Case Report

Pancoast syndrome: A rare presentation of non-Hodgkin's lymphoma

Anirban Sarkar, Anirban Das[1], Sumitra Basuthakur[1], Sudipta Pandit[1], Sibes K. Das[1], Sabyasachi Choudhury[1]

Department of Pulmonary Medicine, Midnapore Medical College, Paschim Midnapore, [1]Department of Pulmonary Medicine, Medical College, Kolkata, West Bengal, India

ABSTRACT

Pancoast syndrome is a common presentation of bronchogenic carcinoma, but other malignancies are rarely cited as its cause. Pancoast syndrome due to non-Hodgkin's lymphoma is rarely described in the literature. Here, we report a case of Pancoast syndrome due to non-Hodgkin's lymphoma to increase the awareness of the clinicians regarding essentiality of tissue diagnosis of Pancoast tumor before starting the treatment.

KEY WORDS: Computed tomography, non-Hodgkin's lymphoma, Pancoast syndrome, tru-cut biopsy

Address for correspondence: Dr. Anirban Das, C/O Mr. Pradip Das, Peon Para, Bhatchala, Near Vidyarthi Girls' High School, P.O.: Sripalili, P.S.: Burdwan Sadar, Burdwan, West Bengal, India. E-mail: dranirbandas_chest@rediffmail.com

INTRODUCTION

Pancoast syndrome is characterized by the shoulder and arm pain radiating along the distribution of eighth cervical, first and second thoracic nerve trunks, weakness and atrophy of the intrinsic hand musculature, and ipsilateral Horner's syndrome. It is most commonly caused by local extension of an apical lung tumor at the superior thoracic inlet, most common histopathological type being squamous cell carcinoma.[1] These tumors are called Pancoast tumor or Superior sulcus tumor. In 1932, Henry Pancoast first described the clinic-radiological features of the thoracic inlet tumors, but he mistakenly thought that it originates from embryonal epithelial rests of fifth brachial cleft.[2] Among malignant etiologies other than lung cancer, lymphoma, especially non-Hodgkin lymphoma (NHL) is reported occasionally to cause Pancoast syndrome. Here, we report such a rare case of anterior mediastinal NHL, which extended to the neck to produce the Pancoast syndrome due to involvement of the lower trunk of brachial plexus and inferior cervical ganglion.

Access this article online

Quick Response Code:

Website: www.lungindia.com

DOI: 10.4103/0970-2113.116266

CASE REPORT

A 16-year-old female presented with a painful swelling on the left side of the neck for 2 months, and gradually increasing weakness and numbness of left upper limb with drooping of the left an upper eye lid for 1 month. She had sharp, shooting pain in the neck and left shoulder, radiating along the inner aspect of the left arm, associated with tingling and numbness of her left forearm extending to the tips of her fourth and fifth fingers, ulnar aspect of the forearm and hand. The pain was not relieved by simple analgesic and her sleep was disturbed because of intense pain. Neck swelling was irregular in shape and progressively increasing in size; initial size was 2 cm × 3 cm. There was no history of fever, weight loss, and anorexia. She was normotensive and non-diabetic. There was no past history of the tuberculosis.

On general examination, there was a firm, tender, irregular swelling of 4 cm × 4 cm size in the left side of the neck extending to left supraclavicular area. The swelling was fixed to underlying structures and lower margin was not delineated. Temperature was not raised and overlying skin was normal. Fluctuation was absent and transillumination test was negative. There was no other peripheral swelling in the body. Clubbing and anemia were absent and her axillary temperature was 97°F, pulse rate 100 beats/min, respiratory rate 20 breaths/min, and the blood pressure was 110/70 mmHg.

On examination of the respiratory system, the movement of the chest wall was restricted over left supraclavicular

and infracalvicular areas along with fullness in the same areas. There was no venous engorgement over the chest wall. Trachea was shifted to the right without shifting of cardiac apex. Percussion note was dull over left second and third intercostal spaces and sternal percussion was also dull. Diminished vesicular breath sound and decreased vocal resonance were noted over left supraclavicular and infraclavicular areas.

Eye examination revealed the partial ptosis of left upper eye lid with ipsilateral miosis, but the vision was preserved with normal light and accommodation reflexes. Ocular movement was normal in all directions.

On neurological examination of upper extremity, the hypothenar muscles of the left hand were wasted. There was a decrease in power of all muscle groups in this hand (grade 2/5). Fine touch and pain sensation were decreased in the C8 and T1 segmental distribution in his left hand and arm. Triceps jerk was diminished.

Complete hemogram and blood biochemistry were normal, except elevated serum lactate dehydrogenase level (340 U/L). Chest X-ray (P.A. view) showed a huge anterior mediastinal mass, compressing left lung and extending to the thoracic inlet, but no rib erosion. Sputum smear for acid fast bacilli was negative. Incisional biopsy from the neck swelling was inconclusive. Contrast enhanced computed tomography (CT) thorax showed heterogeneous huge anterior mediastinal necrotizing mass extending through thoracic inlet to the neck, but no rib erosion [Figure 1]. CT guided fine needle aspiration cytology (FNAC) was inconclusive. Excision biopsy from the left supraclavicular swelling revealed malignant round cell tumor involving lymph nodes. Histopathological examination of CT guided tru-cut biopsy of mediastinal mass showed effacement of normal architecture of lymph gland, which was replaced by sheets of atypical large lymphoid cells admixed with the histiocytes and plasmacytoid cells. Scattered immunoblasts were present. No Reed Sternberg cell or its variant was found [Figure 2]. Hence, histopathology ultimately confirmed the tissue diagnosis of NHL. Immunohistochemistry further substantiated the diagnosis of diffuse large B-cell phenotype of NHL, as the tumor cells expressed CD20 and CD10, but were immunonegative for CD3, CD5, CD23, and CD30 [Figure 3]. Bone marrow biopsy was within normal limit. Ultrasound of whole abdomen was normal. Hence, he was finally diagnosed as a case of diffuse large B-cell phenotype of NHL. The size of the lesion regressed following six cycles of combination chemotherapy, comprising of cyclophosphamide, doxorubicin, vincristine, and prednisolone.

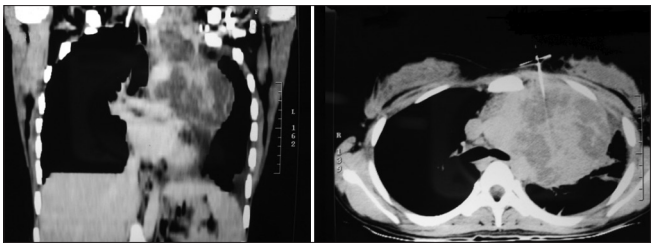

Figure 1: Contrast enhanced computed tomography thorax showing a huge, heterogeneous anterior mediastinal mass extending to neck through the thoracic inlet

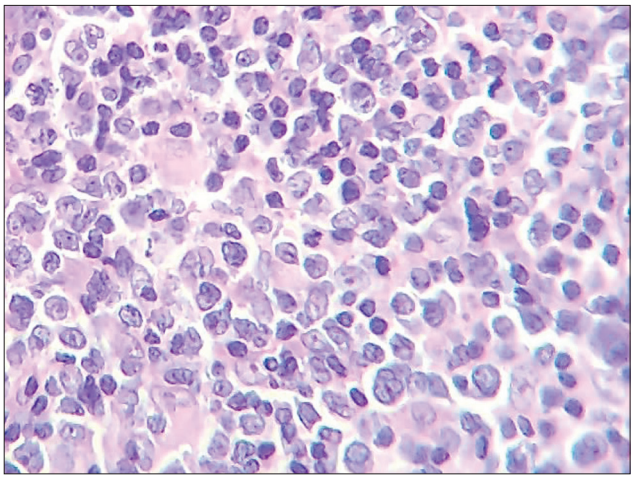

Figure 2: Photomicrograph of computed tomography-guided tru-cut biopsy specimen taken from anterior mediastinal mass showing effacement of normal architecture of lymph node with sheets of atypical large lymphoid cells (H and E, ×40)

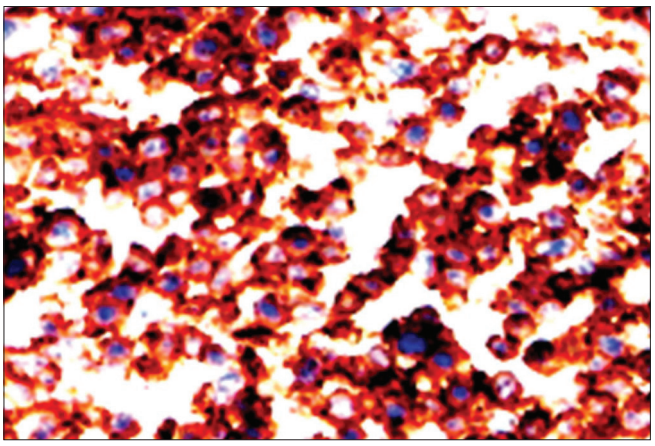

Figure 3: Immunohistochemistry showing CD20 positive tumor cells

DISCUSSION

Pancoast syndrome is rare and primary bronchogenic carcinoma, located at pulmonary apex is the most common etiology, which is a rare location accounting for less than 5% of all bronchogenic carcinomas.[3] These tumors may invade the second and third ribs, the brachial plexus, the subclavian vessels, the stellate ganglion, and adjacent vertebral bodies, and therefore prognosis is definitely poor. Multiple myeloma, metastatic uterine cervical carcinoma, metastatic hepatocellular carcinoma, and lymphomatoid granulomatosis are other malignant causes of may cause Pancoast's syndrome.[3-7] Lymphoma, especially NHL is rarely reported as being a cause of Pancoast syndrome.[8] Benign causes of Pancoast syndrome include pulmonary

tuberculosis, hydatid cyst, histoplasmosis, aspergillosis, destructive sclerosing fibrosis following staphylococcal infection at lung apex.[9-12] Pancoast syndrome is due to involvement of the lower trunk of brachial plexus and inferior cervical ganglion by locally progressive pulmonary apical tumors, which often cause erosion of upper ribs and chest wall destruction before neural involvement.

Brachial neuritis has also been described as a paraneoplastic syndrome of lymphoma, especially of Hodgkin's variety.[13] However, our case is an extremely rare case of diffuse large B-cell NHL, presenting as an anterior mediastinal mass and causing Pancoast syndrome.

Transthoracic lung biopsy may be performed using the fine needle aspiration (FNA) technique or using cutting needle for core biopsy. FNA provides material for cytological analysis and is sometimes sufficient for diagnosis of primary or metastatic lung cancer, but core biopsy is often required to provide the material for the histopathological examination and is often useful for a definite diagnosis of lung cancers, benign non-infectious lung lesions, lymphomas and to distinguish mesothelioma from adenocarcinoma. It also provides sufficient sample for microbiological culture and immunohistochemistry. Most diagnosis of malignancy requires more tissue than FNAC can provide and it often delays a definitive diagnosis.

This case report not only illustrates a rare cause of Pancoast syndrome, but also reemphasizes the importance of the tissue diagnosis in all cases of Pancoast tumor as it significantly alters the treatment options and prognosis.

REFERENCES

1. Flenley DC. Respiratory Medicine. 2nd ed. London: Bailliere Tindall; 1990.
2. Pancoast HK. Superior pulmonary sulcus tumor: Tumor characterized by pain, Horner's syndrome, destruction of bone and atrophy of hands muscles. J Am Med Assoc 1932;99:1931-2.
3. Ahmad K, Fayos JV, Kirsh MM. Apical lung carcinoma. Cancer 1984;54:913-7.
4. Wilson KS, Cunningham TA, Alexander S. Myeloma presenting with Pancoast's syndrome. Br Med J 1979;1:20.
5. Omenn GS. Pancoast syndrome due to metastatic carcinoma from the uterine cervix. Chest 1971;60:268-70.
6. Chang CF, Su WJ, Chou TY, Perng RP. Hepatocellular carcinoma with Pancoast's syndrome as an initial symptom: A case report. Jpn J Clin Oncol 2001;31:119-21.
7. Dolan G, Smith J, Reilly JT. Extrapulmonary lymphomatoid granulomatosis presenting as Pancoast's syndrome. Postgrad Med J 1991;67:914-5.
8. Ngamphaiboon N, Pukiat S, Kandimalla Y, Hernandez-Ilizaliturri FJ, Rooney JJ. Diffuse large B-cell lymphoma presenting as pancoast tumor. J Clin Oncol 2010;28:e254-6.
9. Engleman P, Liebow AA, Gmelich J, Friedman PJ. Pulmonary hyalinizing granuloma. Am Rev Respir Dis 1977;115:997-1008.
10. Stathatos C, Kontaxis AN, Zafiracopoulos P. Pancoast's syndrome due to hydatid cysts of the thoracic outlet. J Thorac Cardiovasc Surg 1969;58:764-8.
11. Simpson FG, Morgan M, Cooke NJ. Pancoast's syndrome associated with invasive aspergillosis. Thorax 1986;41:156-7.
12. Gallagher KJ, Jeffrey RR, Kerr KM, Steven MM. Pancoast syndrome: An unusual complication of pulmonary infection by *Staphylococcus aureus*. Ann Thorac Surg 1992;53:903-4.
13. Pezzimenti JF, Bruckner HW, DeConti RC. Paralytic brachial neuritis in Hodgkin's disease. Cancer 1973;31:626-9.

How to cite this article: Sarkar A, Das A, Basuthakur S, Pandit S, Das SK, Choudhury S. Pancoast syndrome: A rare presentation of non-Hodgkin's lymphoma. Lung India 2013;30:209-11.

Source of Support: Nil, **Conflict of Interest:** None declared.