, generally during exploratory thoracotomy.

1.2. Size of feeding artery

In intralobar disease, the feeding artery is usually of large calibre, in contrast to extralobar disease where it is frequently quite thin.

1.3. Venous drainage

Intralobar disease is always drained by the pulmonary veins whilst extralobar disease is drained by the azygous or portal vein [1,3,5].

1.4. Communication with the pulmonary tree

This is sometimes seen in the case of intralobar sequestration, but is very rarely seen in extralobar disease.

1.5. Sex distribution

Intralobar disease is approximately equally distributed between sexes whilst extralobar disease is found more commonly in men (80% of cases).

1.6. Age at diagnosis

Extralobar disease is usually diagnosed in infancy or childhood whilst intralobar disease often remains unrecognised until after the age of 20.

1.7. Preferential side

Intralobar sequestration demonstrates no preferences for either lung whilst extralobar sequestration is found in the left lung in 80% of cases.

1.8. Associated diseases

Intralobar sequestrations are not associated with other cardio-pulmonary anomalies, but extralobar sequestrations may be found in association with cardiac, or more frequently, diaphragmatic anomalies in 50% of cases [26,27]. The fact that intralobar and extralobar sequestrations can be found simultaneously suggests that the two forms might share a common embryopathogenic basis [1]. Clinically, the malformation remains generally silent as long as there is no secondary infection or communication with the bronchial tree or gastrointestinal tract. The diagnostic investigation of choice remains aortography, but it has to be demonstrated whether digital subtraction angiography or MRI angiography will prove more efficient and less invasive [19,28]. Pulmonary angiography and bronchography

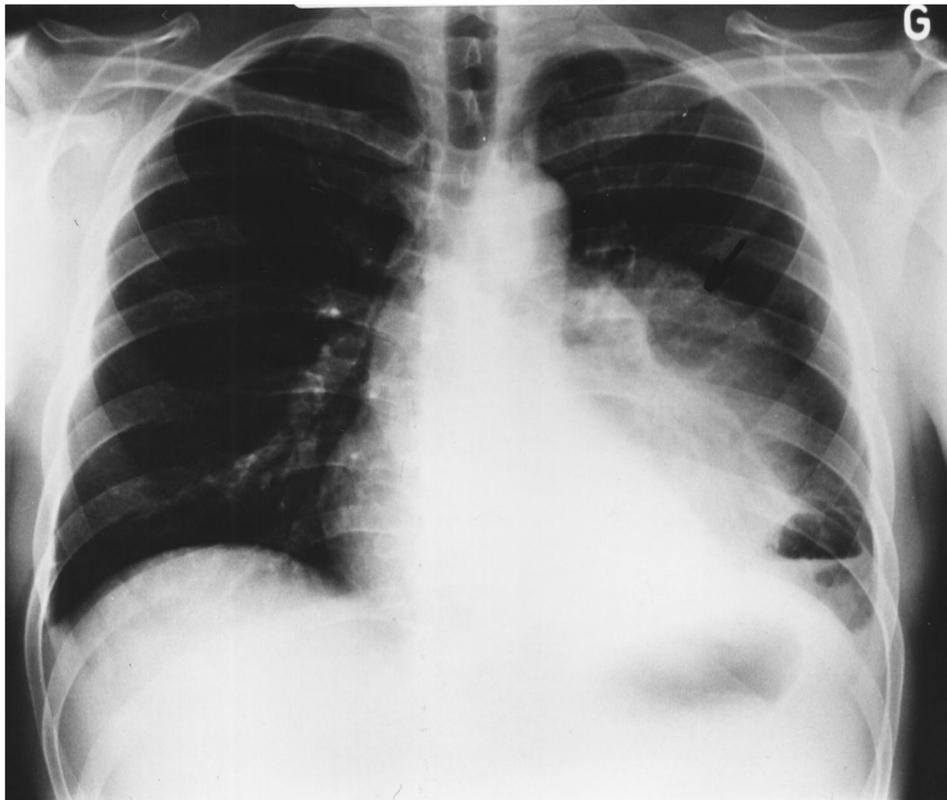


Fig. 1. Pulmonary sequestration. Chest X-ray of an adult with recurrent pneumonia. The film shows radiodensity in the left retrocardiac region.

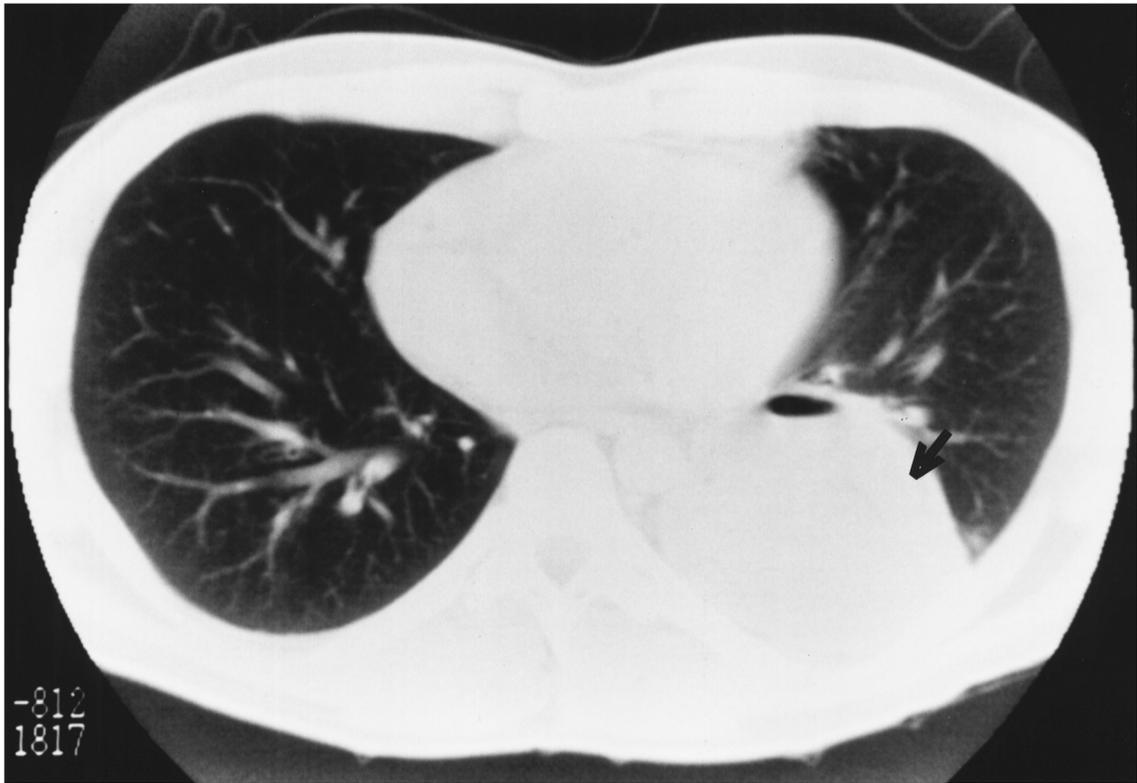


Fig. 2. Computed tomography scan showing the mass in the left lower lobe.

can help with the diagnosis, but are seldom diagnostic by themselves [29–32].

The mainstay of treatment has always been surgical excision. In 1981, Genton et al. suggested that surgical excision is indicated only in the cases of recurrent infection or secondary heart failure [22]. This was supported by Zumbro and numerous other Canadian surgeons who observed that few patients with pulmonary sequestrations had pulmonary symptoms [7,22,23]. They proposed conservative treatment for the majority of patients, and reserved surgical intervention only in the case of complications [2,7,24,25]. We however, would recommend surgical treatment, usually complete excision, even in the absence of symptoms. The diagnosis is often suspected from the results of clinical and radiological examination. On simple chest radiography, an elongated, sometimes cystic lesion, lying adjacent and frequently posterior to the pericardium is strongly suggestive of the diagnosis. However, no distinction can be made between intra- and extralobar disease, and on occasion it is impossible to exclude other cystic structures [8,14,15, 17,18]. Communication with the GI tract can be ruled out by the use of contrast studies. Bronchography allows dilatation of the bronchi to be excluded, may show bronchial obstruction in association with the sequestration, and allows the bronchial anatomy to be mapped. If active infection is present, bronchoscopy may show pus coming from bronchi adjacent to the sequestered segment. Ultrasound will delineate abnormal vessels and pulmonary tissue, but the results

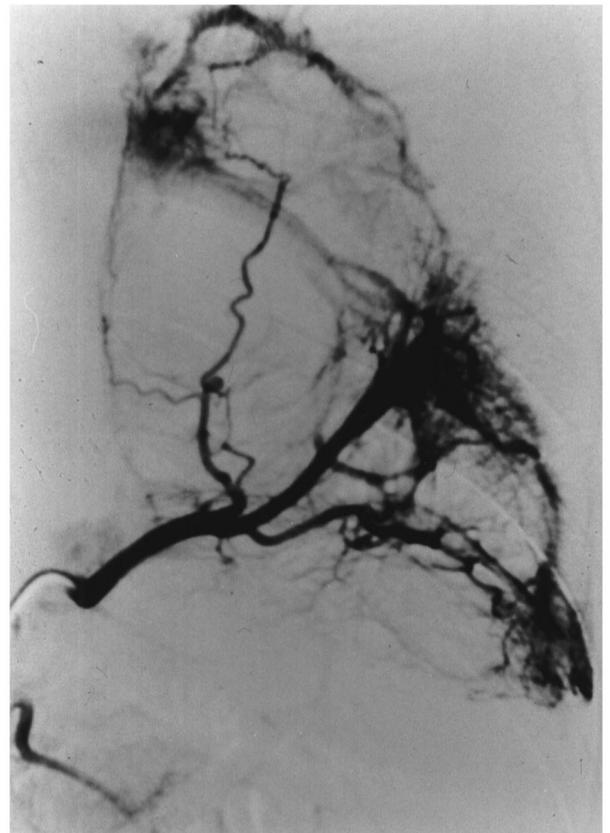


Fig. 3. Thoracic aortography demonstrates a large aberrant artery arising from the distal thoracic artery supplying the lesion.

are only variably interpretable. The examination of choice to display the topographical features and nature of the abnormal pulmonary parenchyma is CT-scan, but for the definitive confirmation of the diagnosis, angiography is required [1,19]. The recent advent of MRI angiography may replace the need for conventional angiography, a procedure which is invasive, and which can be difficult sometimes [1,7,17,20–22].

In the present study, we reviewed the cases diagnosed and operated in Lausanne, in order to assess the diagnostic and treatment accuracy in a non-specialised medium-sized hospital.

2. Patients and methods

We reviewed the files of all patients diagnosed with pulmonary sequestration between May 1959 and May 1997 in the Lausanne University Hospital. The following data were collected for each patient: sex, age, medical history, major complaint at admission, diagnostic procedures, treatment, histological diagnostic and short-term outcome. Operative treatments and findings including exact anatomical location of the sequestered lung and abnormal vasculature were evaluated.

3. Results

In the Department of surgery at the CHUV in Lausanne, 26 cases of pulmonary sequestration were operated on between May 1959 and May 1997. There were 16 males, ten females, four children and 22 adults. The average age of the children operated on was 8 months, ranging from 1 day to 3 years. Amongst the adult population, the average age was 25 years, ranging from 15 to 33. All patients were initially selected on the basis of a suspect chest X-ray. Two patients were diagnosed on the basis of a suspect pulmonary scintigraphy. All diagnoses were confirmed by histology, see Figs. 1–6.

Among 26 patients, 19 presented with signs and symptoms of pulmonary disease: recurrent pneumonia (five), respiratory distress (five), hemoptysis (three), stridor (three), thoracic pain (two) or pleurisy (one). Three cases had secondary infection with *Mycobacterium tuberculosis* (TB), four with *Aspergillus umigatus* and two with *Nocardiose asteroides*. The other infections were due to undetermined agents.

Nineteen of the 26 cases of sequestration were intralobar, and the other seven were extralobar. Of 19 patients presenting with intralobar sequestrations, seven were asymptomatic. Among these, four were discovered during surgery

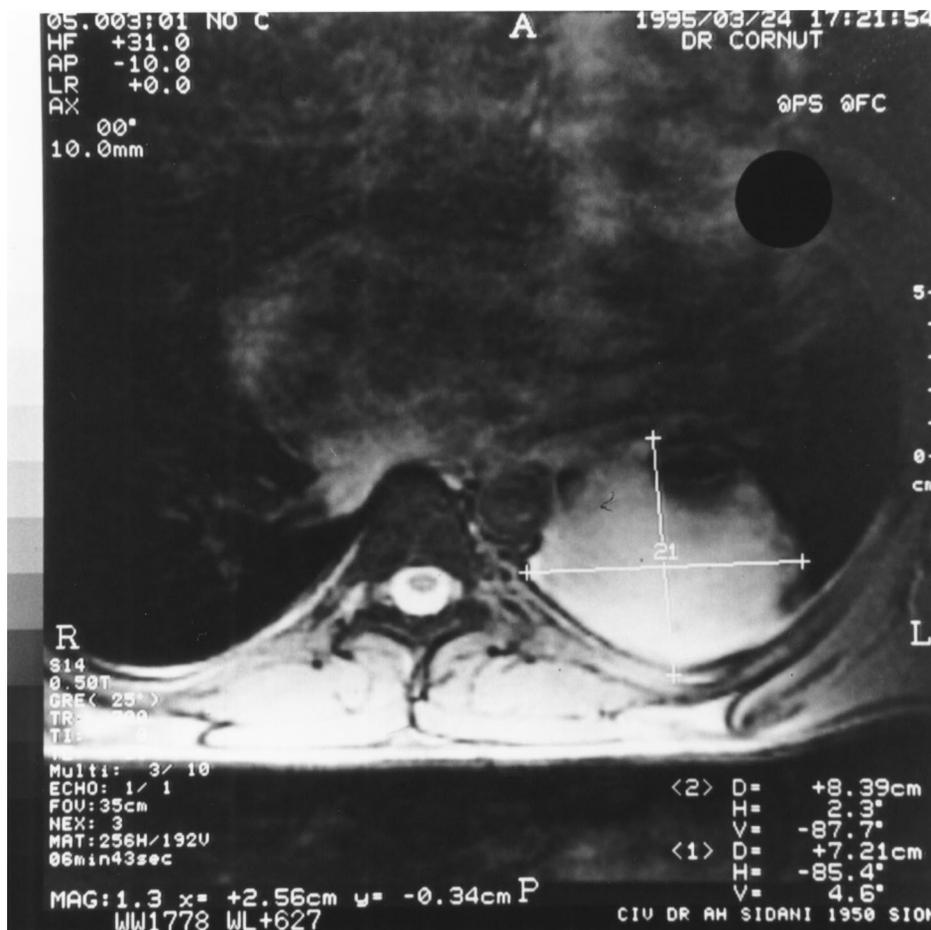


Fig. 4. Pulmonary sequestration. MR image.

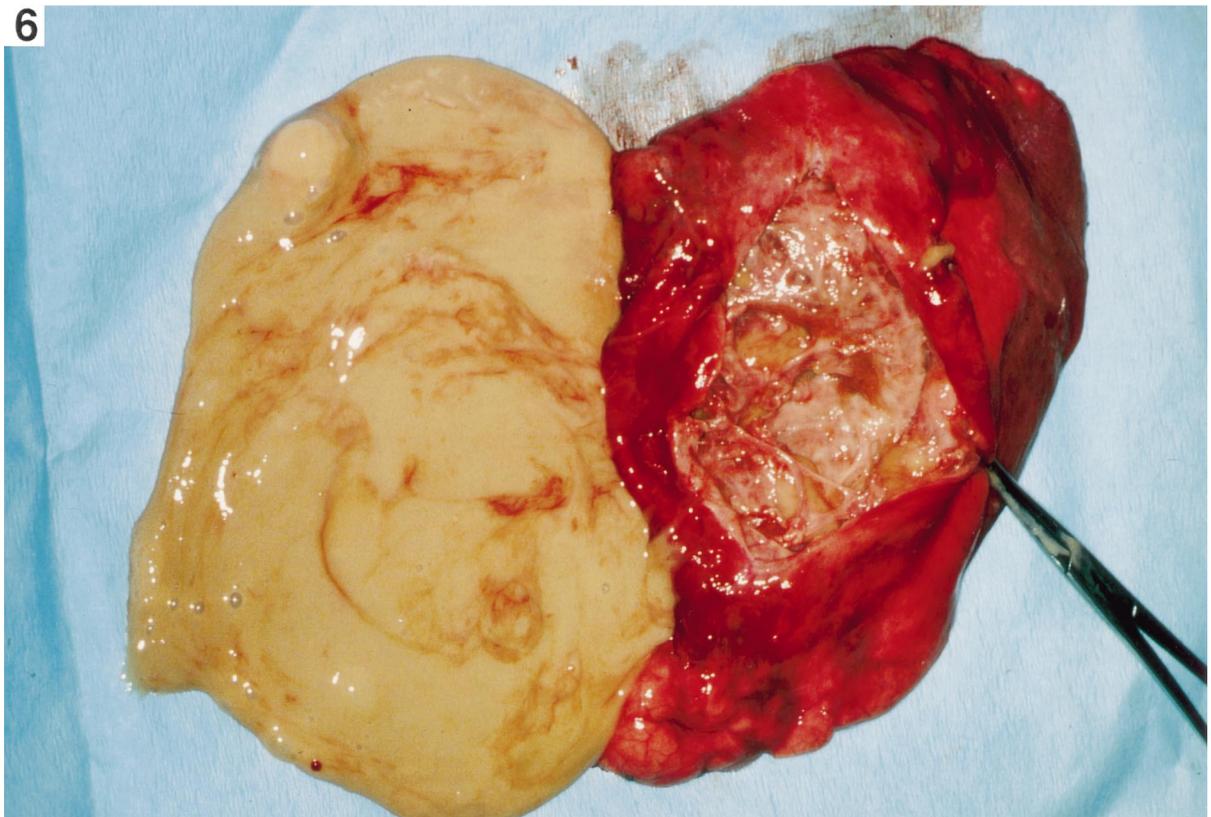
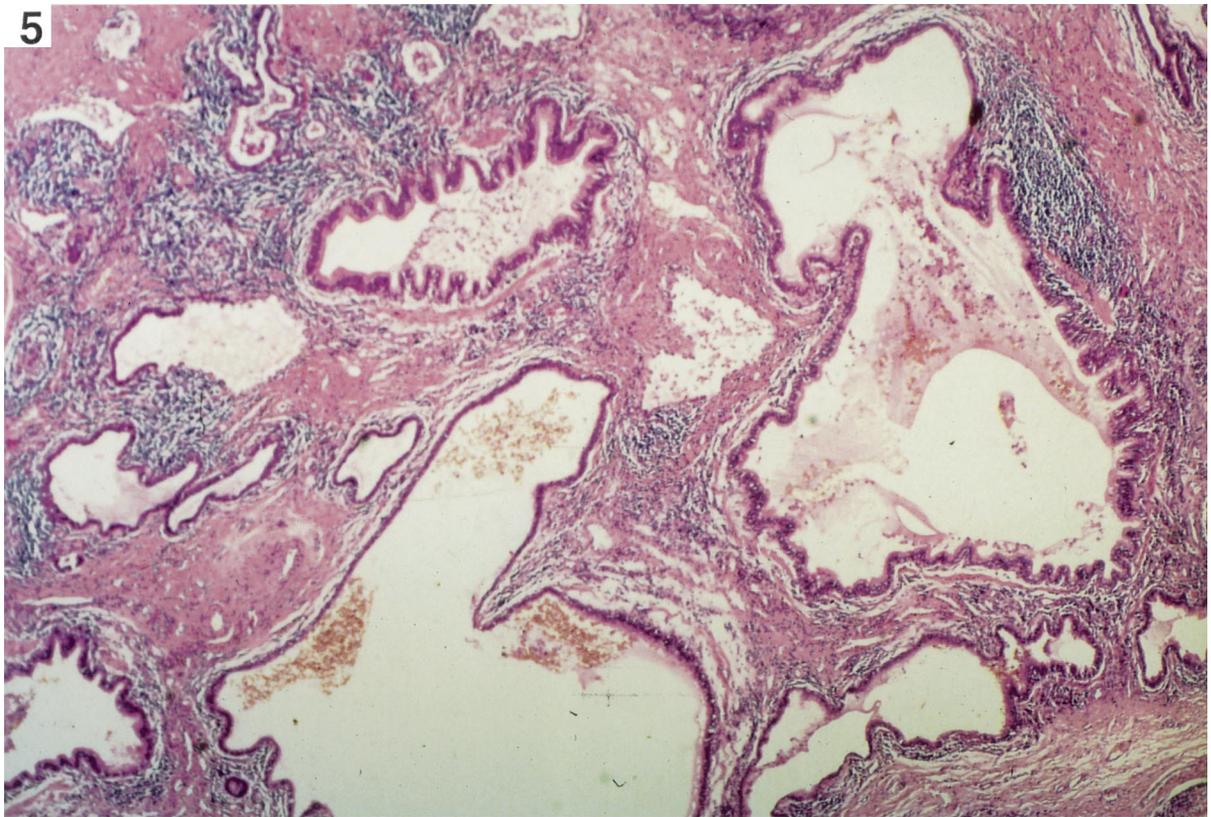


Fig. 5. A typical histological picture of an intrapulmonary sequestration.
Fig. 6. Same patient as in Fig. 1. Note the cyst and the absence of anthracosis in sequestered areas.

Table 1

Major theories of origin of bronchopulmonary sequestration

Year	Author	Theory
1861	Rokitansky and Rektorzik	Fraction theory: separation of normally-developed lung
1878	Ruge	Excess theory: the malformation arises separately from the normal lung as the development of a rudimentary third lung
1946	Pryce	Traction theory: separate artery runs to the embryonal lung-anlagae
1950	Smith	Blood supply theory: parenchymatous changes induced by insufficient arterial blood supply
1959	Gebauer and Mason	Theory of acquired anomalies: result of recurrent infections
1959-today	Several authors	Idiopathic theory: persistent anomalous vessel or branch between dorsal or ventral aorta to splanchnic plexus

for cardiac malformations (two with Fallot's tetralogy, one with an atrial septal defect and one with scimitar syndrome). In the three remaining patients, the definitive diagnosis had been suggested preoperatively by chest X-ray, MRI scanning (including one MRI angiography) and retrograde angiography.

At pathological examination, the size of the specimen varied from a diameter of about 2 cm to a lesion measuring $15 \times 11 \times 8$ cm. The feeding artery varied from 3 mm in diameter in the smallest instance to 14 mm in the largest sequestration. The main vascular supply to the lesions arose from the thoracic aorta in 80% and from the abdominal aorta in 20% of cases. Venous drainage was through the hemiazygous and superior vena cava as well as the pulmonary vein, except for two cases where drainage was via the inferior vena cava or via the portal vein. Seventy-one percent of extralobar sequestrations were found at the level of the lower lobes, with the other 29% in an interlobular fissure.

Treatment evolved with time. Altogether, 12 patients underwent a lobectomy (46%), one had a bilobectomy (4%) and one a pneumonectomy (4%). Four patients underwent simple ligation of the feeding artery (15%) (two by laparotomy, and two by thoracotomy). Eight patients had simple excision of the lesion (30%). There was no short-term postoperative mortality. Complications included three haemothorax after lobectomy, two bronchial fistula, two empyemas and one intra-abdominal haemorrhage after excision of an extralobar sequestration. There were no cases of malignant transformation of the sequestration, either bronchial or pulmonary. Results of long-term postoperative follow-up were not analysed as such. Among patients who came back to the hospital, there were no recurrences, or mid- or long-term complications.

4. Discussion

The present study was aimed at reviewing our experience with a rare disease. Due to its rarity, (540 known cases between the years 1862 and 1975, as quoted by Savic [1]), series from any single hospital are bound to be small. Furthermore, treatment varies along the years, rendering therapeutic modalities difficult to assess. In summary, pulmon-

ary sequestration arises from an abnormal pulmonary development along with pathological vascularisation. Our findings do not allow us to present new proposals concerning the aetiology of this disease (Table 1). In our experience, this pathology reaches clinical significance at the time of infection of the sequestered tissue. It can also be a fortuitous operative finding. Given the probability of developing infectious complications, the sequestered tissue should be excised. In the case of extralobar sequestration, the anomalous artery and vein can be divided close to their origin and the non-ventilated parenchyma removed. As observed in the present study, excision of the sequestered lung tissue is a safe procedure with a low complication rate. Although not used in our hospital during the studied time period, thoracoscopy seems now to find its place in the surgery of this disease [25].

In spite of its rarity, pulmonary sequestration should be considered in presence of certain features on a chest X-ray, prompting further investigations in order to avoid encountering an unexpected pathology during surgery. Diagnosis is best confirmed by CT-scan and retrograde aortography. Except for exceptional inoperable cases, intralobar asymptomatic forms should be excised prophylactically, because of infection hazard.

References

- [1] Savic B, Birtel FJ, Tholen W, Funke HD, Knoche R. Lung separation: report of seven case review of 540 published cases. *Thorax* 1979;34:96–101.
- [2] O'Mara CS, Baker RR, Jeyasingham K. Pulmonary sequestration. *Surg Gynecol Obst* 1978;147:609–616.
- [3] Gdaniez K, Vorpahl K, Piehl G, Hock A. Clinical symptoms and therapy of lung separation. *Prog Pediatr Surg* 1987;21:86–97.
- [4] Carter R. Pulmonary sequestration. *Ann Thorac Surg* 1969;7:68–88.
- [5] Sade MR, Clouse M, Ellis H. The spectrum of pulmonary sequestration. *Ann Thorac Surg*. 1974;18:644–658.
- [6] Kravitz RM. Congenital malformations of the lung. *Ped Clin N Am* 1994;41/3:453–473.
- [7] Vogt-Moykopf I, Rau B, Branscheid D. Surgery for congenital malformations of the lung. *Radiol* 1993;36/2:145–160.
- [8] Boumghar M, Besson A, Saegeseer F. Sequestration pulmonaire intralobaire et surinfection tuberculeuse. *Schweiz. Med. Wschr.* 1979;109:1460–1464.

- [9] Osada, H., Yokote, K., Arakawa, H., Yamate, N. Bilateral intralobar pulmonary sequestration. *J Cardiovasc Surg* 1995;1995:36:611–613.u.
- [10] Saegesser F. Sequestrations pulmonaires. *Schweiz Med Wschr* 1968;49:1919–1927.
- [11] Leijala M, Louhimo I. Extralobar sequestration of the lung in children. *Prog Pediatr Surg* 1987;21:98–106.
- [12] Landing BH. Congenital malformations and genetic disorders of the respiratory tract. *Am P Resp Dis* 1979;120:151–184.
- [13] Rodgers MB, Harman PK, Johnson AM. Bronchopulmonary foregut malformations. *Ann Surg* 1986;203:517–524.
- [14] Schulman MH. Cysts and cystlike lesions of the lung. *Pediatr Chest* 1993;31:631–649.
- [15] Saegesser F, Besson A. Extralobar and intralobar pulmonary sequestration of the upper lobes. *Chest* 1973;63:69–73.
- [16] Jansen D, Schilte PM, De Graaff C, Van Dijk HA. Bronchopulmonary sequestration with an aneurysm of the aberrant artery. *Ann Thorac Surg* 1995;60:193–194.
- [17] Ben Mustapha C. Un piège diagnostique: la sequestration pulmonaire, interet de TDM. *Tunisie Méd* 1999;70:161–164.
- [18] Fu-Jan K, Shi-Chuan C, Wei-Juin S, Reury-Perng P. Extralobar pulmonary sequestration presenting as an anterior mediastinal tumor in an adult. *Chest* 1993;104/1:303–304.
- [19] Jeanbourquin D, Jancovici R, Cordoliani YS, Pons F. Place de l'arteriographie dans le bilan sequestrations pulmonaires. *Ann Chir Thorac Cardio-Vasc* 1994;487/3:275–276.
- [20] Ndiaye M, Fall I, Deme A, Dieng PND, Ndiaye PD, Diop A. Forme kystique de sequestrator pulmonaire extra-lobaire. *Ann Chir* 1996;50/5:409–412.
- [21] Bargetzi MJ, Saltzman J, Maurer W. Bronchopulmonale sequestration. *Schweiz Med Wschr* 1991;121:424–428.
- [22] Begin P, Lamarre A, Blanchette G. La sequestration pulmonaire. *Union Med. Canada* 1985;114:301–307.
- [23] Clements BS, Warner JO. Pulmonary sequestration and related congenital bronchopulmonary malformations: nomenclature and classification based on anatomical and embryologic considerations. *Thorax* 1987;42:401–408.
- [24] Haller JH, Golladay ES, Pickard RL, Tenas JJ, Shorter AM, Shermeta DW. Surgical management of lung bud anomalies: lobar emphysema, bronchogenic cyst, cystic adenomatoid malformations and intralobar pulmonary sequestration. *Ann Thorac Surg* 1979;28:33–43.
- [25] Watine O, Mensier E, Delecluse P, Ribet M. Pulmonary sequestration treated by video-assisted thoracoscopic resection. *Eur J Cardiothorac Surg* 1994;8:155–156.
- [26] Avishai V, Dolev E, Weissberg D, Zajdel L, Priel IE. Extralobar sequestration presenting a massive hemothorax. *Chest* 1996;109/3:843–845.
- [27] Cukier A, Kavakama J, Teixeira LR, Terra-Filho M, Vargas F. Scimitar sign with normal pulmonary venous drainage and systemic arterial supply. *Chest* 1994;105:294–295.
- [28] Donovan CB, Edelman RR, Vrachliotis TG, Frank HA, Kim D. Bronchopulmonary with MR angiographic evaluation. *Angiology* 1994;45:239–244.
- [29] Louie HW, Martin SM, Mulder DG. Pulmonary sequestration: 17 year experience at UCLA. *Surgery* 1993;12:801–805.
- [30] Merlier M, Rochainzmir A, Rojas-Miranda A, Levaseur Ph, Sulzer JD, Verley J-M, Binet LJ. Aspects anatomo-cliniques des sequestrations pulmonaires. *Ann Chir Thorac Cardio-Vasc* 1970;9:511–514.
- [31] Grewal, R.G., Yip, C.K. Intralobar pulmonary sequestration and mediastinal bronchogenic cyst. *Thorax* 1994;49:615–616.
- [32] Cerruti MM, Marmolejos F, Cacciarelli T. Bilateral intralobar pulmonary, sequestration with horseshoe lung. *Ann Thorac Surg* 1993;55:509–510.