Asymptomatic chest wall Hodgkin disease mimicking fibrosing mediastinitis

Charalambos Zisis, MD, PhD,^a Emmanuel M. Kefaloyannis, MD,^a Dimitra Rontogianni, MD, PhD,^b Fani Michalopoulou, MD,^b Ioannis Kalomenidis, MD, PhD,^c Spyros A. Papiris, MD, PhD,^c and Ion Bellenis, MD, PhD, FETCS,^a Athens, Greece

Primarily mediastinal Hodgkin disease (HD) is usually located in the middle or anterior mediastinal compartments and is the most common mediastinal lymphoma,¹ whereas fibrosing mediastinitis is a benign, slowly progressing inflammatory process characterized by excessive fibrotic tissue in the mediastinum. The disease has a tendency to invade and obliterate normal structures such as pulmonary arteries, veins, trachea, main bronchi, superior vena cava, and esophagus.² Imaging and clinical and histopathologic features sometimes overlap with HD.

Clinical Summary

A 36-year-old white man had a slowly growing mass on the right anterior thoracic wall present during the previous 6 months. The patient was free of symptoms, and the chest x-ray film showed mediastinal widening. A computed tomographic scan of the chest revealed a large mediastinal mass occupying the whole anterior mediastinum, invading the anterior thoracic wall and the soft tissues (muscles, subcutaneous fat), and extending into both hemithoraces (Figure 1).

The tumor was not calcified, and developed around the trachea, main bronchi, and superior vena cava in the middle mediastinal compartment. Bronchoscopic examination showed external compression of the trachea and both main bronchi, without an endobronchial lesion. An open biopsy on the right anterior thoracic wall was performed in another hospital and two separate samples were taken (approximately 2 × 2 cm).

Histologic examination showed an inflammatory and fibrotic process with the presence of T and B lymphocytes, compatible with sclerosing or fibrosing mediastinitis.

Median sternotomy was performed and a tumor solid and hard in consistency was found to be densely adherent to the aorta, proximal superior vena cava, and brachiocephalic artery, with invasion of the pericardium. A large part of the tumor was excised, including part of the pericardium. The histologic examination

Figure 1. Chest computed tomographic scan demonstrating the tumor occupying the anterior mediastinum and invading the right anterior chest wall.

showed dense fibrotic tissue containing inflammatory cells with patchy distribution, as well as aggregates of epithelioid cells. The most important finding was the presence of a few mummified cells, as well as very few cells with features of lacunar cells (Figure 2).

Immunohistochemical studies revealed CD30-ki-1 positivity of the few neoplastic cells, whereas the cells were negative to CD15, epithelial membrane antigen (EMA), CD20, CD79a, CD45, CD3, and latent membrane protein–Epstein-Barr virus (LMP-EBV).

The above findings were consistent with the diagnosis of Hodgkin lymphoma nodular sclerosis type, grade I, according to the World Health Organization classification.

Discussion

Nodular sclerosing HD is the most common subtype of HD and has a predilection for the anterior mediastinum, especially the thymus. HD presents as lymphadenopathy or as a discrete mass. Superior vena cava syndrome and chest wall invasion are uncommon.

Although suspicion of the disease can arise from the imaging features, the histologic confirmation may prove strenuous. As nodular sclerosing HD is characterized by a large inflammatory cell reaction within a fibrotic stroma, extensive fibrosis may be the dominant histologic feature. Choice of the suitable surgical access is a difficult point at the decision-making process. In many cases the histologic characteristics of the tumor mandate total resection, whereas for lymphomas a representative specimen is required for an accurate diagnosis to be achieved. Mediastinoscopy, mediastinotomy, video-assisted thoracic surgery, thoracotomy, and even

From the Departments of Thoracic and Vascular Surgery^a and Pathology,^b Evangelismos General Hospital, and the Department of Chest Disease and Intensive Care Unit,^c University of Athens, Evangelismos General Hospital, Athens, Greece.

Received for publication Aug 30, 2005; accepted for publication Sept 9, 2005.

Address for reprints: Charalambos Zisis, MD, PhD, 17a, Patriarchou Grigoriou str, 166 74 Glyfada, Greece (E-mail: chzisis@otenet.gr).

J Thorac Cardiovasc Surg 2006;131:e1-2

0022-5223/\$32.00

Copyright © 2006 by The American Association for Thoracic Surgery

doi:10.1016/j.jtcvs.2005.09.051

Figure 2. Fibrosclerotic tissue with the presence of a lacunar type cell. (Hematoxylin-eosin stain; original magnification $\times 100$.)

sternotomy are the alternatives; a careful and paced procedure is essential to fulfill our diagnostic and therapeutic aims.³

Nearly 10% of the patients with primary mediastinal lymphoma are free of symptoms and the disease is an incidental finding. In the remainder, compression symptoms from the mediastinal anatomic structures are present. It is extremely uncommon for such a sizeable tumor with extended invasion and palpable mass of the muscular and skeletal elements of the anterior chest wall to occur without any symptoms or even pain.

Histologically, the mass had features of mediastinal fibrosing mediastinitis. The previous biopsy specimens obtained showed extended fibrosis with inflammatory reaction. Neoplastic cells were not identified and the findings were compatible with a

chronic sclerosing mediastinal disease. The clinical, radiologic, and histologic findings are similar to HD, and distinguishing between them can be very problematic. Such a case with HD masquerading as sclerosing mediastinitis has been reported once⁴; in that case biopsy from the celiac node had revealed the correct diagnosis of the nodular sclerosing type of HD.

The prognosis for patients with chest wall invasion from HD is suboptimal. Chest wall invasion is considered an adverse prognostic factor among clinical stage I-II Hodgkin lymphoma patients treated with combined modality therapy,⁵ but in all published cases the patients had symptoms.

Conclusion

Distinguishing Hodgkin lymphoma from fibrosing mediastinitis may be very difficult, and safe documentation may demand aggressive surgical management to obtain the correct diagnosis. Thorough histopathologic study and representative tissue material are the prerequisites of an efficacious assessment.

References

1. Strollo DC, Rosado-de-Christenson ML, Jett JR. Primary mediastinal tumors. Part II. Tumors of the middle and posterior mediastinum. *Chest*. 1997;112:1344-57.
2. Rossi SE, McAdams HP, Rosado-de-Christenson ML, Franks TJ, Galvin JR. Fibrosing mediastinitis. *Radiographics*. 2001;21:737-57.
3. Rendina EA, Venuta F, DeGiacomo T, Ciriaco PP, Pescarmona EO, Francioni F, et al. Comparative merits of thoracoscopy, mediastinoscopy, and mediastinotomy for mediastinal biopsy. *Ann Thorac Surg*. 1994;57:992-5.
4. Flannery MT, Espino M, Altus P, Messina J, Wallach PM. Hodgkin's disease masquerading as sclerosing mediastinitis. *South Med J*. 1994; 87:921-3.
5. Hodgson DC, Tsang RW, Pintilie M, Sun A, Wells W, Crump M, et al. Impact of chest wall and lung invasion on outcome of stage I-II Hodgkin's lymphoma after combined modality therapy. *Int J Radiat Oncol Biol Phys*. 2003;57:1374-81.