

Recurrent Atypical Carcinoid Tumor After Eight Years: A Case Report

Sekiz Yıl Sonra Rekürrens Gelişen Atipik Karsinoidli Bir Olgu

Hasan ÇAYLAK, MD,^a
Kuthan KAVAKLI, MD,^a
Sedat GÜRKÖK, MD,^a
Orhan YÜCEL, MD,^a
Armağan GÜNAL, MD^b

Departments of
^aThoracic Surgery,
^bPathology,
Gulhane Military Medical Academy,
Ankara

Geliş Tarihi/Received: 15.10.2008
Kabul Tarihi/Accepted: 19.11.2008

Yazışma Adresi/Correspondence:
Kuthan KAVAKLI, MD
Gulhane Military Medical Academy,
Department of Thoracic Surgery,
Ankara,
TÜRKİYE/TURKEY
dr_kuthan_78@hotmail.com

ABSTRACT Bronchial carcinoid tumors comprise 25% of all carcinoids, but account for 2% of all primary lung cancers. They are classified typical and atypical histologically. Atypical carcinoids are aggressive tumors and tend to metastasize and recur more frequently according to typical carcinoids. Late nodal recurrence of an atypical pulmonary carcinoid tumor from initial resection is very rare. Herein, we presented a case of nodal recurrence of atypical carcinoid tumor in a 40-year-old man who was diagnosed after an insisting approach from 8 years after initial resection and treated with surgery successfully.

Key Words: Neuroendocrine tumors; recurrence; thoracic surgery

ÖZET Bronşial karsinoid tümörler tüm karsinoid tümörlerin %25'ini oluşturur ancak primer akciğer kanserleri için bu oran %2'dir. Histolojik olarak tipik ve atipik olarak sınıflandırılırlar. Atipik karsinoid tümörler tipik karsinoid tümörler göre daha agresif tümörlerdir ve metastaz yapma eğilimleri ve rekürrensleri daha siktir. Atipik karsinoid tümörlerin ilk rezeksiyondan sonra geç nodal rekürrensleri nadirdir. Bizde bu yazıda, ilk rezeksiyondan 8 yıl sonra ısrarlı bir yaklaşım ile tanı konan ve başarılı bir şekilde tedavi edilen nodal rekürrens gösteren atipik karsinoidli 40 yaşında erkek bir olguyu sunduk.

Anahtar Kelimeler: Nöroendokrin tümör; rekürrens; torasik cerrahi

Türkiye Klinikleri Arch Lung 2009;10(1):22-5

Bronchial carcinoid tumors are malignant neoplasms which they are currently included in the spectrum of neuroendocrine proliferations and neoplasm of the lung according to WHO in 1999. Typical Carcinoid (TC) and Atypical Carcinoid (AC) tumors represent low-grade malignancy and intermediate-grade malignancy respectively.^{1,2} TCs have good prognosis with >%87 of patients surviving for 10 years. In contrast, ACs have poor prognosis with approximately %25 to %69 of patients surviving for 5 years.³⁻⁵ Recurrence of carcinoid tumors after surgical resection varies between 2-9% and 5-30% for typical and atypical histologies, respectively. Systemic metastases can be seen particularly in patients with AC with thoracic lymph node involvement within a median time of 17 months.⁶ Nodal recurrence of an atypical pulmonary carcinoid tumor after 8 years from initial resection is a rare entity in the literature. Herein, we presented a case

of late nodal recurrence of AC who was previously treated with surgery.

CASE REPORT

In September 1999, a 40-year-old man admitted with a solitary pulmonary nodule on chest roentgenogram which was discovered incidentally. The computerized chest tomography (CT) revealed a nodule 2 cm in diameter situated in the periphery of the right lung. He underwent right exploratory thoracotomy and the nodule was excised. The frozen section study was reported as neuroendocrine tumor and we achieved middle lobectomy. Mediastinal lymph nodes dissection was not performed because there was no lymph nodes in pathologic appearance. The final pathologic examination revealed atypical carcinoid tumor (Figure 1a). No additional treatment was given and the patient has been followed up with three months intervals for first year, six months intervals for second year and then yearly as our clinic policy. After 8 years from initial resection a lower paratracheal lymph node in 2 cm diameter was determined on CT (Figure 2a). Positron emission tomography (PET) scan revealed that the mass exhibiting a 3.7 of standardized up value (SUV). Cervical mediastinoscopy was performed to get histopathologic diagnosis but it was not confirmed a recurrent atypical carcinoid tumor and re-

ported as a reactive lymphadenopathy. After 6 months from mediastinoscopy the patient was asymptomatic but there was a 5 cm diameter of mass on CT in the same region (Figure 2b). The level of 5-hydroxyindolacetic acid in 24-hour urine was elevated to 120 Mmol (normal value 10-31 Mmol). In September 2007, the lymph node was removed completely via right thoracotomy (Figure 2d). The radical mediastinal lymph nodes dissection was not performed because there was not any lymph nodes except the mentioned lymph node. Thirteen cycle of radiotherapy (39 Gray) was performed as an adjuvant treatment. The histopathologic findings were similar with the primary tumor which was resected 8 years previously (Figure 1b). Histopathological examination of the tumor revealed seven mitoses per 2 mm² (10 high-power field) and necrosis. The patient is doing well and there were no pathologic findings on CT and the level of 5-hydroxyindolacetic acid in 24-hour urine measurement in the last follow-up after 12 months from surgery (Figure 2c).

DISCUSSION

Bronchial carcinoid tumors consist of 1 to 2% of all lung malignancies. The pathologic stage and atypical histology are the most important factors affecting survival.^{3,7} Recurrence is more frequently seen for AC according to TC and usually seen as syste-

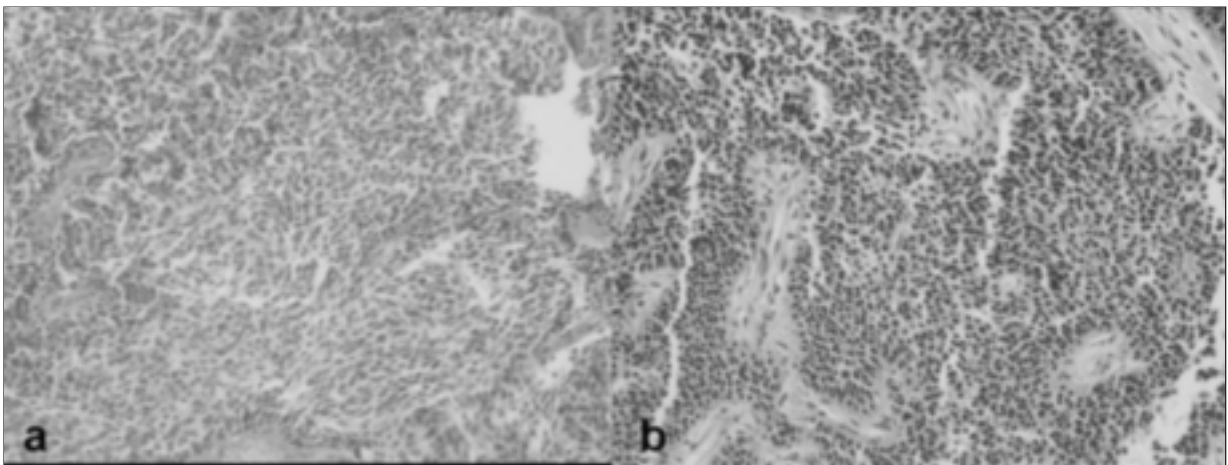


FIGURE 1: Primary (a) and metastatic (b) tumors, those have same neuroendocrine phenotype (HE, x200).

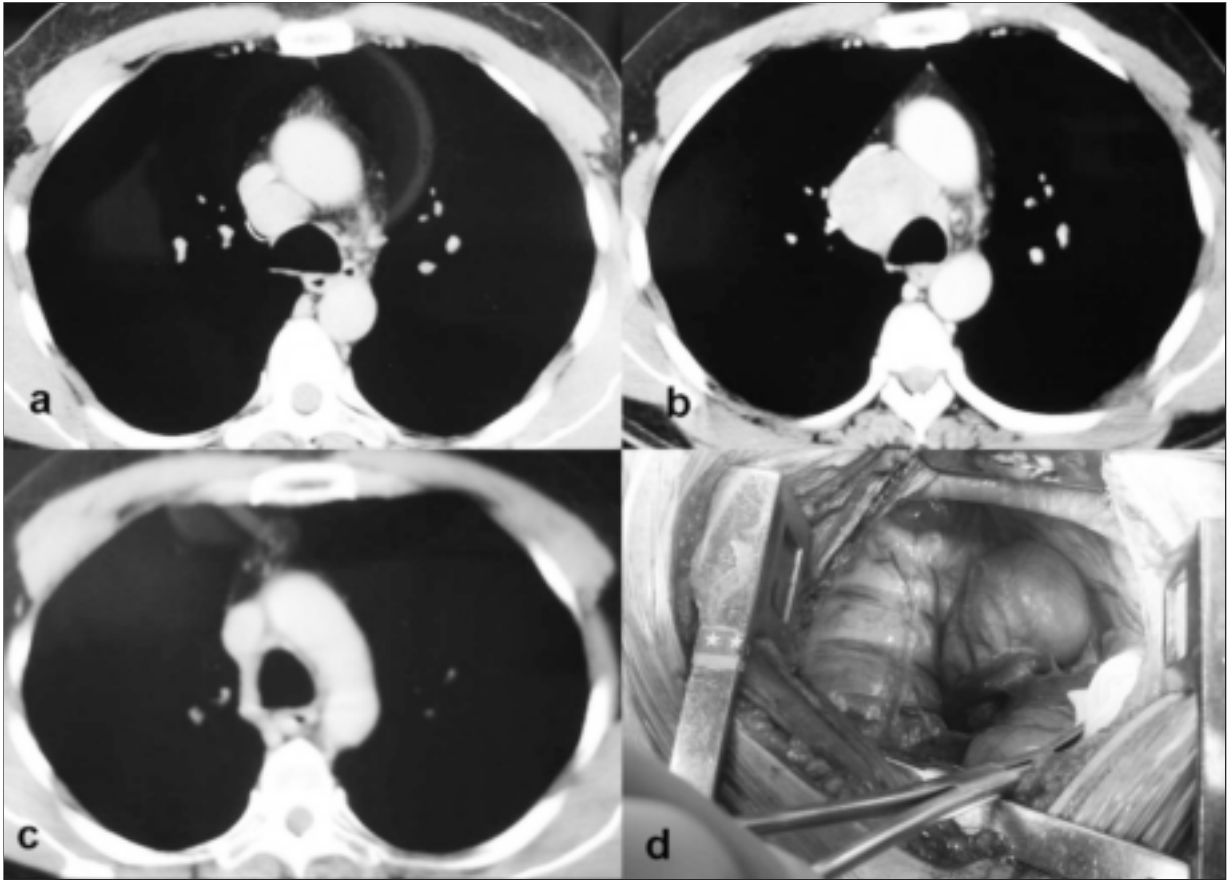


FIGURE 2: Computerized chest tomography revealed a 2 cm sized lymphadenopathy at lower paratracheal region (a), CT image of the 5 cm sized lymphadenopathy discovered at the same region six months later (b), Normal CT image after 12 months from resection (c), Intraoperative view of the resected lymphadenopathy (d).

mic metastases. In a study, patients with typical carcinoid tumors with thoracic lymph node metastases did well after surgery alone and recurrence was seen in 8.7% of 23 patients. In contrast, patients with atypical carcinoid tumor with thoracic lymph node metastases were treated with surgery alone had recurrence rate of 63% of 11 patients.⁶ Schreurs AJ and et al emphasized that presence of lymph node metastases in 9 of 93 patients with TCs patients (all with N1 disease) did not have any prognostic significance.⁸ So the atypic histologies tend to metastase and nodal recurrence more frequently and require close follow up. In our case, although mediastinoscopy was negative for nodal recurrence, insisting approach achieved and successful surgical resection was carried out after six months.

PET has been recently added in the workup of patients with non-small cell cancer. Bronchial carcinoid tumors demonstrate lower uptake than non-small cell lung cancer and it appears to be unreliable in the differentiation of benign process from nonbenign ones.⁹ OctreoScan (using octreotide radiolabeled with Indium111) demonstrates reliable results both in diagnosis of primary carcinoids and detection of early recurrences and metastases even in asymptomatic patients with a sensitivity of more than 90%.¹⁰ However this diagnostic tool is not available for every institutions. The level of 5-hydroxyindolacetic acid in 24-hour urine is an another useful diagnostic method. In our case, there was mass which was growing for 6 months interval on CT images, PET scanning revealed a lower uptake and the level of

5-hydroxyindolacetic acid in 24-hour urine was elevated. All of these findings suggested that the lesion can be the nodal recurrence of AC which was resected 8 years before although mediastinoscopy was negative.

The curative treatment modalities are surgical resection of primary tumor and lymph node dissec-

tion which also gives very important prognostic informations. Bronchial carcinoid tumors have poor response to standard chemotherapy and radiation therapy. So after a adequate surgical treatment including anatomic resection and hilar and mediastinal lymph node dissection, life long-follow up is very important.

REFERENCES

1. Travis WD, Colby TV, Corrin B, Shimasato Y, Brambilla E, Sobin LH. *Histological Typing of Lung and Pleural Tumours* (WHO. World Health Organization. *International Histological Classification of Tumours*). 3rd ed. Berlin: Springer-Verlag; 1999. p.498-9.
2. Hage R, De la Riviere AB, Seldenrijk CA, Van den Bosch JMM. Update in pulmonary carcinoid tumors: a review article. *Ann Surg Oncol*. 2003;10(6):697-704.
3. Travis WD, Rush W, Flieder DB, Falk R, Fleming MV, Gal AA, et al. Survival analysis of 200 pulmonary neuroendocrine tumors with clarification of criteria for atypical carcinoid and its separation from typical carcinoid. *Am J Surg Pathol* 1998;22(8):934-44.
4. Vadasz P, Palfy G, Egervary M, Schaff Z. Diagnosis and treatment of bronchial carcinoid tumors: clinical and pathological review of 120 operated patients. *Eur J Cardiothorac Surg* 1993;7(1):8-11.
5. Filosso PL, Rena O, Donati G, Casadio C, Ruffini E, Papalia E, et al. Bronchial carcinoid tumors: surgical management and long-term outcome. *J Thorac Cardiovasc Surg* 2002;123(2):303-9.
6. Thomas CF, Tazelaar HD and Jett JR. Typical and atypical pulmonary carcinoids: Outcome in patients presenting with regional lymph node involvement. *Chest* 2001;119(4):1143-50.
7. Gould PM, Bonner JA, Sawyer TE, Descamps C, Lange CM, Li H. Bronchial carcinoid tumors: importance of prognostic factors that influence patterns of recurrence and overall survival. *Radiology* 1998;208(1):181-5.
8. Schreurs AJ, Westermann CJ, van den Bosch JM, Vanderschueren RG, Brutel de la Rivière A, Knaepen PJ. A twenty-five-year follow-up of ninety-three resected typical carcinoid tumors of the lung. *J Thorac Cardiovasc Surg* 1992;104(5):1470-5.
9. Erasmus JJ, McAdams HP, Patz EF Jr, Coleman RE, Ahuja V, Goodman PC. Evaluation of primary pulmonary carcinoid tumors using FDG PET. *AJR Am J Roentgenol* 1998;170(5):1369-73.
10. Hauso O, Gustafsson BI, Kidd M, Waldum HL, Drozdov I, Chan AK, et al. Neuroendocrine tumor epidemiology: contrasting Norway and North America. *Cancer* 2008;113(10):2655-64.