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Case Report

Intralobar Pulmonary Sequestration Harbouring Occult Lung Cancer Therapeutic Implications for a Congenital Malformation Case Report and Review of the Literature

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Abstract

Non-small-cell lung cancer within congenital intralobar pulmonary sequestration represents a very rare coincidence. To our best knowledge, only twelve cases of pulmonary neoplasia associated with intralobar sequestration have been published.

A 46 year old female patient, non-smoker, presented with recurrent episodes of fever and fatigue persisting over several months. Routine chest roentgenogram showed a high-density mass in the right lower lobe close to the diaphragmatic surface. CT-scan revealed a 4 x 3 x 8 cm mass in the posterobasal segment of the right lower lobe with systemic arterial supply from two large caliber vessels originating from the celiac artery and draining into the pulmonary vein.

During segmental resection the sequester could not be separated from the diaphragm, a small part of which was also resected. Surprisingly, definitive histopathological examination revealed pulmonary adenocarcinoma pG3, pT3 within the intralobar sequestration. Our case and the review of the cases from the literature underline the suggestion, that pulmonary sequestration should be resected rather than undergoing treatment by embolization.

INTRODUCTION

Intralobar sequestration is a rare congenital disorder representing an area of dysplastic, nonfunctional lung within one pulmonary lobe. Usually a lower lobe, predominantly on the left side is affected. The sequester is supplied by an aberrant systemic artery, originating most frequently from the thoracic or abdominal aorta. Drainage is usually toward the pulmonary veins. Though the condition may remain asymptomatic, complications including congestive heart failure due to arterio-venous shunting, recurrent pulmonary infection or hemoptysis can evolve. Extralobar sequestration where the systemically supplied sequester has its own pleural covering and is always draining into the systemic veins, is even more uncommon. With the exception of arterio-venous shunting or bleeding, the condition remains asymptomatic [1,2]. The development of a malignant lung tumour within lung sequester is an uncommon finding [3] with only 13 cases reported in the literature (Table 1).

CASE PRESENTATION

A 46 year old woman, non-smoker, complained about cough, fever and fatigue within the last five months.

CT-scan revealed a 4x3x8cm intralobar sequestration in close contact to the diaphragm. Two aberrant arteries originating from the celiac artery entered the lesion and drained into the pulmonary vein. Bronchoscopy showed regular findings. After preoperative coil embolization to reduce the risk of bleeding [4,5], transection of the atypical arteries and resection of both the posterobasal segment and of the tightly adhering diaphragm was

Table 1: Clinical and pathological findings and surgery in intra- and extralobar sequestration combined with pulmonary neoplasia.

Author	Year	Age/ sex	Smoking history	Symptoms	Location of sequestration	Type of sequestration	Type of lung neoplasm	Location of neoplasm	Resection
Hertzog	1963	36 f	no information	pain, malaise	right chest	extralobar	squamous cell carcinoma	within sequester	Resection of sequester
Bell-Thomson	1979	69 m	yes	none	RLL	intralobar	squamous cell carcinoma	within sequester	Left lower lobectomy
Peros	1980	24 f	no information	none	LLL	intralobar	squamous cell carcinoma	within sequester	Left lower lobectomy
Jüttner	1985	45 m	no	cough, fever	LLL	intralobar	atypical carcinoid	within sequester	Left lower lobecomty
Gatzinsky	1988	50 f	no	pain, fever, hemoptysis	RLL	intralobar	adeno-carcinoma	within sequester	Left lower lobectomy
Morita	1994	59 m	no information	fever	LLL	intralobar	squamous cell carcinoma	LUL	1/2 and 3 left Sleeve segmentectomy, 9/10 left segmentectomy
Hekelaar	2000	31 f	no	cough	LLL	intralobar	lymphoepithelioma like carcinoma	within sequester	Segmentectomy 8-10 left
Okamoto	2005	69 m	yes	dyspnea	LLL	intralobar	adeno-carcinoma	LUL	Left upper lobectomy, sequester not removed
Lawal	2011	67 m	yes	pneumonia, hemoptysis	LLL	intralobar	adeno-carcinoma	within sequester	Left lower lobectomy
Ma	2011	39 f	no	pneumonia	LLL	intralobar	atypical carcinoid	within sequester	Left lower lobectomy
Westphal	2012	39 f	no information	pneumonia cough, fever, hemoptysis	RLL	intralobar	typical carcinoid	intermediate bronchus	Right lower bilobectomy
Wang	2013	65 m	no information	none	LLL	intralobar	adeno-carcinoma	adjacent to sequester	VATS left lower lobectomy
Nowak	2013	41 m	no information	hemoptysis	LLL	intralobar	typical carcinoid	within sequester	Left lower lobectomy

Abbreviations: F: Female; M: Male; RLL: Right Lower Lobe; LLL: Left Lower Lobe; LUL: Left Upper Lobe

done. The defect was closed by direct suturing. Intraoperative frozen section histology gave no evidence of malignancy.

Surprisingly, by definitive histopathological examination adenocarcinoma pG3, pT3 within the intralobar sequestration, infiltrating the diaphragm was found. Postoperative staging gave no evidence of tumour spread to the mediastinal lymph nodes or of distant metastases. Nevertheless, according to the oncological guidelines right lower lobectomy and complete mediastinal lymph node dissection were done [6]. Histopathological examination confirmed the absence of lymph node metastases. Adjuvant therapy including four cycles of Cisplatin and Navelbine was scheduled. One year after surgery the patient is well and free of tumor recurrence.

DISCUSSION

Intralobar pulmonary sequestration is a rare congenital disorder, characterized by a dysplastic area of lung tissue localized within one lobe, the bronchi of which may or may not have secondary contact to the deformed bronchi of the sequester. The latter is supplied by systemic arteries and usually drains into the pulmonary vein and/or the systemic ones. The condition may remain asymptomatic or become clinically apparent because of a significant arterio-venous shunt, hemoptoe or recurrent infections in the dysplastic tissue [1-3,7]. The even more uncommon extralobar sequestration also represents nonfunctional lung tissue, which, however, is covered by its own pleura. The blood supply derives from systemic arteries which always drain into the systemic veins [1].

Bronchogenic carcinoma associated to or lying within in a pulmonary sequestration is very rare. To our best knowledge, only thirteen cases of pulmonary neoplasms associated with pulmonary sequestration have been reported. There were four squamous-cell carcinomas [6,8-10], four adenocarcinomas [11-14], one lymphoepithelioma-like carcinoma [15] and four carcinoids [16-19]. Twelve out of the sequestrations were of the intralobar type nine of which involved the left and three the right lower lobe, one sequester was extralobar, localized in the right pleural cavity. Ten out of the thirteen tumours had developed within or in contact to the sequester, in three cases the tumour was coincidental and involved different lobes (Table 1).

The symptoms covered the range of typical findings attributed to intralobar sequestration such as hemoptysis, cough, fever, pneumonia and pain. Two patients were asymptomatic. Neuroendocrine or "carcinoid" tumorlets are often associated with bronchiectasis and scarring and may be considered as a precursor of carcinoids [20]. Various authors have described their occurrence in intralobar sequestration [21-23]. They suggested chronic hypoxia within the sequester as cause for the formation of the tumorlets. The development of a carcinoid tumour in intralobar sequestrations could be explained by this mechanism. Recent data suggest that ongoing inflammatory reactions in the lungs may be the cause of malignant transformation [24-26]. Thus, also considering the fact that at least 40% of the patients with sequestration and lung neoplasia were nonsmokers, a pathogenetic pathway of chronic infection promoting tumor formation can be suspected. For the incidence of pulmonary neoplasia in sequestration only estimates can be made: Within a series of 45 intralobar sequestrations in adults within 20 years Liu reported one case "accompanied by carcinoid", corresponding to an incidence of 2.2%. Larger series of intralobar sequestration have only been reported in infants and children who are unlikely to develop pulmonary neoplasia within the malformation.

Though resection is generally considered as the optimum treatment of pulmonary sequestration, recently also endovascular embolization of the atypical feeding arteries has been advocated [27-30]. In view of the possibility of malignancy developing in sequestration, resection – which can also be done thoracoscopically [3,14] - rather than embolization should be preferred in adults as well as in children [7].

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