

Pancoast Syndrome and Endobronchial Tumor Infiltration as the First Manifestation of Hodgkin Lymphoma

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The most common cause of Pancoast syndrome is bronchogenic carcinoma. Other less common causes are solid tumor metastases, other chest tumors, infections, and hematologic neoplasms. Pancoast syndrome due to lymphoma is very rare, and cases described in the literature are essentially associated with non-Hodgkin lymphomas. In a review of the literature we found a single case of Pancoast syndrome secondary to a Hodgkin lymphoma; however, the syndrome manifested during recurrence of disease in that patient. We report a case of nodular sclerosis Hodgkin lymphoma which first manifested clinically as Pancoast syndrome and which was initially diagnosed by bronchial biopsy.

Key words: *Hodgkin lymphoma. Pancoast syndrome. Diagnosis. Bronchial biopsy.*

Síndrome de Pancoast e infiltración tumoral endobronquial como primera manifestación de un linfoma de Hodgkin

La causa más frecuente del síndrome de Pancoast es un carcinoma broncogénico. Otras causas menos frecuentes son metástasis de tumores sólidos, otros tumores intratorácicos, infecciones y neoplasias de estirpe hematológica. El síndrome de Pancoast por un linfoma es muy raro y los casos descritos corresponden fundamentalmente a linfomas no hodgkinianos. En una revisión de la bibliografía encontramos un único caso de linfoma de Hodgkin, pero que se manifestó durante la recidiva de la enfermedad. Nosotros aportamos un caso de linfoma de Hodgkin de tipo esclerosis nodular cuya primera manifestación clínica fue este síndrome y cuyo diagnóstico inicial se realizó mediante biopsia bronquial.

Palabras clave: *Linfoma de Hodgkin. Síndrome de Pancoast. Diagnóstico. Biopsia bronquial.*

Introduction

Pancoast syndrome is not common and is usually secondary to a bronchogenic adenocarcinoma or squamous cell carcinoma. Other causes are solid tumor metastasis, other primary chest tumors,¹ and, in rare cases, infections² and hematologic tumors.³⁻⁵ Pancoast syndrome as the first manifestation of a Hodgkin lymphoma is exceptional. In a bibliographic review we found a single case of Hodgkin lymphoma that presented with Pancoast syndrome, but only during the recurrence of this disease.⁶

We report the case of a woman with Pancoast syndrome secondary to nodular sclerosis Hodgkin lymphoma that was diagnosed through fiberoptic bronchoscopy. We emphasize the rarity of a Hodgkin

lymphoma first manifesting as a large lung mass which led to Pancoast syndrome as well as the extensive endobronchial tumoral infiltration which made diagnosis by endoscopic biopsy possible.

Case Description

The patient was a 51-year-old woman who was a former smoker (20 pack-years) who had quit 6 years earlier and who had a history of atopic dermatitis and bronchial asthma starting in childhood. At the time of presentation she was not undergoing treatment. In the past year she had experienced coughing and breathlessness, mainly associated with exercise, and she attributed those manifestations to her asthma. Three months prior to admission she presented with increased coughing, fever of 38°C, left palpebral ptosis without vision loss, and subsequent shoulder and left shoulder blade pain radiating to the left arm. On physical examination the 38°C fever and left palpebral ptosis with a left miotic but responsive pupil were confirmed. No other neurologic alterations were detected. Two small lymph nodes above the left clavicle and a larger swollen lymph node on the right side of the neck were observed. Heart and lung sounds were

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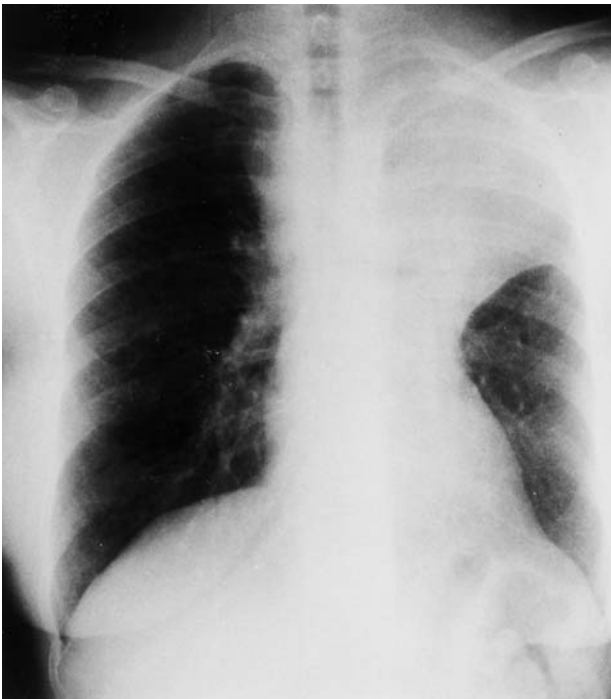


Figure 1. Mass occupying the entire left upper lobe and widening of the mediastinum due to involved lymph nodes.



Figure 2. Computed tomography scan of the chest showing a solid mass in the lung apex.

normal and no abdominal visceromegaly or enlarged lymph nodes in other areas were observed. Hemoglobin was 11.6 g/dL; hematocrit, 36%; white blood cell count, $10.9 \times 10^9/L$ (74% polymorphonuclear cells and 21% lymphocytes); platelet count, $685 \times 10^9/L$, and erythrocyte sedimentation rate, 63 mm/h. Glucose, urea, uric acid, cholesterol, triglyceride, total protein, quantitative immunoglobulin A (IgA), IgM, and IgG determinations were normal. Alkaline phosphatase was 477 U/L (normal range: 91-258 U/L); total bilirubin, 0.28 U/L; lactate dehydrogenase, 546 U/L (normal: 219-439 U/L); aspartate aminotransferase, 28 U/L (normal: 5-40 U/L); alanine aminotransferase, 43 U/L (normal: 5-40 U/L), and γ -glutamyl transpeptidase, 198 U/L (normal: 7-65 U/L). Hepatitis B and C serologies were negative. Urine sediment was normal. Blood gases and lung function were within normal ranges.

The chest x-ray (Figure 1) revealed a large mass that occupied the entire left upper lobe. A computed tomography scan (Figure 2) showed the mass to be solid, without cavitation or calcification, with a diameter of 6x5 cm in the left upper lobe and involved lymph nodes in the aortopulmonary window, right subcarinal and paratracheal region. No alterations were observed in the abdomen or pelvis.

Fiberoptic bronchoscopy showed irregular, edematous, and hyperemic mucosa that caused partial narrowing of the left bronchus and segments of the left upper lobe. Bronchial biopsies confirmed infiltration by a tumor mainly consisting of binucleate lymphocytes with very prominent nucleoli, some of them large and resembling Reed-Sternberg cells. A monoclonal antibody assay showed these lymphocytes to be mostly T cells with only isolated B cells found. Tests for large lymphocytes were positive for CD30 and negative for CD3 and CD20. Subsequently, the involved right neck lymph node was biopsied, and the microscopic and immunohistochemical findings were similar to those of the bronchial biopsy. A bone marrow biopsy revealed no alterations. Combination chemotherapy with adriamycin, bleomycin, vinblastine, and dacarbazine was started.

Discussion

Pancoast syndrome is normally secondary to a primary lung cancer located in the pulmonary apex, which is a rare location accounting for only 2% to 5% of all bronchogenic carcinomas.⁷ Usually the cell type is a well-defined and slow developing squamous carcinoma with a tendency to spread to the subpleural lymph nodes and adjacent structures, such as the roots of the eighth cervical and the second and third dorsal nerves, as well as to the sympathetic chain and bones. Invasion of these structures gives rise to the defining symptoms of Pancoast syndrome:⁸ Horner syndrome, shoulder pain, and weakness and atrophy of upper-limb muscles. Although these symptoms were first described by Pancoast, currently any apical lung disease causing some of the symptoms mentioned above (usually brachial plexus pain) is considered Pancoast syndrome.

Pancoast syndrome secondary to a lymphoma is rare as reflected by the scarcity of reported cases in the literature. The majority of cases are related to high-grade non-Hodgkin lymphomas.³⁻⁵ We found only 1 case of Pancoast syndrome secondary to a Hodgkin lymphoma, and in that patient the syndrome appeared during relapse rather than with presentation, as was the case for our patient.⁶

The origin of the tumor in our case was not clear. It may have originated in the lung parenchyma or in the subpleural lymphatic tissue, later spreading to the mediastinal lymph nodes and neck as occurs in primary extranodal Hodgkin lymphoma of the lung.⁹ Another hypothesis is that the tumor began in the lymph nodes of the neck or the mediastinum with retrograde spread to the subpleural lymph nodes and lymphatic tissue of the bronchial submucosa. The latter situation would better explain the endobronchial infiltration noted on diagnosis, a very unusual finding in a Hodgkin lymphoma,¹⁰ as such

a tumor generally involves the mediastinal lymph nodes.¹¹ In our case the large mass in the lung parenchyma along with the onset of cough and Horner syndrome supports the hypothesis that the tumor probably arose in the left upper lobe and subpleural lymph nodes.

Diagnosing the cause of Pancoast syndrome is difficult due to its location in the extreme periphery of the lung, and, therefore, diagnosis is generally established through transthoracic needle aspiration.¹² In some cases, given the technical difficulties in obtaining a histologic diagnosis, the high probability of bronchogenic carcinoma, and the frequent presence of severe pain may provide sufficient grounds for initial radiotherapy merely based on a clinical and radiological diagnosis. Nevertheless, other diseases are possible, explaining the need for an accurate diagnosis. Our patient's case was one of those in which identifying the etiology resulted in a change of treatment. We therefore believe the exceptionality of the diagnosis by bronchial biopsy merits special attention.

REFERENCES

1. Hatton MQ, Allen MB, Cooke NH. Pancoast syndrome: an unusual presentation of adenoid cystic carcinoma. *Eur Resp J* 1993;6:271-2.
2. Arenas Gordillo M, Ortega Ruiz F, Otero Candeira R, Caballero Oliver A, Orozco Blanco A, Calderón Osuna E. Síndrome de Pancoast producido por tuberculosis pulmonar. *Arch Bronconeumol* 1998;5:266-8.
3. Wang JC, Finn NG, Nimmagadda N, Reddy D. Pancoast syndrome in a patient with malignant lymphoma. *Cancer* 1989;64:2588-90.
4. Villena MV, Echave-Sustaeta J, López Encuentra A, Blasco A. Síndrome de Pancoast secundario a linfoma de Hodgkin. *Arch Bronconeumol* 1995;31:35-6.
5. Arcasoy SM, Bajwa MK, Jett JR. Non Hodgkin's lymphoma presenting as Pancoast's syndrome. *Resp Med* 1997;91:571-3.
6. Goswami C, Dinshaw KA, Shrivastava SK, Muckaden MA, Sharma V, Advani SH. Hodgkin's disease relapse presenting as Pancoast's syndrome. *Indian J Cancer* 1996;33:21-3.
7. Ahmad K, Fayos JV, Kirsh MM. Apical lung carcinoma 1984;54:223-7.
8. Pancoast HK. Superior pulmonary sulcus tumor: tumor characterized by pain, Horner's syndrome, destruction of bone and atrophy of hands muscles. *JAMA* 1932;99:1931-2.
9. Chetty R, Slavin JL, O'Leary JJ, Ansari NA, Gatter KC. Primary Hodgkin's disease of the lung. *Pathology* 1995;27:111-4.
10. Gallagher CJ, Knowles GH, Habeshaw JA, Green M, Malpas JS, Lister TA. Early involvement of the bronchi in patients with malignant lymphoma. *Br J Cancer* 1983;48:777-81.
11. Trédaniel J, Peillon I, Ferme C, Brice P, Gisselbrecht C, Hirsch A. Endobronchial presentation of Hodgkin's disease: a report of nine cases and review of the literature. *Eur Resp J* 1994;7:1852-5.
12. Bonaventura I, Mir I, Marcos JA, Bastus R, Rami R, Martínez I, et al. Tumor de Pancoast: dificultades para su diagnóstico. *Rev Clin Esp* 1993;193:435-7.