

Fig 4.

cutaneous CD30-positive lymphoproliferative disorders.<sup>1</sup> These entities constitute a spectrum of related cutaneous conditions originating from a transformed or activating CD30-positive T-lymphocyte. LP is a chronic recurrent skin disease characterized by appearance of papules and/or nodules, which regress spontaneously, typically within 3 to 6 weeks. Lesions greater than 2.5 cm may not regress completely and may leave residual scars with hypo- or hyperpigmented skin.<sup>2</sup> Histologically, there is an atypical T-cell infiltrate, which mimics a T-cell lymphoma. The putative cell of origin is an activated skin homing T-lymphocyte.<sup>3</sup> Fully developed papules show wedge-shaped dermal infiltrates of atypical T-cells admixed with varying proportions of inflammatory cells such as neutrophils, eosinophils, macrophages, and small lymphocytes.<sup>4</sup> The atypical T-cells may either simulate RS features akin to Hodgkin's lymphoma or their nucleus may resemble the cerebriform appearance of mycosis fungoides. The presence of RS-like cells along with numerous inflammatory cells constitutes type A lesions, whereas accumulation of cells with cerebriform nuclei in the absence of inflammatory cells exemplifies type B lesions.<sup>5</sup> Both lesions may exist in the same patient. Immunophenotypic features include CD3-positive, CD4-positive, and CD8-negative T-lymphocytes. CD30 is present in type A lesions but may be absent in type B

lesions. Anaplastic lymphoma kinase protein is consistently absent. Although clonally arranged *TCR* genes have been noted in the majority of type B lesions and occasionally in type A lesions, the t(2;5) translocation, which is a characteristic feature of ALCL, is universally absent.<sup>6</sup> Due to the overlapping clinical (waxing and waning in borderline lesions), histologic (RS-like cells in Hodgkin's lymphoma and cerebriform cells in mycosis fungoides), and immunohistochemical (CD30-positive in ALCL and Hodgkin's lymphoma) features, the diagnosis of LP requires vigilance by the hematopathologist and the oncologist to avoid unnecessary cytotoxic therapy, as highlighted in this case.

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## CASE 2. Mediastinal Liposarcoma in a Patient With Previous Testicular Cancer

A 40-year-old man was treated in January 2002 with three cycles of adjuvant chemotherapy (BEP: bleomycin, etoposide, platinum) because of a nonseminomatous testicular cancer pathologically staged as pT1. His brother had died at age 34 years with a diagnosis of

metastatic testicular seminoma. Familial and personal history was otherwise unremarkable. Patient was followed, and a chest computed tomography (CT) scan in March 2004 showed a solid and homogeneous anterior mediastinal mass measuring 7 cm at longest diameter (Fig 1, arrow). He had no symptoms. Recurrent tumor, thymic rebound, and thymic tumor were suspected. To differentiate thymic hyperplasia—frequently found in

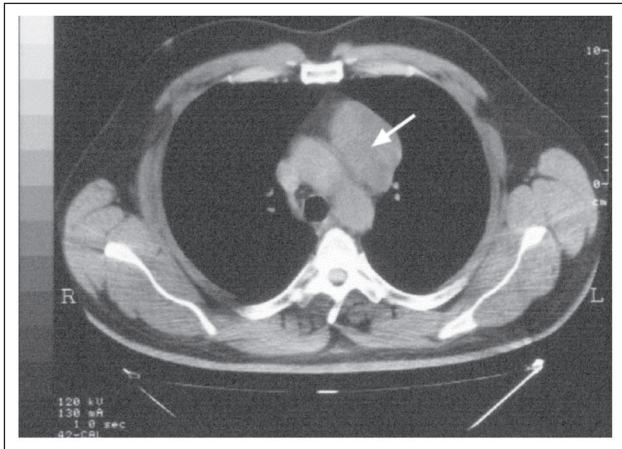


Fig 1.

adult patients treated with chemotherapy<sup>1,2</sup>—from other mediastinal masses, the patient underwent [<sup>111</sup>In-DTPA-D-Phe1]-octreotide scintigraphy according to previously described methods.<sup>3</sup> The images collected after 24 hours from tracer injection revealed uptake in the region corresponding to the left hilum, thus suggesting more a pathologic process than a benign thymic hyperplasia (Fig 2, arrow). CT-guided, fine-needle biopsy was not conclusive. Therefore, surgical intervention was carried out. At exploration, a bumpy mass measuring approximately 9 cm appeared to be closely related to left thymic horn and phrenic nerve. The mass was partly solid with cystic areas filled by mucoïd material. Microscopically, atypical spindled cells similar to lipoblasts were scattered within a diffuse myxoid matrix (Figs 3A and 3B). By immunohistochemistry, vimentin and S-100 (Fig 3C) protein were expressed while carcinoembryonic antigen (Fig 3D), epithelial membrane antigen, and high and low molecular weight cytokeratins were not. The

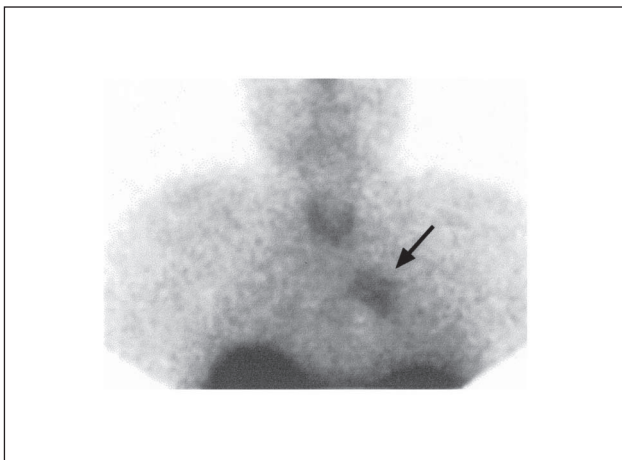


Fig 2.

histologic appearance and immunohistochemical profile were indicative of myxoid liposarcoma. Thymic residual tissue removed with the mass was free from tumor.

Liposarcoma is the most commonly diagnosed soft tissue sarcoma in adults and occurs predominantly in the lower limbs and retroperitoneum.<sup>4</sup> Whereas neurogenic tumors, thymoma, and lymphoma are found in approximately 65% of mediastinal tumors, mesenchymal neoplasms account for approximately 6%.<sup>4</sup> Malignant liposarcoma develops more commonly in the posterior mediastinum. Myxoid liposarcoma accounts for 40% to 50% of all liposarcomas and has been characterized by a t(12:16)(q13-14; p11) translocation.<sup>4</sup> Mediastinal liposarcomas are rare: there are less than 100 cases published worldwide. Liposarcomas of the anterior mediastinum to our knowledge have not been reported.<sup>5</sup> While in early stages they are often asymptomatic, when growing to large size they exhibit various clinical symptoms mimicking lung or heart disease by compression of adjacent intrathoracic organs. This behavior may be attributed to their expansive rather than infiltrative growth. The myxoid liposarcomas seem to have a somewhat more aggressive course than the well-differentiated tumors.<sup>6</sup> Some features displayed by CT imaging may be helpful in differentiating the fat-containing masses arising in the mediastinum.<sup>7</sup> However, the definitive confirmation comes only from pathologic examination. In our case, somatostatin-receptor scintigraphy revealed tracer uptake by the mass. In vivo expression of somatostatin receptor was identified in 71% of 17 studied patients with metastatic sarcoma.<sup>8</sup> However, no liposarcoma was included in this series. In vitro, one group did not find somatostatin-receptor expression in four liposarcomas by autoradiography. The different subtypes of somatostatin receptors appear to be variably expressed in a study on 31 human soft tissue tumors evaluated by reverse transcription polymerase chain reaction. In particular, four of seven liposarcomas expressed the somatostatin receptor subtype 2 mainly responsible for in vivo visualization, and two of these cases were positive for all five subtypes of somatostatin receptor. Besides the role in detection for thymic mass, [<sup>111</sup>In-DTPA-D-Phe1]-octreotide scintigraphy could functionally define mediastinal masses of different histotypes, and its role merits further studies.

The metachronous association of testicular cancer and mediastinal liposarcoma in the same patient has not been described previously. The absence of significant environmental and therapeutic risk factors (no radiotherapy and only three BEP cycles) and the concomitant occurrence of germ cell tumors in two brothers outline, once again, the need for detailed investigation in the



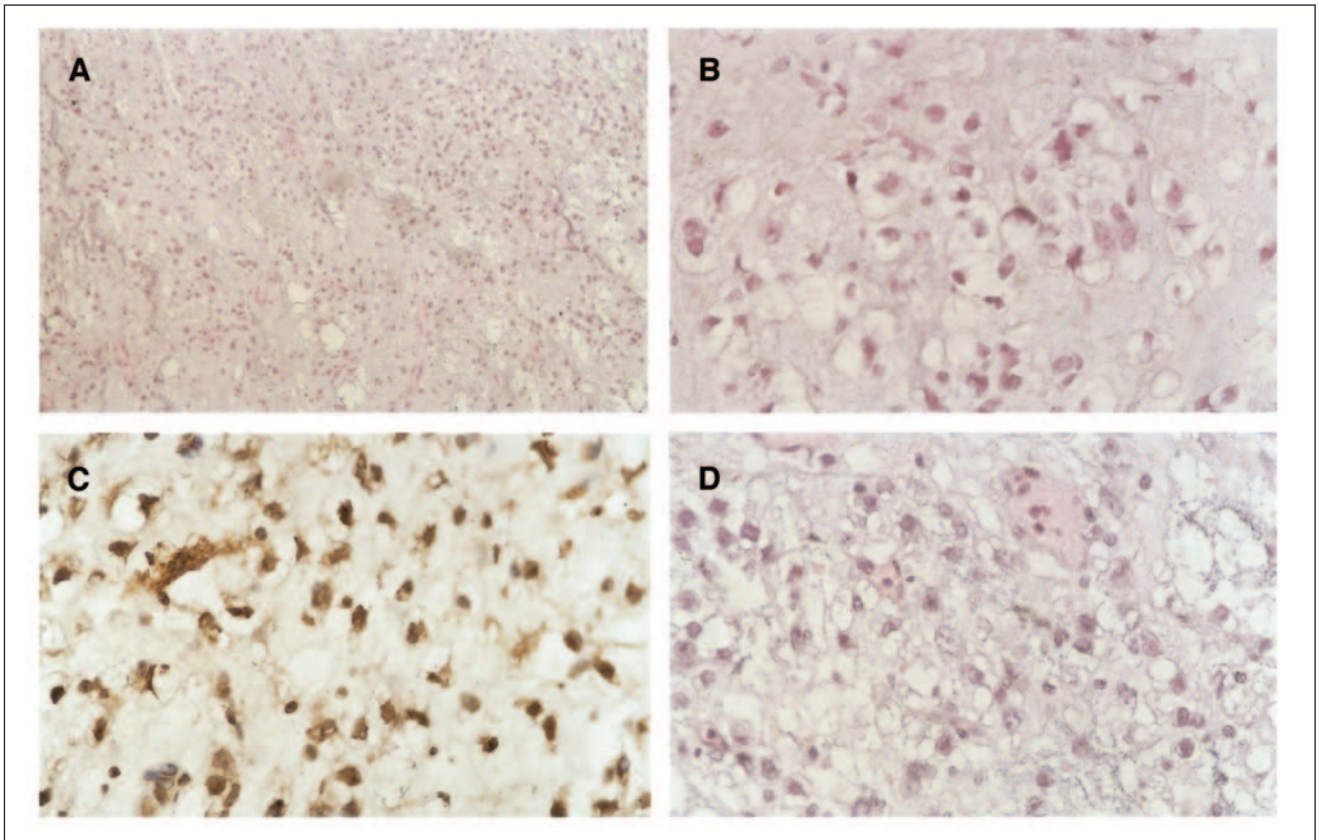


Fig 3.

intriguing field of cancer epidemiology, genetic diseases, and tumor antecedents.

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