Ann Thorac Surg
2003;75:1018–9

CASE REPORTS

Surgical Resection of a Solitary Plasmacytoma Originating in a Rib of a Patient With Castleman's Disease

Simon Wilkinson, MRCS, and Christopher P. Forrester-Wood, FRCS

Department of Thoracic Surgery, Bristol Royal Infirmary, Bristol, United Kingdom

Castleman's disease is a rare disorder characterized by lymphoid hyperplasia. It may present as asymptomatic involvement of one lymph node group or as a multicentric disease with systemic features. We report a patient with Castleman's disease who presented with axillary lymphadenopathy associated with a solitary plasmacytoma originating from a rib. The affected rib was surgically resected and radical radiotherapy was subsequently administered to the axillary lymph nodes. In this particular case, a joint surgical and oncologic approach resulted in a successful outcome.

(Ann Thorac Surg 2003;75:1018–9)

Castleman's disease is a heterogeneous group of disorders characterized by unregulated growth of lymphoid tissue. We encountered a solitary plasmacytoma in a rib of a patient with Castleman's disease, a rare association.

A 35-year-old, otherwise healthy man presented with axillary lymphadenopathy, hepatosplenomegaly, and skin erythema. On physical examination, multiple firm lymph nodes were palpated in the right axilla, and erythema was seen over the right chest wall.

No abnormalities were detected in peripheral blood. A chest roentgenogram showed a solitary expanding lesion in the right 10th rib corresponding to the area of erythema (Fig 1). Computed tomographic imaging of the chest and abdomen demonstrated multiple discrete lymph nodes in the right axilla (Fig 2). The liver and spleen were both enlarged and a focal lytic lesion was confirmed in the 10th rib (Fig 3). The radiologic features suggested a malignant process of metastatic or lymphoproliferative nature. Axillary lymph nodes were biopsied, yielding a diagnosis of Castleman's disease of the hyaline vascular type.

Whole-body bone scintigraphy revealed a solitary abnormal accumulation and, therefore, a surgical approach was taken to remove the rib lesion. At operation, a right thoracotomy incision was performed over the 10th rib. The underlying tissues were edematous and hemorrhagic. A mass was palpable in the 10th rib, also extending up on to the 9th rib. Three quarters of the 9th and 10th ribs were resected en bloc with the surrounding tissue. The postoperative course was uneventful.

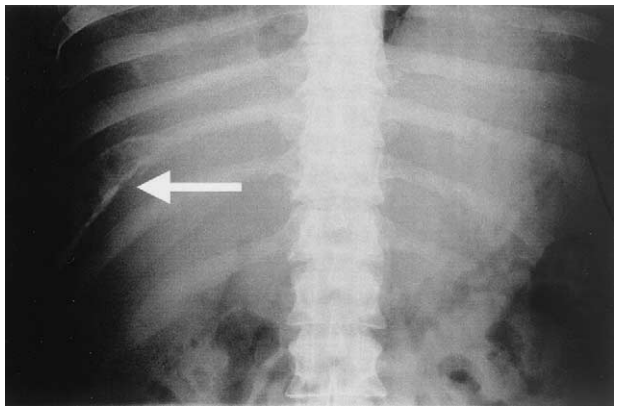

Fig 1. Chest roentgenogram showing a solitary expanding lesion in the right 10th rib (arrow).

The resected specimen contained an abnormal rib expanded to approximately 2.4 cm in diameter. The cut surface showed the medulla to be replaced by a uniformingly pale tumor. Histologic examination showed complete replacement of the medulla by a dense population of mildly atypical plasma cells together with large masses of interstitial amyloid. These characteristics were consistent with a plasmacytoma. The resection margins were negative.

After recovery from surgery, radical radiotherapy was administered to the axillary lymph nodes affected by Castleman's disease. The patient remains well 2 years after surgery, with no evidence of recurrence.

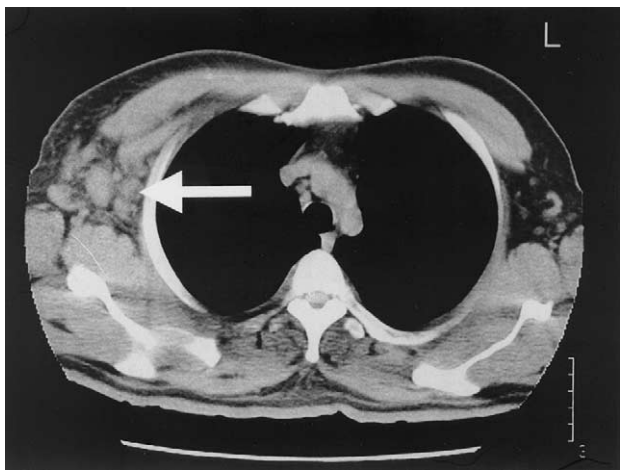

Fig 2. Computed tomographic image of the chest showing multiple discrete lymph nodes in the right axilla (arrow).

Accepted for publication Sept 6, 2002.

Address reprint requests to Mr Wilkinson, 57 Oakfield Rd, Clifton, Bristol BS8 2BA, UK; e-mail: simonmwilkinson@hotmail.com.

Published by Elsevier Science Inc

0003-4975/03/$30.00
PII S0003-4975(02)04476-4

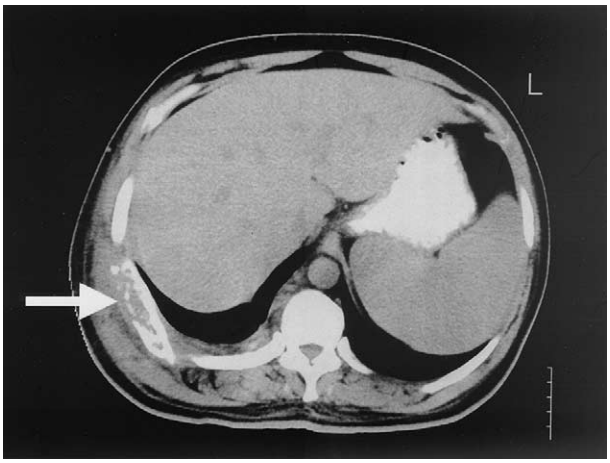

*Fig 3. Computed tomographic image of the abdomen showing hepatosplenomegaly and a focal lytic lesion in the 10th rib (*arrow*).*

Comment

Castleman's disease is a rare lymphoproliferative disorder that was first described in 1954 [1]. Clinical manifestations vary from a localized mass to a systemic disorder with widespread lymphadenopathy, fevers, recurring infections, and autoimmune manifestations. It can affect any part of the body that contains lymphoid tissue, although 70% of cases are intrathoracic [2].

Two variants of Castleman's disease have been described based on histologic features of the affected lymph nodes: a hyaline vascular type and a plasma cell type [2]. The hyaline vascular variant of the disease accounts for 90% of cases and is usually characterized by a benign clinical course with no constitutional symptoms. The clinically more aggressive plasma cell variant accounts for 10% of cases and is frequently associated with systemic manifestations and an uncertain prognosis. Either type may, however, be localized or widespread. When the disease is widespread or "multicentric," the course tends to be more aggressive.

The etiology of Castleman's disease remains elusive, although dysregulated overproduction of interleukin-6 (IL-6) is thought to be central to disease progression [3]. The multicentric variant of Castleman's disease is associated with human herpes virus 8 in many cases [4]. This virus encodes a functional analogue of IL-6, providing further evidence that this cytokine has a pivotal role in the disease.

Functioning as a B-cell growth and differentiation factor, IL-6 has also been implicated in the pathophysiology of B-cell neoplasms, including plasmacytoma [5]. Therefore, it appears that IL-6 may represent a common link between Castleman's disease and plasmacytoma, although only a few cases of this association have so far been reported in the literature [6, 7].

Surgical resection is the mainstay of treatment for patients with localized Castleman's disease and is curative in most cases [8]. Radiation therapy has been used with mixed success in patients who are poor surgical candidates or in those with unresectable lesions [9].

Patients with multicentric disease are sometimes successfully treated with corticosteroids. Patients who do not respond to corticosteroids can be treated with combination chemotherapy regimens [10]. Other treatments include retinoic acid [11], humanized anti–IL-6 receptor antibodies [3], anti–IL-6 antibodies [12], and bone marrow transplantation [10].

Patients with a solitary plasmacytoma have a good indication for surgical intervention, and complete resection is expected to be curative.

In conclusion, Castleman's disease is a rare cause of lymphadenopathy, and the diagnosis is usually one of exclusion after all other causes have been eliminated. The correct diagnosis, however, is indispensable, as localized disease can be cured by surgical resection or radiotherapy. The role of chemotherapy for multicentric disease remains inconclusive. Solitary plasmacytomas are rarely associated with Castleman's disease but should be considered in the differential diagnosis of bone lesions associated with this disease. Surgical resection of these localized bone lesions can be curative.

References

1. Castleman B. Hyperplasia of mediastinal lymph nodes. N Engl J Med 1954;250:26–30.
2. Keller AR, Hochholzer L, Castleman B. Hyaline-vascular and plasma-cell types of giant lymph node hyperplasia of the mediastinum and other locations. Cancer 1972;29:670–83.
3. Nishimoto N, Sasai M, Shima Y, et al. Improvement in Castleman's disease by humanized anti-interleukin-6 receptor antibody therapy. Blood 2000;95:56–61.
4. Soulier J, Grollet L, Oksenhendler E, et al. Kaposi's sarcoma-associated herpes virus-like DNA sequences in multicentric Castleman's disease. Blood 1995;86:1276–80.
5. Burger R, Wendler J, Antoni K, et al. Interleukin-6 production in B-cell neoplasias and Castleman's disease: evidence for an additional paracrine loop. Ann Hematol 1994;69:25–31.
6. Dupuy O, Dourthe LM, Mayaudon H, et al. Castleman disease and solitary plasmacytoma of the acromion. Rev Med Interne 1999;20:83–4.
7. Gould SJ, Diss T, Isaacson PG. Multicentric Castleman's disease in association with a solitary plasmacytoma: a case report. Histopathology 1990;17:135–40.
8. Bowne WB, Lewis JJ, Filippa DA, et al. The management of unicentric and multicentric Castleman's disease: a report of 16 cases and a review of the literature. Cancer 1999;85:706–17.
9. Chronowski GM, Ha CS, Wilder RB, Cabanillas F, Manning J, Cox JD. Treatment of unicentric and multicentric Castleman disease and the role of radiotherapy. Cancer 2001;92:670–6.
10. Repetto L, Jaiprakash MP, Selby PJ, Gusterson BA, Williams HJ, McElwain TJ. Aggressive angiofollicular lymph node hyperplasia (Castleman's disease) treated with high dose melphalan and autologous bone marrow transplantation. Hematol Oncol 1986;4:213–7.
11. Rieu P, Droz D, Gessain A, Grunfeld JP, Hermine O. Retinoic acid for treatment of multicentric Castleman's disease. Lancet 1999;354:1262–3.
12. Beck JT, Hsu SM, Wijdenes J, et al. Brief report: alleviation of systemic manifestations of Castleman's disease by monoclonal anti-interleukin-6 antibody. N Engl J Med 1994;330:602–5.