Anterior mediastinal mass in a patient with Cushing's syndrome

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A 35-year-old male patient presented with complaints of weight loss, facial puffiness, and pedal edema. On physical examination, he was found to be hypertensive (blood pressure - 180/110 mm Hg) and showed centripetal obesity, buffalo hump, and purple striae over body. On biochemical evaluation, patient was found to be diabetic. Fasting blood sugar was 155 mg/dl. He had elevated levels of serum cortisol with loss of circadian rhythm and elevated levels of adrenocorticotropic hormone (ACTH). 8 am cortisol levels were 51 µg/dl and plasma ACTH was 207 pg/ml. The mean plasma cortisol after dexamethasone suppression test (DST) showed non-suppressibility. Overnight DST was >1750 and low dose DST was 1209. Magnetic Resonance Imaging showed no pituitary adenoma. Chest radiograph [Figure 1] showed a well-defined lobulated mediastinal mass silhouetting right heart border. Contrast-enhanced computed tomographic (CT) [Figure 2] in mediastinal window showed a well-defined lobulated heterogeneously enhancing mass lesion in anterior mediastinum. No lung parenchymal lesion was evident.

Question

What is the Diagnosis?

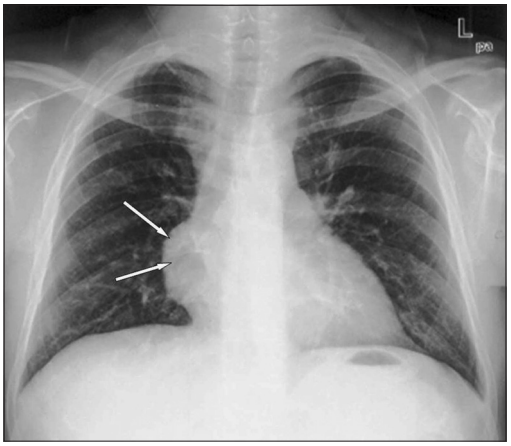

Figure 1: Chest radiograph showing a well-defined lobulated mediastinal mass

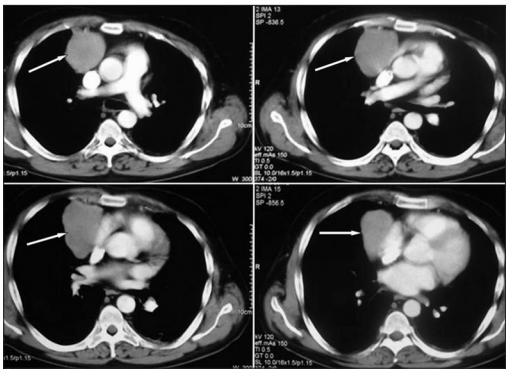

Figure 2: Contrast-enhanced CT showing a well-defined lobulated heterogeneously enhancing mass lesion in anterior mediastinum

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Answer

Thymic carcinoid

CT-guided fine needle aspiration cytology was performed which suggested the diagnosis of thymic carcinoid. With the diagnosis of a thymic carcinoid leading to Cushing's syndrome due to ectopic adrenocorticotropic hormone (ACTH) production, the patient underwent surgery. Mediastinal mass lesion was seen arising from right lobe of thymus and patient underwent extended thymectomy. Histopathological examination confirmed the diagnosis of neuroendocrine tumor arising from thymus.

Discussion

Differential diagnosis of common anterior mediastinal masses includes four T's, i.e., thymic masses, thyroid (goiter with intrathoracic extension), teratoma and other germ cell tumors, and terrible lymphoma. Thymus origin masses include thymoma, thymic cyst, thymic hyperplasia, thymolipoma, thymic carcinoma, and thymic carcinoid. Thymic carcinoid is a rare cause of Cushing's syndrome.

Cushing's syndrome is a disorder caused by prolonged exposure of tissues to high levels of glucocorticoids. Source of glucocorticoids can either be endogenous or exogenous. Patient presents with moon faces, facial puffiness, buffalo hump, centripetal obesity, skin pigmentation, proximal muscle weakness, hirsutism, hypertension, and diabetes mellitus. Endogenous Cushing's syndrome is caused by ACTH secreting pituitary adenomas in 75 to 80% and the remaining 20 to 25% is due to adrenal neoplasms (10-15%) and ectopic ACTH secretion (10%). Ectopic sources of ACTH include small cell carcinoma of the lung and various neuroendocrine tumors such as bronchial and thymic carcinoid, and rarely gastro-intestinal tract carcinoids.[1]

Contrast-enhanced CT chest and abdomen is considered best examination when looking for an occult ACTH-secreting neoplasm. However, in case of failure to localize the ectopic source of ACTH secretion with one modality, multimodality imaging techniques should be considered to identify the source. CT, Magnetic resonance imaging (MRI), and octreotide scan/positive emission tomography (PET) scan all should be employed.[1] Thymic carcinoids are rare. They are seen in all age groups, with a 3:1 male predominance.[2] Symptoms are generally due to mass effect on adjacent structures or invasion of the mediastinum. Most of the thymic carcinoids are of intermediate grade histologically and half of them will have metastatic deposits at the time of diagnosis. 15% of thymic carcinoids are associated with MEN 1 syndrome and approximately 25% of the patients present with Cushing syndrome. Carcinoid syndrome is very rare but has been reported in two cases.[2] Thymic carcinoids are diagnosed as anterior mediastinal mass on chest radiograph or CT. Although chest X-ray is the initial imaging modality, it is not adequate either for screening or follow-up because the lesion may be obscured by adjacent mediastinal structures.[2] CT scan on the other hand will clinch the diagnosis in appropriate clinical setting. The role of somatostatin receptor scintigraphy in detection of small thymic lesions has not been completely established.[2] MRI has the potential role to detect pericardial and large-vessel invasion before surgery.[2] A PET scan with 18-fluorodeoxyglucose may be useful in detecting distant metastases and recurrence, but concern is being raised about the physiological uptake of 18-fluorodeoxyglucose in the thymus.[2] Thymic carcinoids are aggressive lesions and often have invaded adjacent structures at the time of presentation.[3]

Radical surgical excision of the thymus is considered treatment of choice in case of a thymic carcinoid. Even If the tumor is non-resectable, debulking is considered best.[4] Chemotherapy and radiotherapy may be considered either preoperatively or postoperatively, although because of small number of patients, standard guidelines about optimal treatment modalities have not been completely established.[3] Survival in an individual patient will depend upon stage at presentation, histologic degree of differentiation, associated endocrine features, and resectability rate. Recurrences can be locoregional or distant and surgery should be considered even in the treatment of locoregional recurrences.[3] However, distant metastasis is still an issue due to lack of better systemic form of therapy.[5]

In conclusion, thymic carcinoids are rare; however, when a diagnosis of ectopic Cushing's syndrome is suspected, these lesions merit consideration and a chest radiograph followed by CT should be performed to look for this important source of ectopic ACTH secretion leading to Cushing's syndrome.

References

1. Bhansali A, Walia R, Rana SS, Dutta P, Radotra BD, Khandelwal N, *et al*. Ectopic Cushing's syndrome: Experience from a tertiary care centre. Indian J Med Res 2009;129:33-41.
2. Elsayes KM, Menias CO, Bowerson M, Osman OM, Alkharouby AM, Hillen TJ. Imaging of carcinoid tumors: Spectrum of findings with pathologic and clinical correlation. J Comput Assist Tomogr 2011;35:72-80.
3. Ruffini E, Oliaro A, Novero D, Campisi P, Filosso PL. Neuroendocrine tumors of the thymus. Thorac Surg Clin 2011;21:13-23.
4. Van Brandt V, Heyman S, Van Marck E, Hendriks J, Van Schil P. Atypical presentation of an atypical carcinoid. Ann Thorac Surg 2009;88:2004-6.
5. Dutta R, Kumar A, Julka PK, Mathur SR, Kaushal S, Kumar R, *et al*. Thymic neuroendocrine tumour (carcinoid): Clinicopathological features of four patients with different presentation. Interact Cardiovasc Thorac Surg 2010;11:732-6.

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