Mediastinal carcinoid tumour with unusual manifestations

Praveen Aggarwal¹, Surender Kumar Sharma¹, Tushar Kanti Chattopadhyay² and Ashok Kumar Mukhopadhyay³

Departments of ¹Medicine, ²Surgery and ³Pathology, All India Institute of Medical Sciences, New Delhi 110029, India.

Summary: A rare case of mediastinal carcinoid tumour with superior vena cava obstruction, and osteolytic as well as osteoblastic secondaries in the bones is described. The relevant literature is reviewed.

Introduction

Carcinoid tumours occur most commonly in the gastrointestinal tract. Other reported sites include the lung, gall bladder and bile duct, parotid, pancreas, ovary, cervix and testis. In recent years, similar tumours occurring in the anterior mediastinum have been described.¹ In this report, we describe certain unusual features in a patient with mediastinal carcinoid.

Case report

A 36 year old male presented with pain in the right arm radiating along its medial aspect, of 3 years duration, worse on coughing and sneezing. One year before admission he noticed progressive thinning of the right leg. At about the same time, he also developed sudden nasal regurgitation, slurring of voice and deviation of tongue to the left side and these symptoms persisted at the time of admission. Six months prior to admission, he noticed facial puffiness along with mild dyspnoea on exertion. He had lost 12 kg in weight in the past year.

Examination revealed signs of superior vena cava obstruction. Chest examination revealed dullness in the right upper parasternal area. The nervous system examination showed evidence of lower motor neurone palsies of IX, X and XII cranial nerves on the left side along with slight wasting of the right calf muscles and decreased sensation along D₁ distribution in the right arm. The ankle jerk on the right side was absent.

The chest X-ray showed a superior mediastinal mass (Figure 1). The skeletal survey showed multiple lytic lesions in the D₄, D₅ and L₄ vertebrae; multiple areas of sclerosis in the pelvis and upper femora and multiple lytic lesions in the skull (Figure 2). There was no

M-spike in the blood or Bence-Jones protein in the urine. An ultrasonography of the abdomen and computed tomography of the skull were normal. Repeated attempts to obtain tissue by needle aspiration of the mediastinal mass were unsuccessful.

At thoracotomy, a large mass occupying the superior mediastinum with infiltration of the great vessels was found. Histopathology of a biopsy revealed groups of small, rounded cells, separated by dilated vascular endothelial channels. The cells had scant cytoplasm and darkly-stained nuclei, occupying almost the whole of the cells. The cytoplasm contained

Figure 1 Chest X-ray showing a superior mediastinal mass.

Figure 2 Skull X-ray showing multiple lytic lesions.

fine granules. Numerous mitotic figures were seen. The appearances were compatible with carcinoid tumour.

In a search for carcinoid tumour elsewhere, radiographic evaluation of the gastrointestinal tract and a fiberoptic bronchoscopy were performed. There was no evidence of tumour at both these sites. Twenty-four hour urinary 5-hydroxyindoleacetic acid [2.2 mg (normal 2–8 mg)] and 17-hydroxysteroid [6 mg (normal 2–10 mg)], and plasma cortisol levels were not elevated.

A final diagnosis of malignant carcinoid tumour of the mediastinum with superior vena cava obstruction and multiple osteolytic and osteoblastic secondaries was made. The neurological involvement was thought to be due to metastatic deposits causing pressure on the nerve roots. The patient received local radiotherapy to the mediastinal mass followed by chemotherapy with 5-fluorouracil without any response.

Discussion

The mediastinal carcinoids have been regarded as tumours arising from thymus because of the presence

of Kulchitsky's cells (the origin of these tumours) in thymus and also because of the demonstration of remnants of thymic tissue in many of these tumours. Fewer than 100 cases of thymic carcinoids have been reported.² The male/female incidence is 3:1 and the tumours present most commonly in the fourth and fifth decades of life.

The symptoms are nonspecific and consist of dyspnoea, cough and chest pain.¹ Out of the 8 cases reported by Rosai and Higa,¹ 5 were asymptomatic. Wick *et al.*³ reported 7 cases, of which two had superior vena cava obstruction. Out of 3 cases reported by Salyer *et al.*⁴ two patients developed superior obstruction during follow up. Carcinoid tumours of the thymus have also been reported to be associated with multiple endocrine adenomatosis⁵ and with Cushing's syndrome.^{3,4} Systemic features in the form of polyarthritis and paraesthesiae, weight loss, clubbing, hepatomegaly and proximal myopathy have also been reported.⁶ Carcinoid syndrome has not yet been described in any patient with thymic carcinoid tumours.

In patients with malignant carcinoid tumours, the metastatic bone deposits are mostly osteoblastic while osteolytic lesions are rare.⁸ Osteolytic as well as osteoblastic lesions in the same patient have been reported rarely.⁹ To our knowledge, osteolytic metastatic bone lesions have not been previously reported in thymic carcinoids.

The primary mode of therapy in thymic carcinoids is surgical removal. In a review of 16 cases,¹ the visible tumour could be removed in 12 patients out of whom 9 were alive for 1–9 years postoperatively. The response to radiotherapy is poor.¹

The present case is of interest not only because of the location of the tumour but also because of the presence of the superior mediastinal obstruction and the osteolytic as well as the osteoblastic secondaries in the bones. The possibility of thymic carcinoid, although quite rare, should be considered in the differential diagnosis of tumours in this location.

References

1. Rosai, J. & Higa, E. Mediastinal endocrine neoplasm, of probable thymic origin, related to carcinoid tumour. Clinicopathologic study of 8 cases. *Cancer* 1972, **29**: 1061–1074.
2. Kogan, J. Carcinoid tumour of the thymus. *Postgrad Med* 1984, **75**: 291–296.
3. Wick, M.R., Scott, R.E., Li, C-Y. & Carney, J.A. Carcinoid tumour of thymus. A clinicopathologic report of seven cases with a review of the literature. *Mayo Clin Proc* 1980, **55**: 246–254.
4. Salyer, W.R., Salyer, D.C. & Eggleston, J.C. Carcinoid tumours of the thymus. *Cancer* 1976, **37**: 958–973.
5. Rosai, J., Higa, E. & Davie, J. Mediastinal endocrine neoplasm in patients with multiple endocrine adenomatosis. A previously unrecognized association. *Cancer* 1972, **29**: 1075–1083.
6. Lowenthal, R.M., Gumpel, J.M., Kreel, L., McLaughlin, J.E. & Skeggs, D.B.L. Carcinoid tumour of the thymus with systemic manifestations: a radiological and pathological study. *Thorax* 1974, **29**: 553–558.
7. Thomas, B.M. Three unusual carcinoid tumours, with particular reference to osteoblastic bone metastases. *Clin Radiol* 1968, **19**: 221–225.
8. Bichile, L.S., Pandit, S.P., Kuloor, P.L., Abhyankar, S.C., Hedge, A. & Nanivadekar, S.A. Carcinoid tumour of lung with haemorrhagic pleural effusion and extensive metastasis. *Indian J Chest Dis Allied Sci* 1984, **26**: 51–53.
9. Norman, A., Greenspan, A. & Steiner, G. Case report 170: Varified osteolytic metastasis in the clavicle from a bronchial carcinoid tumour and probable osteoblastic metastasis in the humerus from the same lesion. *Skeletal Radiol* 1981, **7**: 155–157.