Mediastinal follicular dendritic cell sarcoma

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A 35-year old man was admitted to our hospital with a right cervical mass and a 4 month history of a cough and mild dysphagia. One year earlier, he had been submitted to right hemi-thyroidectomy for an encapsulated follicular carcinoma of the thyroid gland, with negative lymph-nodes.

At the time of admittance to our hospital he showed a good performance status with blood cell count and biochemical tests within the range of normal values. Physical examination revealed a right cervical mass of about 4 cm of maximum size. A total-body CT scan showed enlarged and confluent mediastinal nodes, with diffuse hypodense areas, extending from the right supraclavicular area to the tracheal carena (figure 1, panel A). A fibrobronchoscopy showed a stenosis and rotation of the trachea, without any endoluminal lesions. A biopsy of the cervical mass was performed. Histologic examination showed a relatively uniform population of oval to spindle neoplastic cells, admixed with reactive T-lymphocytes (figure 1, panels B-C). The neoplastic cells were arranged in a vaguely nodular, storiform pattern, with focal necrosis and nuclear pleomorphism . Tumor cells were diffusely immunoreactive for vimentin, CD21, CD23 and CD35, focally for neuron-specific enolase, epithelial membrane antigen and CD68 (KP-1), and not for actin, myeloperoxidase, S-100, ALK-1, CD1a, CD3, CD5, CD20, CD30, CD68R (PGM-1), HMB 45, desmin, actin, placental alkaline phosphatase, thyreoglobulin and low-molecular weight cytokeratins. Histopathologic and immunohistochemical features were diagnostic for a follicular dendritic cell sarcoma (FDCS).

Chemotherapy with cyclophosphamide, adriamycin, vincristine and prednisone (CHOP regimen) was given for one course, without any response. A second-line chemotherapy with ifosfamide and epirubicin was consequently administered for two cycles, but once again, no change was observed . At thoracotomy, surgical resection was hampered by tumor infiltration of trachea and esophagus; only a biopsy was taken, which confirmed FDCS. Due to the occurrence of disphagia and cough after surgery, a radiation therapy on the mediastinal area was started (planned dose 45 Gy), but the treatment was stopped after two weeks (18 Gy) due to symptomatic worsening, and to the occurrence of mild fever. Oral amoxicillin was given together with supportive care; a severe Stevens-Johnson syndrome developed. The patient died one month later (at seven months from diagnosis) from a generalized sepsis. Autopsy was not performed.

FDCS is a rare neoplasm originating from follicular dendritic cells (FDC) [1]. FDC are located in primary and secondary lymphoid follicles, and play an important role in immune response, acting as antigen-presenting cells for the B-cell compartment. FDCS exhibits distinctive immunohistochemical features: diagnosis relies on the reactivity for FDC markers (mainly CD21 and CD35) [2]. The most common presentation is lymph-node enlargement, even though extranodal sites of disease are frequent as well (mainly head and neck locations). Only 5 cases of mediastinal localization have been reported so far in literature [3-5]. In spite of the clinical and radiological pattern, the behavior of FDCS resembles that of soft tissue sarcomas rather than that of lymphomas [6]. A significant recurrent and metastatic potential has been reported [5]. The optimal therapy for FDCS has yet to be defined. Complete resection, when feasible, is the treatment of choice. The role of adjuvant chemotherapy and radiotherapy is not well established, because of the rarity of the disease [5]. Treatment should be tailored to the individual patient according to the putative predictors of aggressive behavior, such as intra-abdominal location, high mitotic count, and presence of significant necrosis and cellular atypia [7]. Variable degrees of response to the chemotherapy regimens commonly used for lymphomas or soft tissue sarcomas have been reported in unresectable patients; in these cases, the role of radiation therapy is unclear [5].

This case suggests that FDCS, although uncommon, should be considered in the differential diagnosis of mediastinal masses arising within lymphoid milieu.

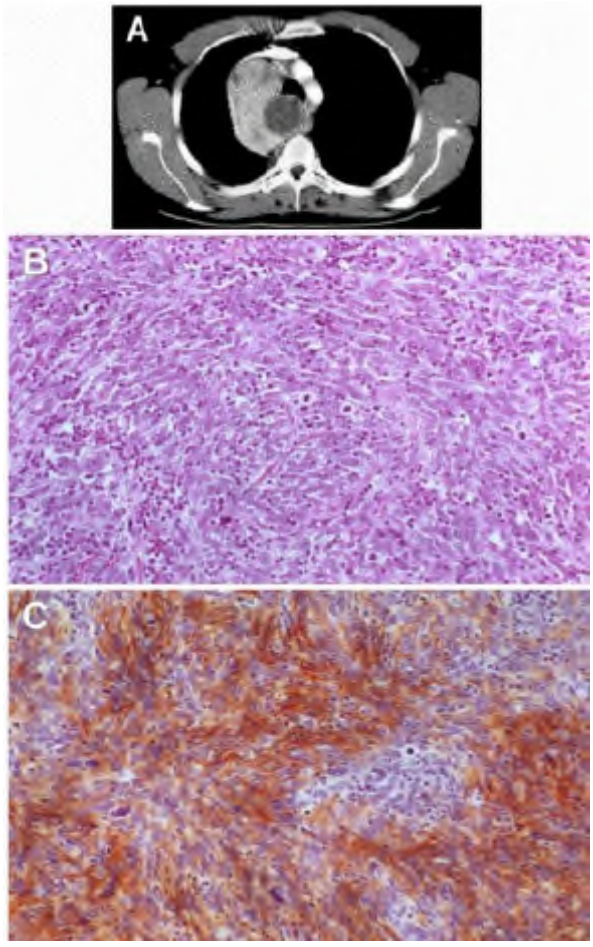

Figure 1. CT scan showing a mediastinal mass with hypodense areas (A). Neoplastic cells in a storiform pattern with admixed lymphocytes (B) are immunoreactive for CD21 (C).

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