In 1946, Pryce used the term sequestration for the first time. Intralobar sequestrations (ILS) are masses of non-functioning lung parenchyma that are contiguous with the normal adjacent lung tissue and separate from the normal tracheobronchial tree, with no distinct pleura, but with a systemic artery supply coming from the descending thoracic aorta, the coeliac trunk or from intercostal arteries. There are numerous case reports about pulmonary sequestration, and large series reported are extremely rare. A systemic artery supply was found during the preoperative period in 11. Surgery consisted of lobectomy (n = 20), bilobectomy (n = 1), segmentectomy (n = 4), and pneumonectomy (n = 1). There were no postoperative deaths, and the postoperative course was uneventful in 20 patients. All patients were alive and faring well at long-term follow-up (mean follow-up 36.5 ± 7.2 months). Surgery consisted of lobectomy in most cases. The arterial supply came from the descending thoracic and abdominal aorta. Hemoptysis and/or recurrent infections were present in 14/26 (54%) of patients. These are the same symptoms as those leading to the diagnosis of bronchectasis. This suggests, for similar reasons, that ILS in adults should be nosologically very similar to acquired lesions, such as bronchectasis.

Keywords: Sequestration

1. Introduction

In 1946, Pryce used the term sequestration for the first time. Intralobar sequestrations (ILS) are masses of non-functioning lung parenchyma that are contiguous with the normal adjacent lung tissue and separate from the normal tracheobronchial tree, with no distinct pleura, but with a systemic artery supply coming from the descending thoracic aorta, the coeliac trunk or from intercostal arteries. There are numerous case reports about pulmonary sequestration, and large series reported are extremely rare [1–4] and make no distinction between intra- and extralobar sequestration (ELS). In this study, we examined the characteristics and the outcomes of ILS.

2. Patients and methods

We retrospectively reviewed data we obtained from the medical records of patients with ILS who underwent surgery at Laennec, Georges Pompidou European, and Amiens South Hospital. From 1985 to 2010, 26 consecutive adults patients underwent surgery for ILS. There were 14 males and 12 females. The average age was 37.3 years, ranging from 14 to 73 years. The ILS was right-sided in 11 patients (42.3%) and left-sided in 15 patients (57.7%). A systemic artery supply was found during the preoperative period in 11. Surgery consisted of lobectomy (n = 20), bilobectomy (n = 1), segmentectomy (n = 4), and pneumonectomy (n = 1). There were no postoperative deaths, and the postoperative course was uneventful in 20 patients. All patients were alive and faring well at long-term follow-up (mean follow-up 36.5 ± 7.2 months). Surgery consisted of lobectomy in most cases. The arterial supply came from the descending thoracic and abdominal aorta. Hemoptysis and/or recurrent infections were present in 14/26 (54%) of patients. These are the same symptoms as those leading to the diagnosis of bronchectasis. This suggests, for similar reasons, that ILS in adults should be nosologically very similar to acquired lesions, such as bronchectasis.

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A systemic artery supply was found during the preoperative period in 11 patients and the diagnosis of sequestration was only made in 42% of cases. There was one feeding vessel in eight patients, two in one patient and three in two patients. The feeding vessel came eight times from the descending thoracic aorta, three times across the pulmonary ligament, two times from the coeliac trunk, and once from a phrenic artery. The venous drainage was by mortality, occurring within 30 days or before discharge. Follow-up information was obtained at the time of office visits or by contacting the patients or their relatives or physicians by mail.

3. Results

From December 1985 to April 2010, 26 consecutive patients underwent surgery for an ILS. There were 14 males and 12 females, all were adults. The sex ratio was 0.54. The average age was 37.3 years, ranging from 14 to 73 years.

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pulmonary veins in 25 cases and by the azygos vein in one case.

Surgery consisted in lobectomy (n=20), bilobectomy (n=1), segmentectomy (n=4), and pneumonectomy (n=1). The preoperative systemic blood supply was confirmed for all 11 patients during surgery, and systemic vascularization was demonstrated in all other 15 patients: across the pulmonary ligaments (n=2), presenting as important bronchial arteries (n=7), and across pulmonary adhesions (n=6).

There were no postoperative deaths, and the postoperative course was uneventful in 20 patients.

Postoperative complications (25%) were: pleural empyema (n=1), hemoptysis (n=1), prolonged air leak (n=2), arrhythmia (n=1), fistulae (n=1) (5%). All patients were alive and faring well at long-term follow-up (mean follow-up 36.5±7.2 months ranging from 7 to 140 months).

At pathologic examination, the size of the ILS ranged between 2 and 10 cm, mean = 5.3±2.7 cm. Pathological examination confirmed the diagnosis of pulmonary ILS and demonstrated that six lesions were infected with *Aspergillus fumigatus*.

4. Discussion

ILSs are rare congenital bronchopulmonary anomalies, most of them being published as case reports. To our knowledge, five series have been reported. The number of patients varied between 7 and 21 [1–5]. In the latter series [4] ELS were included (n=8). To our knowledge, our series is the largest report focusing on ILS.

ILS rarely causes problems before the age of two years [5] and 15% of adult patients do not have any complaints when the sequestration is discovered [5]. Presentation is similar whatever the sex of the patient and the anomaly is generally isolated, not associated with other malformations. ILS affect the basal segments of the lower pulmonary lobes and location in the upper lobes as we observed in one case, is extremely rare, and was reported by Savic et al. in only 2% of 540 patients reviewed in the literature. ILS are located on the left side in 60% of cases [6] which we also observed. The arterial supply comes from the descending thoracic and abdominal aorta (73% and 27%, respectively) [5], either directly or by means of other arteries. We observed bronchial arteries supplying the ILS territory in seven patients. At pathological examination in these patients was typical of ILS, but such vascularization was the question of an acquired origin.

Hemoptysis [7] and/or recurrent infections permitted diagnosis of ILS in two-thirds of our patients. These are the same symptoms as those leading to the diagnosis of bronchectasis. Some authors suggest, for similar reasons, that ILS in adults are acquired lesions [8].

Superimposed infection by fungal pathogens and especially by *Aspergillus fumigatus* are extremely rare (to our knowledge only 18 cases have been reported [9]), and should be supplementary reasons to consider ILS as acquired lesions [10]. In our series, only three patients had no aberrant systemic blood supply but only hypertrophic vessels (two with recurrent pneumonia and one with aspergil- lus), which does not favor this hypothesis.

Presentation as pseudo-tumoral masses is not uncommon, and was observed in 6% of patients in the review made by Savic and colleagues [5]. In our series, the pseudo-tumoral masses were all located on the left. In these patients, preoperative work-up did not permit to disclose the systemic aberrant artery. Attention must be paid with these pseudo-tumoral forms because peroperative massive bleeding from the aberrant vessel may occur in cases of sub-diaphragmatic origin, the artery retracting into the abdomen cavity and causing fatal haemorrhage as reported by Savic et al. [5]. More rarely, the aberrant vessel can be complicated by aneurysmal evolution [11] or severe hemotherox due to the rupture of the supplying artery inside the pleura [12–14].

Surgery consists in lobectomy in most cases reported in the literature, however, a more limited resection is possible in case of limited ILS as we observed in four patients and as it was reported [1]. Pneumonectomy was needed in one patient because previous tuberculosis history led to associated destroyed lung. To our knowledge, no pneumonectomy was reported in case of ILS in the literature we reviewed.

5. Conclusion

ILSs are rare congenital bronchopulmonary anomalies and affect preferentially the basal segments of the lower pulmonary lobes. The arterial supply comes from the descending thoracic and abdominal aorta. Venous drainage is usually by the pulmonary veins. Hemoptysis and/or recurrent infections are present in two-thirds of patients. These are the same symptoms as those leading to the diagnosis of bronchectasis. This suggests, for similar reasons, that ILS in adults should be nosologically very similar to acquired lesions, such as bronchectasis.

Surgery is safe and consists in lobectomy in most cases, but attention must be paid with these pseudo-tumoral forms because peroperative massive bleeding from the aberrant vessel may occur.

References


972


