Bronchial Carcinoid Tumors of the Thorax: Spectrum of Radiologic Findings

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Learning objectives

- To understand the pathophysiology of bronchial carcinoid tumors.
- To become familiar with the clinical and radiologic findings of bronchial carcinoid tumors.
Background

We report a retrospective study that evaluates 20 patients with bronchial carcinoid tumors: 15 females and 5 males with a mean age of 38.7 years (range, 25-63 years), seen during a four-year period from January 2009 to December 2012. Patients were admitted with variant clinical features: cough, wheezing, hemoptysis, and chest pain. Data confirming the presence of paraneoplastic syndrome due to ectopic production of hormones were not found for any of the patients.

All our patients underwent chest CT scan. The final pathologic diagnosis for all cases was confirmed by review of cytologic or histologic specimens (bronchoscopy or CT-guided biopsy).

Introduction

Bronchial carcinoid tumors are classified as neuroendocrine neoplasms of the lung. They arise in the bronchial and bronchiolar epithelium and may derive from existing Kulchitsky cells, neuroepithelial bodies, or pluripotential bronchial epithelial stem cells (1, 2).

Carcinoid tumors possess an indolent clinical course with infrequent metastases, but they are true malignancies with the potential to cause significant morbidity and mortality (3).

Radiological findings include hilar or perihilar masses, endobronchial nodules, findings related to bronchial obstruction, and peripheral nodules (4).

The histologic diagnosis is made with bronchoscopic biopsy (proximal tumor) or with computed tomography-guided transthoracic needle biopsy (peripheral tumor) (4).

Prognosis is highly dependent on tumor size, histologic subtype, and nodal involvement at initial diagnosis (5).

Epidemiology

In this study, as mentioned before, there were fewer male patients than females (25% males compare to 75% females) and the average age of our patients was 38.7 years; likewise, Melissa et al. 1999, in an article entitled "Thoracic carcinoids: radiologic-pathologic correlation" noted that affected population had significantly lower male to female ratio (6). Results of a study by Descovich P et al. in 2000 showed the M: F ratio of 0.6 and the average age of 42.5 years (7).
According to harpole et al: Bronchial carcinoids affect mainly young patients and there is no relationship to smoking, inhalation of carcinogens, radiation or environmental factors (8).

Clinical Presentation

Our study showed the most frequent symptoms to be cough (70%), hemoptysis (59%), wheezing (53%) and chest pain(50%). It has been noted in the literature that patients with bronchial carcinoids are often symptomatic and the most common symptoms include cough, hemoptysis, wheezing, and pneumonia as a result of central airway involvement. Rare associations of pulmonary carcinoid tumors include carcinoid syndrome, Cushing disease from ectopic adrenocorticotropic hormone (ACTH) production, and acromegaly from ectopic production of growth hormone (3, 9).

However, about 19-51% of the patients are asymptomatic and bronchial carcinoid is diagnosed because of their abnormal findings on chest radiographs (1, 10).

Tumor Location

According to our results, neoplasms were located more in the right lung compared to the left (80% and 20% respectively).

The most common sites in decreasing order of frequency were right middle lobe, right upper lobe, right lower lobe, left upper lobe, and left lower lobe.

The SEER registry (1973-2003) identified the location of BP-carcinoids in 5123 cases (91%) (Fig. 1), of which right-sided lesions were the most common (59.0%), whereas 10.4% were located in the main bronchi (3).

In 2001, a study on 142 patients with pulmonary carcinoids by Fink G et al. showed the neoplasm to be located in the right lung in 60% of patients and in the left lung in 40% (12); the most common sites were the same as our study.

Bronchoscopic examination: (Fig. 2)

On bronchoscopic examination Bronchial carcinoids are usually red-brown to bluish-tan endobronchial masses with a smooth surface. They are often highly vascular and in some circumstances have been reported to bleed considerably when biopsied. Care should therefore be exercised and cautery always available (11).
*Histology*

Bronchial carcinoids arise in the bronchial and bronchiolar epithelium and may derive from existing Kulchitsky cells, neuroepithelial bodies or pluripotential bronchial epithelial stem cells (4).

They are categorized into two major groups: About 90% are typical carcinoid tumors. The remaining 10% of the lesions are atypical carcinoid tumors, which tend to have a higher rate of metastasis and are larger at the time of diagnosis (12).

The WHO diagnostic criteria for TC are: a tumor with carcinoid morphology and <2 mitoses/2 mm² (10 HPH), lacking necrosis, and tumor 0.5 cm or larger. An AC is defined as a tumor with carcinoid morphology with 2 to 10 mitoses/2 mm² and/or necrosis (13). *(Fig. 3)*

Our study shows typical and atypical carcinoids in respectively 85% and 15% of the patients.
Images for this section:

Fig. 1: Distribution of 5123 lung carcinoids, registered in the Surveillance Epidemiology and End Results (SEER) registry. The anatomical locations are main bronchi, upper, middle, and lower right lung lobes, and upper and lower left lobes

Fig. 2: Typical carcinoid obstructing the medial segment of the right middle lobe bronchus

<table>
<thead>
<tr>
<th>Tumor Type</th>
<th>Criteria</th>
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</thead>
<tbody>
<tr>
<td>Typical carcinoid tumors</td>
<td>Carcinoid morphology and &lt;2 mitoses/2mm² (10 HPFs), no necrosis and &gt;0.5 cm</td>
</tr>
<tr>
<td>Atypical carcinoid tumors</td>
<td>Carcinoid morphology with 2-10 mitoses/2mm² (10 HPFs) or necrosis (often punctual)</td>
</tr>
</tbody>
</table>

**Fig. 3:** 2004 WHO Criteria for Diagnosis of Neuroendocrine Tumors

Findings and procedure details

The imaging features of typical and atypical carcinoids are too similar to be separated. Although the location of carcinoids varies, the majority are centrally located and therefore related to airways.

Chest Radiography

Plain X-rays are nonspecific, although BP-carcinoids often appear as:

- An isolated, well-defined hilar or perihilar mass (3, 14).
- Associated parenchymal change as a consolidation, which is suggestive of atelectasis, obstructive pneumonitis, or recurrent pneumonia may be also seen in some cases (14).

CT scans

Once a suspicious lesion is detected on a chest X-ray, a computed tomography (CT) of the chest and upper abdomen should be undertaken to determine the size, characteristics, extent of the primary tumor, involvement of mediastinal lymph nodes, and presence of distant metastases (3).

CT scans findings include hilar or perihilar masses, endobronchial nodules, findings related to bronchial obstruction, and peripheral nodules (4).

Review of CT scans in our study revealed the most frequent finding to be a well-defined and round border mass with a close anatomic relationship to the bronchus and distal parenchymal disease (60%). Also, endobronchial nodule (25%), Solitary pulmonary nodule in the lung periphery distal to the segmental bronchi (15%) was other frequent findings.

Hilar or Perihilar Masses

- Central bronchial carcinoids most frequently manifest as a hilar or perihilar mass. The mass is usually a well-defined round or ovoid lesion that narrow, deform, and/or obstruct airway (3,15,16). The tumors range from 2 to 5 cm in size (15, 16). They are located close to central bronchi, often near the bifurcation area (17). (Fig. 4,5,6)
- Calcification is evident in up to 30% of tumors and manifests in a punctuate or diffuse pattern (3). (Fig. 7)

- Carcinoids tend to be vascular with marked, homogeneous contrast enhancement (3,4). Furthermore, atypical carcinoids may have irregular contours and less uniform contrast enhancement (18). (Fig. 8)

- Both typical and atypical carcinoids may be associated with hilar infectious or malignant lymphadenopathy (3). (Fig. 9)

**Endobronchial Nodules**

- Central carcinoids often demonstrate radiologic evidence of an endobronchial Nodules component (Fig. 10). Although most carcinoids are primarily endobronchial lesions, they may only extend into the adjacent parenchyma (Fig. 11). Such tumors may display a dominant extra-luminal component with a very small endobronchial portion ("iceberg" lesion) (19). (Fig. 12)

- Some small carcinoids are located entirely within the bronchial lumen. CT can demonstrate that these lesions are entirely intraluminal without extraluminal components (4). (Fig. 13)

**Findings Related to Bronchial Obstruction**

Findings related to bronchial obstruction such as atelectasis (Fig. 14), air trapping, obstructing pneumonitis (Fig. 15), and mucoid impaction (bronchocele, mucocele) may also be seen. (Fig. 16, 17)

**Peripheral Nodules**

- In about 20% of cases, a bronchial carcinoid manifests as a solitary pulmonary nodule in the lung periphery distal to the segmental bronchi. These lesions are also usually round or ovoid with smooth or lobulated borders (4). (Fig. 18)

- Typical carcinoids in the periphery are slow growing and should be considered in the differential diagnosis of slow-growing solitary pulmonary nodules (15).
- Atypical carcinoids are more likely to occur in the lung periphery and are usually large (20).

**MRI**

- MRI is not a routine diagnostic modality for lung imaging and is usually utilized to resolve ambiguous CT findings (21).

- MRI should be considered when an ACTH-producing carcinoid is suspected but not found at CT. MRI may also be useful if there is concern about neural foramen or brachial plexus involvement (3).

- Bronchial carcinoids have high signal intensity on T2- weighted and short-inversion-time inversion recovery MR images, thus MR imaging may be helpful in distinguishing small carcinoids from the adjacent normal vascular structures (22).
**Fig. 4:** Typical carcinoid in a 36-year-old woman with hemoptysis. Contrast-enhanced CT scan demonstrates a well-defined ovoid, perihilar mass with narrowing of the intermediate bronchus (arrow).

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Fig. 5: Typical carcinoid in a 48-year-old man with a 3-month history of hemoptysis. Contrast-enhanced Chest CT with mediastinal window shows a well-defined ovoid mass close to carina (arrow).

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**Fig. 6:** The same case as Fig. 5 Images of VR reconstruction of trachea and bronchial tree show a narrowing of the carina (blue arrow) and the right main bronchus (red arrow).

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Fig. 7: Typical carcinoid in a 46-year-old woman with a 1-year history of cough and chest pain. CT scan (lung and mediastinal windowing) demonstrates a well-defined ovoid perihilar mass (blue arrow), located between the lower and middle lobes and containing diffuse calcification (red arrow), with a close anatomic relationship to the intermediate bronchus.

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Fig. 8: Typical Central carcinoid in a 46-year-old woman with hemoptysis who had given birth 5 months earlier. Contrast-enhanced chest CT scan (mediastinal windowing) shows a right hilar ovoid mass with marked and homogeneous contrast enhancement (arrow). The lesion narrows the intermediate bronchus.

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Fig. 9: Typical Central carcinoid in a 65-year-old woman with hemoptysis. Contrast-enhanced chest CT scan (mediastinal windowing) shows a right hilar ovoid mass (blue arrow) with a left hilar adenopathy (red arrow).

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Fig. 10: Typical carcinoid in a 46-year-old man with a 1-year history of cough and wheezing. Contrast-enhanced chest CT scan (mediastinal windowing) shows a well-circumscribed, ovoid, endoluminal nodule (arrow).

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**Fig. 11:** Typical carcinoid in a 42-year-old woman with a 3-month history of cough and hemoptysis. Contrast-enhanced chest CT scan (mediastinal windowing) shows a well-defined ovoid mass in the right upper lobe bronchus (blue arrow) extending into the adjacent parenchyma (red arrow) with evidence of consolidation of the upper lobe (asterisk).

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**Fig. 12:** Typical carcinoid in a 60-year-old woman with a 6-month history of cough and chest pain. Contrast-enhanced chest CT scan (mediastinal windowing) shows a dominant extra-luminal component (red arrow) with a small endobronchial portion (blue arrow) "iceberg" lesion"

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**Fig. 13:** 56-year-old woman with a 1-year history of cough and hemoptysis. Chest CT scan (lung windowing) shows a well-circumscribed, ovoid, entirely endoluminal nodule (blue arrow).

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Fig. 14: Atypical carcinoid in a 53-year-old woman with a 3-year history of cough and recurrent pneumonia. CT scan (lung and mediastinal windowing) shows a well-circumscribed lobular mass with slight enhancement in the lung periphery (arrow) that close the right lower lobar bronchus producing complete atelectasis (asterisk).

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**Fig. 15:** Atypical carcinoid in a 67-year-old man with recurrent pneumonitis. Contrast-enhanced CT scan (mediastinal windowing) shows a lobulated mass with slight enhancement in the right lower lobe (blue arrow) with distal pneumonitis (red arrow).

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**Fig. 16:** Typical carcinoid in a 36-year-old woman with a 2-year history of cough and recurrent pneumonitis. Contrast-enhanced Chest CT with mediastinal window demonstrates a single mass in the left lung base (blue arrow) that causes narrowing of the left lower bronchus. Chest CT with lung window shows a several bronchiectasis distal to the tumor site, some occupied by mucus (red arrow).

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**Fig. 17:** Typical carcinoid in a 32-year-old woman with a 8-months history of recurrent pneumonitis. Contrast-enhanced Chest CT (mediastinal window) demonstrates a right perihilar mass (blue arrow) with distal bronchocele (red arrow).

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**Fig. 18:** 55-year-old woman with peripheral typical carcinoid tumor at lobectomy. CT contrast enhancement of right lower lobe nodule. A, Unenhanced CT scan shows density of nodule (arrow) is 5 HU. B, CT scan obtained 60 seconds after contrast administration shows density of nodule is 78 HU.

© Meisinger et al. CT Features of Peripheral Pulmonary Carcinoid Tumors. AJR:197, November 2011
Conclusion

Bronchial carcinoids are neuroendocrine tumors that range from low-grade typical carcinoids to more aggressive atypical carcinoids.

It is imperative that radiologists understand the spectrum of imaging features to facilitates accurate diagnosis and helps optimize surgical planning.
References


