

Metastatic Thymic Carcinoid: Does Surgeon Have a Primary Role?

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Abstract

Thymic carcinoids are rare mediastinal tumours. These are aggressive tumours that often present late and have poor prognosis. Primary surgical treatment is recommended even in metastatic tumours since the role of adjuvant therapy is not well established. We present a case of metastatic thymic carcinoid managed with surgical excision.

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Key words: Neuroendocrine carcinoma, Thymic carcinoid, Metastasis.

Introduction

Thymic carcinoids are rarely reported malignancies. Since these were categorised as a separate entity in 1972,¹ some series have been published, the largest being from Mayo clinic.² Patients with thymic carcinoid tumour can be asymptomatic or present with symptoms of local invasion or endocrine syndromes. We discuss the clinical profile and surgical management of a patient diagnosed with thymic carcinoid.

Case Report

A 40-year-old labourer presented with complaints of right sided chest pain. Although the pain subsided with analgesics, an abnormal chest radiograph (Figure 1) prompted referral to our centre. Clinical examination revealed a significant left supra-clavicular lymphadenopathy. Respiratory system evaluation was normal except for a mild reduction of air entry on the right side. He underwent contrast-enhanced computed tomography (CECT) which revealed a well-defined anterior mediastinal mass partly compressing the right main bronchus and superior vena cava (Figure 2). Further evaluation with positron emission tomography-computed tomography (PET-CT) delineated a large well-defined heterogeneous mass having increased flurodeoxy glucose (FDG) uptake with multiple areas of necrosis and calcification. The left supra-clavicular node also showed increased uptake of FDG. Rest of the thoracic and abdominal viscera were normal. Fine needle aspiration cytology (FNAC) from the mass raised a suspicion of a neoplastic lesion. Histopathological examination of the core biopsy obtained from the mass was suggestive of a thymic neuro-endocrine carcinoma with diffuse positivity for synaptophysin and

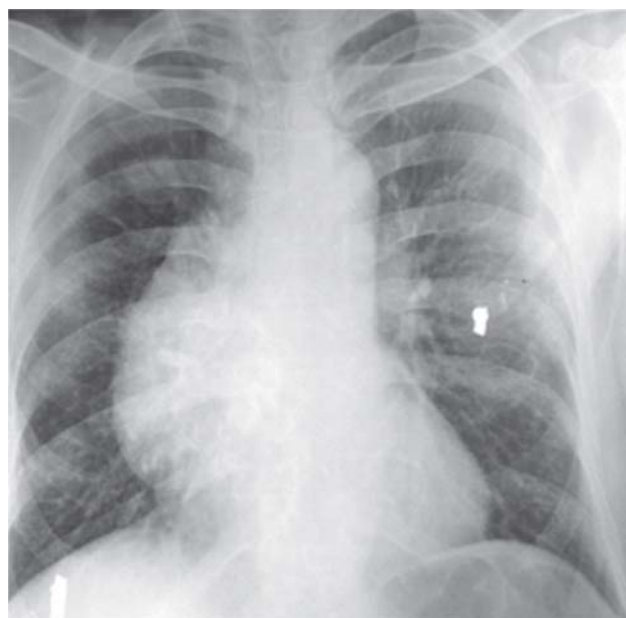


Figure 1. Pre-operative chest radiograph (postero-anterior view) showing a mediastinal mass lesion.

chromogranin and patchy positivity for CD56. The FNAC from the left supra-clavicular node was also suggestive of neuro-endocrine cell neoplasm possibly carcinoid.

Haematological investigations were normal. Pulmonary function testing revealed moderate restriction. Since the patient did not have any clinical feature of endocrine syndromes, detailed hormonal evaluation was not carried out.

A right postero-lateral thoracotomy exposed a well encapsulated mass 13cm x 8cm x 7cm adherent to the right hilum and pericardium without invasion. The tumour was seen to extend to the left, crossing the midline.

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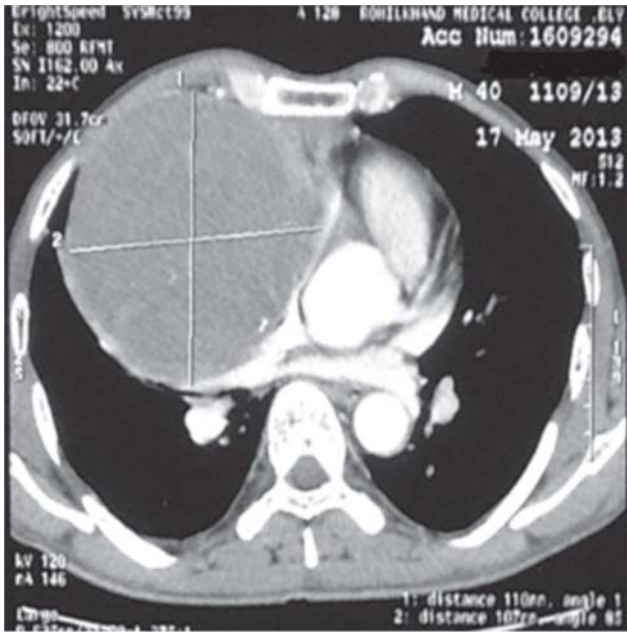


Figure 2. CECT chest showing a well-defined anterior mediastinal mass partly compressing the right main bronchus and superior vena cava.

Great vessels were free of infiltration and no enlarged lymph nodes were identified. The right pleura and lung were normal. With meticulous dissection, the tumour could be excised en bloc (Figure 3). A cut-section of the specimen had variegated appearance with fleshy, haemorrhagic and calcified areas. Histological picture was consistent with a well differentiated neuro-endocrine carcinoma of the thymus with absence of mitosis.

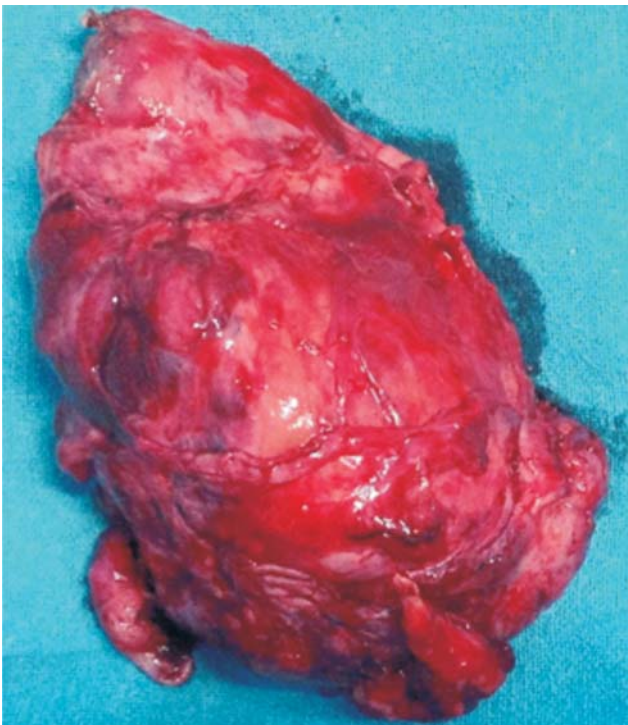


Figure 3. Photograph of the excised surgical specimen showing a lobulated and encapsulated appearance.

The patient had an uneventful post-operative course (Figure 4) and was referred to the oncologist for consideration of adjuvant therapy.

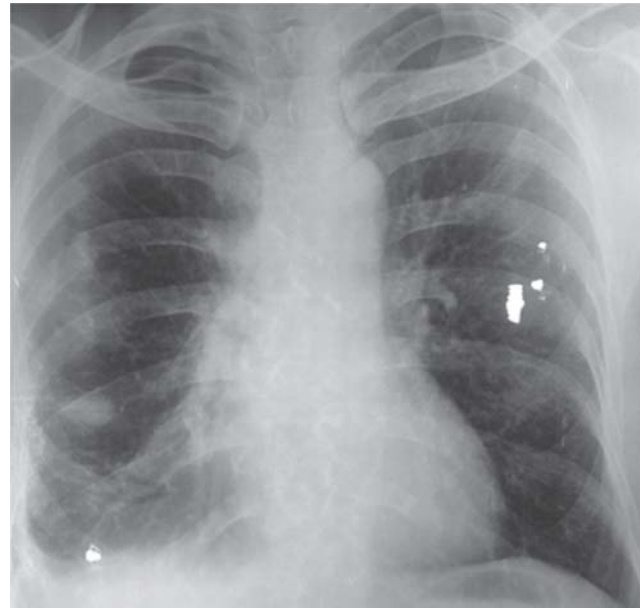


Figure 4. Post-operative chest radiograph of the patient.

Discussion

Primary thymic neuro-endocrine carcinomas constitute <5% of all mediastinal tumours.³ These can be incidentally detected or present to the clinician with Cushing's syndrome or as part of multiple endocrine neoplasia (MEN) syndromes. Clinically, these may be locally confined or metastatic. On histopathology these tumours can vary from being well-differentiated to poorly-differentiated tumours. According to the current classification,⁴ well-differentiated thymic neuro-endocrine carcinomas and thymic carcinoid tumours are synonymous.⁴ This condition has a uniformly poor prognosis irrespective of the histological character of the lesion.⁵ Thymic neuro-endocrine carcinomas are known for a poor 5-year survival and high chance of local recurrence as well as distant metastasis which cannot be predicted from the initial presentation.⁶ A separate staging for neuro-endocrine carcinomas has not been proposed for thymus.⁷ But considering the staging of other thymic carcinomas, supra-clavicular lymph node involvement is grouped under stage 4 along with distant metastasis. Hence, management of present case also goes along the line of metastatic thymic carcinoid. An aggressive surgical approach is recommended for thymic neuro-endocrine carcinomas whether the tumour is localised or metastatic. Even resectable distant metastasis should be surgically dealt with. As the role of adjuvant radiotherapy and chemotherapy have not been well established, the reduction of tumour burden by an aggressive surgical approach may help to prolong the time period for future development of local recurrence and distant metastasis. Resection of the primary tumour

is not indicated only in those small stable ones with unresectable distant metastasis. Surgery is the primary mode of therapy except in those with huge unresectable tumours and with carcinoid syndrome where octreotide is suggested as the fine-line treatment.⁷ Thus, the management of advanced stage thymic neuro-endocrine carcinomas differ from other advanced thymic carcinomas in that the latter is preferably managed with chemotherapy before surgical resection.⁸ In our patient, in addition to the large tumour size, radiographic evidence of partial compression of the right main bronchus and superior vena cava prompted surgical excision, although it was a metastatic tumour.

Although thymic carcinoids are rare tumours with aggressive behaviour and poor prognosis, primary surgical excision is indicated even in advanced disease to reduce the tumour burden and reduce the local symptoms.

References

1. Rosai J, Higa E. Mediastinal endocrine neoplasm of probable thymic origin related to carcinoid tumor. *Cancer* 1972;29:1061-74.
2. Moran CA, Suster S. Neuroendocrine carcinomas (carcinoid tumor) of the thymus: a clinicopathologic analysis of 80 cases. *Am J Clin Pathol* 2000;114:100-10.
3. Duh QY, Harbarger CP, Geist R, Gamsu G, Goodman PC, Gooding GAW, *et al.* Carcinoids associated with multiple endocrine neoplasia syndromes. *Am J Surg* 1987;154:142-8.
4. Klimstra DS, Modlin IR, Coppola D, Lloyd RV, Suster S. The pathologic classification of neuroendocrine tumours. *Pancreas* 2010;39:707-12.
5. Ahn S, Lee JJ, Ha SY, Sung CO, Kim J, Han J. Clinicopathological analysis of 21 thymic neuroendocrine tumors. *Korean J Pathol* 2012;46:221-5.
6. Fukai I, Masaoka A, Fujii Y, Yamakawa Y, Yokoyama T, Murase T, *et al.* Thymic neuroendocrine tumor (thymic carcinoid): a clinicopathologic study in 15 patients. *Ann Thorac Surg* 1999;67:208-11.
7. National Comprehensive Cancer Network (NCCN). Clinical practice guidelines in oncology. Version 2. 2013. Neuroendocrine tumours. Available at URL: <http://www.nccn.org/>. Accessed on September 4, 2014.
8. National Comprehensive Cancer Network (NCCN). Clinical practice guidelines in oncology, Version 2. 2013. Thymomas and thymic carcinomas. Available at URL: <http://www.nccn.org/>. Accessed on September 4, 2014.