Bronchial carcinosarcoma

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ABSTRACT

Carcinosarcoma is an uncommon mixed tumor of the lung. We present the case of a 65 year-old-male with cough and a right lower lobe radio-opacity who underwent resection, showing a large endobronchial tumor with an epithelial component of non-small cell carcinoma and malignant mesenchymal elements. The radiologic and histopathologic features are reviewed with reference to relevant literature.

CASE REPORT

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A 65-year-old man presented for evaluation of a lung mass. His medical history was significant for 4 months of productive cough, low-grade fever and 25 pound weight loss. A chest radiograph showed a very large lobulated right lower lobe opacity (Fig 1). Bronchoscopy at the referring institution demonstrated a large yellow mass in the bronchus intermedius. Brushings only revealed Aspergillus. A non-diagnostic thoracoscopic biopsy was performed. He was referred for further evaluation. A chest computed tomography (CT) scan was obtained with intravenous contrast enhancement. (Siemens Sensation 64, Siemens Medical Solutions, Forcheim, Germany - 3mm slices with 2mm overlap, 100 kV, and 180 mAs). A well-defined, lobulated mass of soft-tissue density was demonstrated in right lower lobe, measuring 9.6 x 7.4 cm in axial plane (Fig 2a,b). Coronal reconstructions better demonstrated the endobronchial tubular and branching distribution of the mass resulting in a "finger-in-glove" appearance (Fig 3a,b). An associated mild to moderate pleural effusion was present. The patient had no history of malignancy and there were no additional parenchymal lesions nor definite pathologically enlarged mediastinal or hilar lymph nodes. Due to the persistent symptomatology, suspicious imaging findings, and underlying severe bronchiectasis in the right lower lobe and a lack of a diagnosis, a right middle and lower lobectomy was performed with partial resection of the diaphragm because of uncertain invasion, followed by mediastinal lymph node dissection.

On gross examination, the tumor replaced most of the right lower lobe, measuring up to 11 cm in diameter (Fig 4). Histopathologic examination revealed a subtype of carcinosarcoma, a rare primary pulmonary malignancy, in this case composed by concurrent presence of poorly differentiated/undifferentiated squamous cell carcinoma, spindle cell foci and malignant mesenchymal elements forming cartilage and bone (Fig 5). Metastases were noted to subcarinal and paraesophageal lymph nodes.

The patient's performance status deteriorated within the first month after surgery, and he died before starting chemotherapy.

DISCUSSION

We present a case of bronchial carcinosarcoma, a malignant biphasic tumor that accounts for less than 1% of all lung cancers. It is defined by coexisting histologic elements of carcinomatous parenchyma and sarcomatous stroma. The WHO classification of this neoplasm states that the epithelial component defines the various subtypes. Among 66 cases reviewed by Koss et al [1], the most common finding was squamous cell carcinoma (69%), followed by adenocarcinoma (20%), and large cell carcinoma (11%) [6]. Unlike spindle cell

carcinoma or homologous sarcomatoid pleomorphic carcinoma whose stromas lack differentiation, mesenchymal components of carcinosarcomas are poorly to moderately differentiated, mostly rhabdomyosarcoma, but also osteosarcoma, chondrosarcoma, a combination of these elements, or less frequently liposarcomatous and angiosarcomatous elements. [2].

These tumors usually present as solitary masses, with a male predominance of 7.25:1, of average age of 65 years and a history of tobacco use [1]. The location correlates with the predilection of the epithelial component. Central polypoid or cast-like growth is the characteristic pattern of the squamous cell type, causing associated symptoms like cough, hemoptysis, chest pain, shortness of breath, and dyspnea. Peripheral locations are more frequent for adenocarcinomas, which tend to be less symptomatic until they become large. Metastasis occurs early and widely, and can be carcinomatous, sarcomatous, or both.

Our patient presented with chronic cough and a large lobulated-tubular opacity on radiograph, which on CT was better shown to create the classic "finger-in-glove" appearance associated to impacted dilated bronchi [3]. The surrounding lung remains aerated by the interalveolar pores and canals of Lambert. This imaging sign has been demonstrated in a heterogeneous group of conditions: aspergillosis, congenital bronchial atresia, bronchiolithiasis, and foreign body aspiration. Less commonly, benign neoplasms such as bronchial hamartoma and lipoma present by luminal obstruction.

The CT findings of bronchial carcinosarcoma share the characteristics of the entirely epithelial neoplasms. [1,2]. The mean size of these tumors is 7 cm, with areas of hemorrhage, pseudocyst formation, and central low-attenuation representing necrosis [1,4]. Tumoral calcifications may imply ossification of the osteosarcomatous component [4].

Prior to resection, the differentiation of carcinosarcoma is difficult. Bronchoscopic bronchial lavage and percutaneous needle biopsy often demonstrate only the epithelial component of the tumor [1,4]. In seven cases presented by Huwer et al, the sarcomatous component was never diagnosed preoperatively [5].

The standard therapy is the combination of resection and external radiation, while chemotherapy is reserved for cases with remote metastasis [6]. The reported 5-year survival rate is only 21.3% [1,4]. The tumor has a marked tendency for distant metastasis and has a high rate of local recurrence. Postoperative survival is 9 months on average [5]. However, tumors with endobronchial localization have been reported to have a better prognosis than peripherally located cases as they cause symptoms earlier. The most important predictors of poor prognosis are a size larger than 5 cm [4] and the presence of a rhabdomyosarcoma component which is thought to play a role in the development of hematologic metastases [5].

In this case report, we describe the radiologic manifestations of an uncommon lung tumor. Diagnosis of this particular type of carcinosarcoma cannot be made on the basis of imaging results alone. Chest CT is presented as the best imaging method for assessing the extent of involvement for preoperative and pretreatment planning.

TEACHING POINT

Carcinosarcoma is a malignant tumor having a mixture of carcinoma and sarcoma, presenting as a solitary mass with the location and features of the epithelial component. The endobronchial proliferation of this tumor creates the appearance of "finger-in-glove" in chest radiograph. Although the CT characteristics are suggested by the epithelial component, definitive anatomopathologic diagnosis is needed for appropriate treatment.

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FIGURES

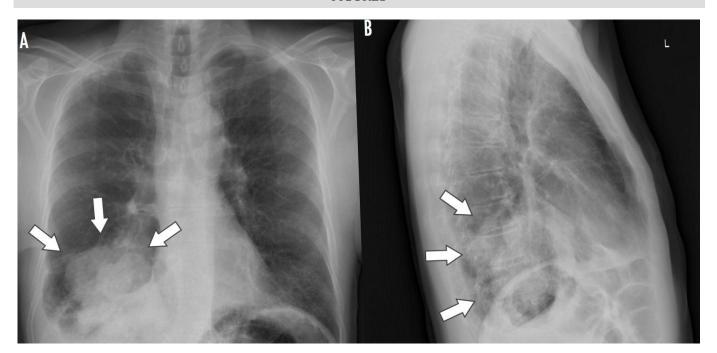


Figure 1: 65 years old male with lung mass that turned out to be bronchial carcinosarcoma. Posteroanterior and lateral chest radiograph show lobulated opacity that radiates from the right lower bronchus (finger-in-glove sign; white arrows), measuring approximately 12 x10 x 13 cm.

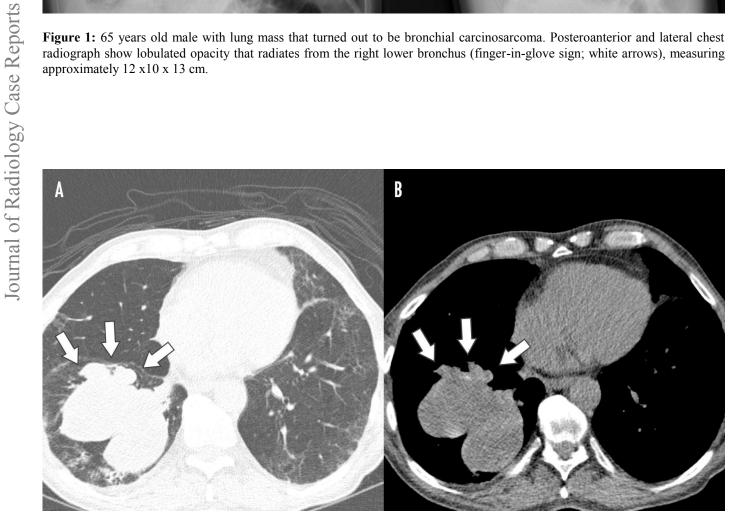


Figure 2: 65 years old male with lung mass that turned out to be bronchial carcinosarcoma. Non-enhanced axial CT in both lung (2A) and soft tissue (2B) window settings shows a low attenuation lobulated lesion in the central portion of the right lower lobe (white arrows) originating from the hilar region. (Siemens Sensation 64 CT scanner, 180 mAs, 100 kV, 3mm slice thickness)

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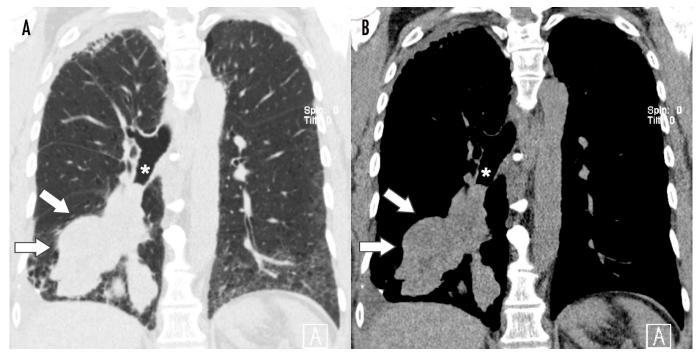


Figure 3: 65 years old male with lung mass that turned out to be bronchial carcinosarcoma. Coronal reconstruction of the non-enhanced CT in both lung (3A) and soft tissue (3B) window settings reveals the multilobulated appearance of the mass arising from the right lower lobe (white arrows) causing markedly dilated bronchi (*). A small right pleural effusion is noted. (Siemens Sensation 64 CT scanner, 180 mAs, 100 kV, 3mm slice thickness)

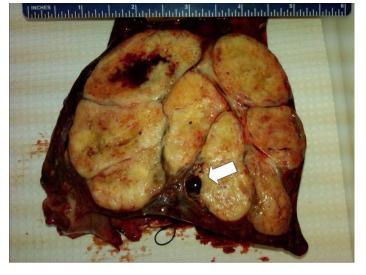


Figure 4: 65 years old male with lung mass that turned out to be bronchial carcinosarcoma. Photograph of gross specimen shows polypoid mass growing in close relationship to bronchial lumen.(white arrow)

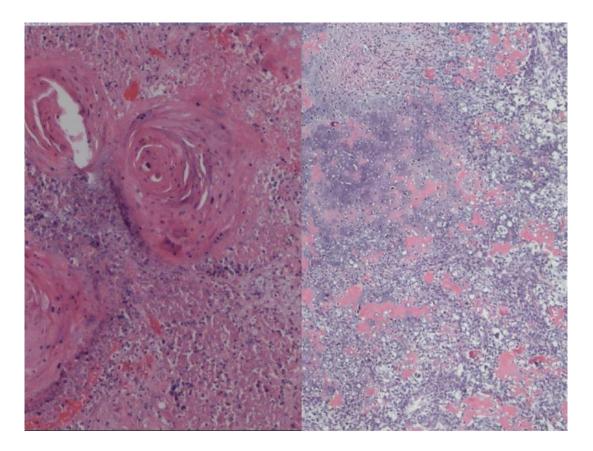


Figure 5: 65 years old male with lung mass that turned out to be bronchial carcinosarcoma. Histologic specimen shows the biphasic portions of the tumor, squamous cell carcinoma component (a), and sarcomatoid heterologous component (b). (H&E stain, 10x)

Etiology	Unknown. It has been associated to tobacco smoking			
Incidence	Accounts for less than 1% of all lung cancers			
Gender ratio	Male:female 7.25:1			
Age predilection	Average age of 65 years			
Risk factors	History of tobacco use			
Treatment	Standard therapy: combination of resection and external radiation. Chemotherapy is reserved f cases with remote metastasis			
D .				
Prognosis	Size larger than 5 cm and the presence of a rhabdomyosarcoma component			
Findings on imaging	Intrabronchial growth associated to impacted dilated bronchi.			

Table 1: Summary of bronchial carcinosarcoma

Disease	Clinical	Plain x ray	CT	Preoperative anatomopathology
Lung carcinosarcomas:	Chest pain, shortness of breath, and dyspnea.		Pseudocyst formation, and central low-attenuation+/-calcifications	Sarcomatous component hardly ever diagnosed
-squamous cell type	Cough, hemoptysis	Central polypoid opacity	Endobronchial opacity	Epithelial component
-adenocarcinoma type	Later presentation of symptoms: chest pain, shortness of breath, dyspnea	Peripheral consolidation	Consolidation with air bronchogram opacity in the periphery of lung	Adenomatous component
Aspergillosis	Shortness of breath, productive cough, fever	Central polypoid or lobar infiltrate	Centrilobular nodules, central bronchiectasis and mucoid impaction	Inflammatory necrotic background with aggregates of fungi

Table 2: Differential diagnosis table of endobronchial carcinosarcoma squamous cell type.

ABBREVIATIONS

CT = Computed Tomography WHO = World Healh Organization

KEYWORDS

Carcinosarcoma; "Finger-in-glove"; endobronchial tumor

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