

# Bronchial carcinoid tumors: A rare malignant tumor

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## Abstract

Bronchial carcinoid tumors (BCTs) are an uncommon group of lung tumors. They commonly affect the young adults and the middle aged, the same age group affected by other more common chronic lung conditions such as pulmonary tuberculosis. Diagnosis is commonly missed or delayed due to a low index of suspicion. Surgery is the mainstay of treatment with an excellent outcome. There are many reports of this rare group of tumors in the Western and Asian regions. The only report around our sub-region is a post mortem report of an atypical variant. We wish to report a case of the typical variant and increase our index of suspicion. A 25-year-old male presented with a 4 years history of cough and haemoptysis. He was repeatedly treated for bronchial asthma and pulmonary tuberculosis with no improvement of symptoms. Chest X-ray and chest computed tomography scan revealed a left upper lobe tumor. Histology reported a typical variant of BCT which was confirmed by immunohistochemistry. He had a left upper lobectomy and has done excellently well thereafter. A high index of suspicion is needed to reduce the risk of missing or delaying the diagnosis.

**Key words:** Bronchial carcinoid tumor, diagnosis, outcome, treatment, West Africa

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## Introduction

Bronchial carcinoid tumors (BCTs) are an uncommon group of lung tumors. The first report of BCT was by Laennac in 1831. They represent about 1% of all primary lung tumors. About 80% are centrally located, within the lobar and segmental bronchi.<sup>[1,2]</sup> BCTs were initially classified as benign tumors but now more appropriately classified as malignant tumors. They arise from the Kulchitsky cells in the bronchial mucosa, more often in the cartilaginous portion of the tracheobronchial tree, covered by intact epithelium. They are highly vascular and may have a broad based or polypoid attachment. BCTs are capable of secreting biologically active substances. Over 80% of the tumors express somatostatin receptor subtype 2.

The cause of BCTs is not proven and there is no strong association with smoking and environmental carcinogens. Familial BCTs have been reported with or without multiple endocrine neoplasia type 1. Leotlela *et al.* attributed BCTs to loss of heterogeneity in multiple chromosomes.<sup>[3]</sup>

The World Health Organization classifies BCTs into two groups. Typical BCTs are well differentiated with very low malignant potential, have <2 mitotic figures/10 high power field and no evidence of necrosis. Atypical BCT have >2–10 mitotic figures/10 high power field and evidence of necrosis. Typical BCTs are about four times more common than atypical BCTs.

We are reporting a case of typical BCT since none has been reported before in the sub-region, to the best of our knowledge.

## Case Report

Mr. O.O., a 25-year-old graduate, Ibo, from SE, Nigeria presented at the Cardiothoracic Unit, University of Nigeria Teaching Hospital, Enugu, Nigeria with a 4 years history of cough, haemoptysis, shortness of breath and left sided

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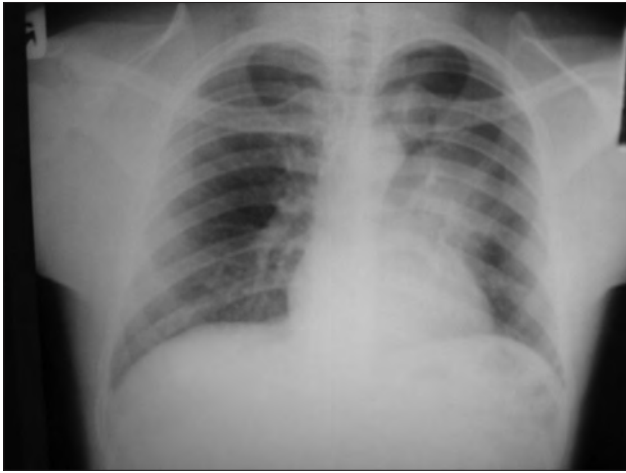
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**Figure 1:** Chest X-ray showing left peri-hilar ground glass consolidation extending laterally



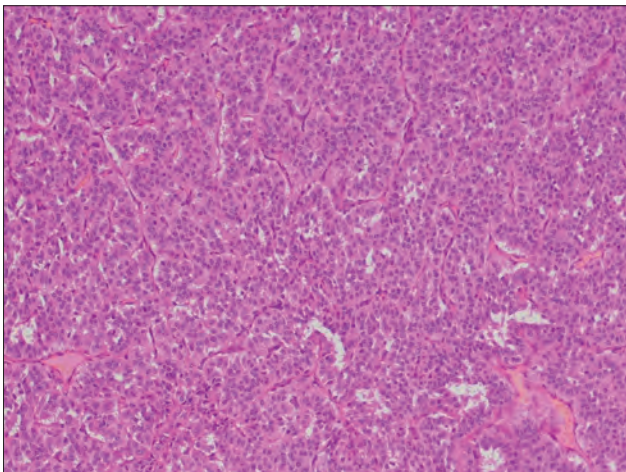
**Figure 2:** Axial computed tomography at the level of tracheal bifurcation showing an oval, enhancing left upper lobe mass



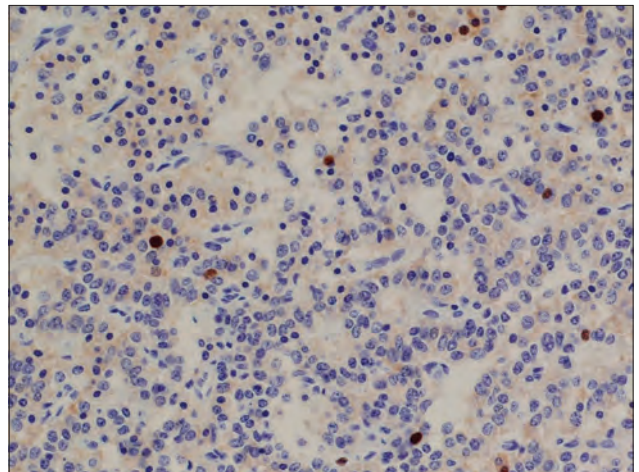
**Figure 3:** Axial computed tomography at the level of main stem bronchus; note the close relationship between the left main stem bronchus and the mass



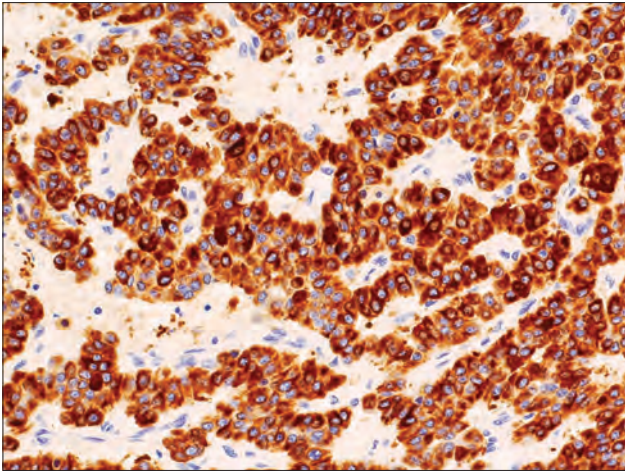
**Figure 4:** Postoperative chest X-ray showing the left lower lobe, unaffected by the tumor and filling up the left hemi-thorax



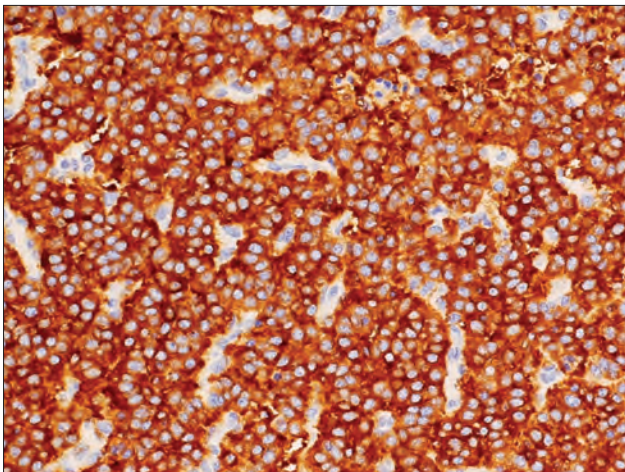
**Figure 5:** Proliferation of uniformly small round to oval cells forming nests, rosettes and small acinar structures. The stroma is scanty and vascular. There are no foci of necrosis (H and E,  $\times 40$ )



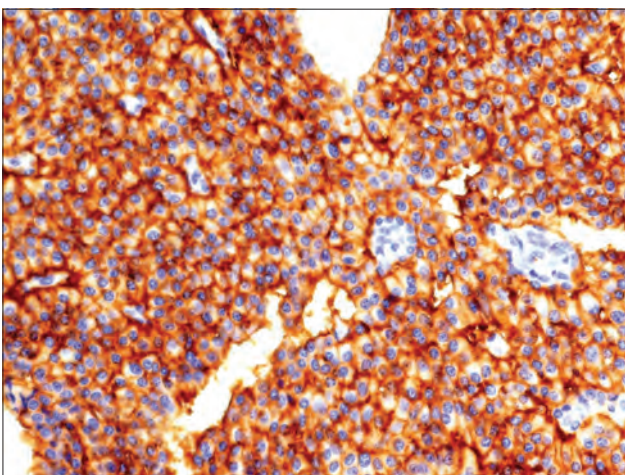
**Figure 6:** Poor staining of the tumor cells for Ki-67 ( $\times 400$ ). The Ki-67 index is about 1%



**Figure 7:** Strong staining of the tumor cells for broad spectrum cyto-keratins (AE1/3,  $\times 200$ )



**Figure 8:** Strong staining of the tumor cells for (synaptophysin,  $\times 200$ )



**Figure 9:** Strong staining of the tumor cells for CD56 (N-CAM,  $\times 200$ )

chest pain. There was no weight loss or excessive sweating. He is not a known asthmatic and never smoked. He had

a complete course of anti-tuberculosis medication, despite screening negative for pulmonary tuberculosis, without any improvement. On examination, he looked well-nourished and active. There was a dull percussion note and a reduced air entry in the left upper lung zone. Chest X-ray (CXR) showed a significant left hilar opacity [Figure 1]. Chest computed tomography (CT) scan revealed an oval mass in the left upper lobe with a widest diameter of 6.19 cm obstructing the left upper lobar bronchus with collapse of the upper lobe segments. There was no enlargement of the left hilar or mediastinal lymph nodes [Figures 2 and 3]. Histology of a trans-thoracic needle biopsy specimen reported a typical BCT [Figure 4]. His lung function test and cardiac performance status were favorable. He had a left upper lobectomy with regional lymph node dissection through a left postero-lateral thoracotomy. At surgery, a firm nodular left upper lobe mass measuring 7 cm in the widest diameter with areas of atelectasis and bronchiectasis was seen. There were no enlarged hilar or mediastinal lymph nodes. The left lower lobe was free of any tumor [Figure 5]. Histology of surgical specimen reported a typical BCT (central type) with tumor free resection margins and no lymph node involvement. They tumor cells strongly expressed broad spectrum cytokeratins, synaptophysin, CD56 and neuron-specific enolase on immunohistochemistry [Figures 6-9]. There was no expression of chromogranin and thyroid transcription factor-1.

## Discussion

The patient presented with the classical features widely described in the literature. There is no such report from our sub-region so far. The reported global incidence is about 0.2–3 per 100,000 per year and appears to be rising because of improved imaging techniques.<sup>[2]</sup> There is no wide sex or racial variation. Typical BCTs have been reported in all age groups but peak around fourth decade while atypical BCTs occur more around the fifth decade.<sup>[1,2]</sup> Central tumors are usually symptomatic with features of bronchial obstruction, irritation and ball valve effect for polypoid tumors, while peripheral tumors are usually asymptomatic. They can present with carcinoid syndrome, Cushing's syndrome, acromegaly, etc., CXR reveals the tumor mass, parenchyma changes due to obstruction and/or evidence of pleural effusion. Chest CT scan better defines the tumor mass, location, relationship with the tracheobronchial tree and the lymph node status. There is a greater contrast enhancement for BCTs than with other lung tumors due to the higher vascularity and tumor portions outside the airway are better appreciated than with bronchoscopy. Bronchoscopy offers the best choice for tissue diagnosis. Tumor resection, laser or photodynamic therapy can be offered at the same setting. Trans-thoracic needle biopsy, video-assisted thoracic surgery or thoracotomy are other

options for tissue diagnosis. Histology is by routine light microscopy and confirmed by immunohistochemistry. Surgery is the mainstay of treatment.<sup>[2,4]</sup> Lung resection and lung parenchymal sparing bronchoplastic procedures with regional lymph node dissection have been offered with excellent outcomes especially for typical BCTs. Chemotherapy and radiotherapy are not yet convincing.<sup>[2]</sup> 5 years survival for typical BCTs is over 80% and about 50% for atypical BCTs. Recurrence is very low, about 3–5%, and is usually due to incomplete resection.<sup>[1,4]</sup>

Bronchial carcinoid tumors commonly affect the same age group affected by other more common chronic lung conditions such as pulmonary tuberculosis. Manash *et al.* in 2012 reported BCT as a rare differential of pulmonary tuberculosis.<sup>[5]</sup> Dixit *et al.* in 2009 reported a case of BCT with concomitant pulmonary tuberculosis.<sup>[6]</sup> This underscores the need for a good index of suspicion with the resurgence of pulmonary tuberculosis and the rising incidence of BCTs.

Diagnosis is missed or delayed even in symptomatic patients due to a low index of suspicion, paucity of knowledge and lack of facilities for accurate diagnosis. Faduyile *et al.* in 2012 reported a postmortem case of a 53-year-old man who died from atypical BCT while being wrongly treated for pulmonary tuberculosis.<sup>[7]</sup> In many reports diagnosis is made more than 1-year after presentation. This may be worse in our setting due to earlier highlighted reasons, poor

referral system and the high tendency to resort to alternative health care services.

## Conclusion

Bronchial carcinoid tumors also occur in our sub-region. A good index of suspicion and knowledge of the tumor is very important for a good outcome.

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