Papillary Carcinoma of Thyroid Presenting as Posterior Mediastinal Mass with Superior Vena Cava Syndrome

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Abstract
Enlarged thyroid with retrosternal extension presenting as anterior mediastinal mass is known. Superior vena cava (SVC) syndrome due to direct invasion from a primary thyroid malignancy is a rare phenomenon. We present a unique case of papillary carcinoma of thyroid extending into the posterior mediastinum with superior vena cava syndrome along with internal jugular and azygous vein thrombosis.

INTRODUCTION
In the past, SVC obstruction occurred due to syphilitic aortic aneurysm or mediastinal fibrosis secondary to tuberculosis. Presently malignant tumours account for 86% of all cases of SVC syndrome. Bronchogenic carcinoma [70%], lymphomas [8%] and other metastatic involvements [8%] are common malignancies which invade mediastinum. We report a rare presentation of papillary carcinoma of thyroid extending into the posterior mediastinum while also causing SVC obstruction syndrome.

CASE REPORT
A 50 year old male, non-smoker was admitted with complaints of midline neck swelling for 10 years, mild puffiness of face, breathlessness on lying down and dry cough for last 5 months. He also had dysphagia to solids as well as liquids and chest pain localized to right side which was continuous and dull aching. He did not give history of fever, palpitations, syncope, weight loss, abdominal distension, edema feet, bone pain or stridor.

Physical examination revealed a pulse of 88/min, blood pressure of 100/70mmHg and respiratory rate of 28/min. He had no pallor, clubbing, cyanosis or pedal edema. Face was plethoric and puffy. Right jugular vein was distended but nonpulsatile. Distended veins were seen over right side of chest demonstrating downward flow. Medial group of right maxillary lymph nodes were palpable. Thyroid gland was palpable [12cmx10cm]; firm, nontender, without bruit, with a hard nodule at inferior margin of right lobe. The auscultation of chest and systemic examination were normal. A diagnosis of SVC syndrome possibly due to thyroid malignancy was made.

Investigations revealed hemoglobin=12gm/dl, WBC = 3600/cmm, ESR 98mm at 1 hr, Thyroid function tests were normal with T3=123.9 ng/dl [60-181], T4=3.2 (µg/dl [3.2-12.6], TSH=1.3[IU/ML [0.35-5.5], prothrombin time was 13/11 with an INR value of 1.2. The tests for HIV and HBsAg were negative; all biochemical parameters were normal.

X-ray chest showed superior mediastinal widening which is extending upwards to neck, ultrasonography of neck revealed large right lobe of thyroid with necrotic enlargement of right carotid lymph nodes. CT scan of chest and neck confirmed primary neoplasm arising from the right lobe of thyroid gland with cervical and axillary lymphadenopathy, rib destruction and thrombotic obstruction of the azygous and right internal jugular vein. The trachea was severely compressed and reduced to a slit at the T4 vertebral level. Thyroid mass shows extension into posterior mediastinum [Fig. 1]. Fine needle aspiration cytology of thyroid showed tumor cells arranged in sheets forming papillae with large nuclei and dusty chromatin with internuclear cytoplasmic inclusions suggestive of papillary carcinoma [Fig. 2].

During the stay in ward, patient had several episodes of worsening breathlessness and hemoptysis. He was treated with intravenous steroids, antibiotics and radiotherapy. He died of severe respiratory distress after indoor stay of one month. A postmortem specimen from thyroid gland showed marked pleomorphic population of tumour cells comprising of round and spindle shaped cells with abundant cytoplasm against necrotic background suggestive of an anaplastic change in the papillary carcinoma.
SVC syndrome is a critical condition in which an intrathoracic mass lesion compresses the SVC and promotes the development of head and upper extremity edema and cyanosis. Either a luminal obstruction or an extrinsic compression can impair flow in the thin walled low pressure SVC. Luminal obstruction can be due to neoplastic infiltration or by thrombosis occurring in the setting of a thrombophilic state associated with some tumors. Tumor and lymph node masses, aortic aneurysms, bronchogenic and esophageal carcinomas cause extrinsic compression. The Azygous vein is the first available bypass others being inferior mammary vein, long thoracic venous system and vertebral veins. Compression of azygous vein [as in our patient] will increase superficial venous distension.3 Diagnostic modality of choice is contrast CT scan of chest and neck. Tissue diagnosis should be obtained whenever possible.4,5 Conservative measures like bedrest, supplemental oxygen, corticosteroids and diuretics may provide symptomatic relief along with local thrombolytic therapy for clot formers. In the setting of malignancy local radiotherapy or stenting to relieve obstruction can provide palliation. Complications include thromboembolism, excessively high venous pressure with cerebral and life threatening laryngeal edema.4,5

Our patient had papillary carcinoma of thyroid gland, which is usually found in the 3rd to 4th decade with a female preponderance. The tumors tend to be biologically indolent with excellent prognosis. Poor prognostic factors are old age at diagnosis, male sex, large tumour size, extra thyroidal growth and adverse histological subtypes (solid arrangement pattern with differentiation).6 The anaplastic change can explain the aggressive behavior of papillary carcinoma with rapid worsening and death within a period of one-month inspite of radiotherapy.

Thyroid malignancy invading posterior mediastinum is very rare and has not been reported in literature, Coexistence of SVC syndrome as a complication of papillary carcinoma of thyroid makes the case unique.

REFERENCES