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Case report

Adenocarcinoma arising in an extralobar sequestration: a case report and review of the literature

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ABSTRACT

Extralobar sequestration is a type of bronchopulmonary foregut malformation defined as an isolated portion of lung tissue with a systemic arterial supply, its own pleural investment, and no bronchial communication. While it may be recognized *in utero* or in the neonatal period, depending on its location and associated anomalies, it can also go unrecognized until later in life when it may present as a mass. We report the first case of adenocarcinoma arising in an extralobar sequestration. The patient was a 70-year old man with a 55 pack year smoking history who presented with chest discomfort and was found to have a 6.5 cm right lower lobe mass. Percutaneous biopsy of the mass was positive for adenocarcinoma. At surgery, the mass was noted to have a separate arterial connection, no bronchial communication, and its own pleural investment, consistent with an extralobar sequestration. Malignancy arising in pulmonary sequestrations is rare and the few reported cases have been in intralobar types. Carcinoma arising in this setting adds to the dilemma of whether or not these developmental anomalies should be excised or followed. Our tumor, while small, did have vascular invasion.

1. Introduction

Pulmonary sequestrations (PS) form a subset of bronchopulmonary foregut malformations, defined as portions of lung tissue lacking a bronchial connection and possessing their own systemic arterial blood supply. They are further segregated into extralobar (ELS) and intralobar (ILS) variants depending on whether or not they are encased in their own visceral pleura or are enclosed within the normal visceral pleura of the lung [1]. ELS's are more likely than ILS's to be associated with other congenital anomalies such as diaphragmatic hernia or congenital heart disease. While PS's are typically identified early in life, approximately 15% may go unrecognized until adolescence or adulthood. At that time patients may present with hemoptysis, mass effect and secondary symptoms, or signs of infection [2]. We present the first case of malignancy arising in an ELS.

2. Case report

The patient was a 70-year old man with a 55 pack year smoking history who presented with chest discomfort on December 2012. A chest x-ray was read as abnormal and subsequent chest CT scan confirmed the presence of a 6.5 cm right lower lobe mass (Fig. 1A) and subcarinal lymphadenopathy. The lesions were hypermetabolic with an SCV of 2.2. Percutaneous biopsy identified a moderately differentiated adenocarcinoma. The patient was stage IIA (T2BN0M0) by PET/CT and staging mediastinoscopy. Preoperative pulmonary function testing was within normal limits and a thoracoscopic right lower lobectomy was performed.

At surgery, isolated lung tissue with a separate pleural investment and an independent blood supply arising within fatty tissue above the right hemi-diaphragm was identified (Fig. 1B). The fissure between the pulmonary sequestration and the right lower lobe was divided; no communication with the right lung was appreciated—consistent with an extralobar sequestration.

The specimen consisted of a 108 g (post fixation) $8.5 \times 6.0 \times 4.0$ cm segment of lung tissue. The entire specimen was invested by pleura. An 8.0 cm white tan rubbery pleural plaque was present. On sectioning, a firm lesion with an associated 4 mm calcified nodule was noted.

Pathologic examination revealed a 6 mm moderately differentiated TTF-1 positive adenocarcinoma (Fig. 2A). The tumor could be seen arising from a bronchiole (Fig. 2B.) A focus of large vessel invasion was identified (Fig. 2C). Next generation sequencing identified a BRAF-mutation. The background lung parenchyma is distorted with diffuse interstitial fibrosis and pulmonary hypertensive changes (Fig. 2D). An associated congenital pulmonary airway

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Table 1
Patient characteristics.

Age	Sex	Smoker	Presentation	Location	Type	Diagnosis	Location of tumor	Treatment	Follow-up	Additional pathologic findings	Refs.
50	F	N	Non-productive cough, hemoptysis, fever	R	ILS	Adeno	Medial	Lobectomy	None		[3]
69	M	Y	Incidental	R	ILS	Squam	Subpleural	Lobectomy	AWNED, 5 years	Fibrosis, HTN vascular changes, squamous metaplasia	[4]
59	M	?	Fever, infection	L	ILS	Squam	L upper division bronchus	Sleeve segment-ectomy	AWNED 10 months		[5]
65	M	?	Incidental	L	ILS	Adeno	Subpleural	Lobectomy	None		[6]
36	F	?	Chest pain	R	?ELS	Squam	?		Widely metastatic		[7]
31	F	N	Cough, digital clubbing	L	ILS	LEL-EBV+	Hilus	Pneumo-nectomy	AWNED, 4 years		[8]
67	M	Y	Hemoptysis, recurrent pneumonia	L	ILS	Adeno	Proximal margin	Lobectomy	None		[9]
40	M	N	Long standing dysphagia	R	BPFM	Squam	?	Lobectomy	DOD	Fibrosis, HTN vascular changes	[10]
70	M	Y	Chest discomfort	R	ELS	Adeno	Subpleural				This report

Key: M, male; F, female; Y, yes; N, no; ?, unknown; R, right lung; L, left lung; ILS, intralobar sequestration; ELS, extralobar sequestration; BPFM, communicating bronchopulmonary foregut malformation; Adeno, adenocarcinoma; Squam, squamous cell carcinoma; LEL, lymphoepithelial lesion; EBV+, Ebstein-Barr Virus positive; AWNED, alive with no evidence of disease; DOD, dead of disease; HTN hypertensive.

Fig. 1. (A) CT scan of mass (arrow), (B) intraoperative photograph illustrating the vascular pedicle (arrowhead) and the pleural plaque (thick arrow).

malformation (CPAM) was absent. A pleural plaque was noted. A small bronchus with associated cartilage was noted in one section, suggestive of bronchial atresia. Precursor mucinous cells were not identified in the examined sections.

3. Discussion

Malignancy is a rare complication of bronchopulmonary foregut malformations. To our knowledge this is the first report of adenocarcinoma arising in an extralobar sequestration. Our review of the literature revealed seven other cases of carcinoma arising in a pulmonary sequestration and one arising in a sequestered portion of lung tissue with an esophageal communication (communicating bronchopulmonary foregut malformation) (Table 1) [3–10]. Six of these were ILS and in one case the type of sequestration was unclear. Including our case, the patients' ages ranged from 31 to 70, and there were six men and three women. All of the sequestrations were in the lower lobes except for case 3 where the exact location within the left lung was not described. Three of the patients smoked, three were non-smokers and in three the smoking history was not indicated. Five of the tumors were squamous cell carcinomas, three were adenocarcinoma, and one was an EBV associated lymphoepithelioma–like carcinoma. The location of the tumor within the malformation was variable and not mentioned in every case. The patients were treated by surgical resection and most did well. One patient had widely metastatic disease and it remains uncertain if the tumor within the sequestration was a metastasis or primary.

The exact pathway leading to the development of carcinoma in the sequestrations is not known. One hypothesis is that sequestrations are manifestations of an underlying cancer predisposition syndrome like cystic pleuropulmonary blastoma, where there is