CASE 1. ANTerior MEDIASTINAL RHABDOMYOSARCOMA

A 21-year-old white female presented with a 9-month history of progressively worsening vague chest pressure and discomfort, dyspnea, anorexia, and a 20-lb weight loss. Her medical history was unremarkable, and physical examination was normal, aside from a tachycardia of 120 beats per minute. Computed tomography (CT) of the chest revealed a large, somewhat lobulated, homogeneous anterior mediastinal mass measuring approximately $13 \times 10 \times 8$ cm (Fig 1). Despite its size, the mass seemed resectable, without definite invasion of adjacent structures. CT-directed fine-needle aspiration biopsy was performed, and histopathologic examination of the biopsy specimen showed a neoplasm with prominent myxoid differentiation without evidence of lymphoma or thymoma. Given this tentative diagnosis of a soft tissue sarcoma, the resectable appearance of the mass on CT, and a negative metastatic work-up, surgical resection via median sternotomy was undertaken. At the time of operation, the mass appeared larger than reported by the preoperative CT scan and was found to be densely adherent to the pericardium, the mediastinal pleura, and the left lung. Complete surgical resection with negative margins was achieved by wide en bloc resection of these structures along with the tumor (Fig 2). The patient’s recovery was uneventful. Final pathologic examination revealed a high-grade malignant neoplasm with clear-cut features of skeletal muscle differentiation. An area of focal residual thymus was evident (Fig 3, upper left corner) along with the high-grade malignant neoplasm with large atypical cells (Fig 3, lower portion of slide). High-power magnification revealed large tumor cells with prominent round to oval nuclei and intensely eosinophilic cytoplasm—so-called myoblast strap cells. Cross striations in the myoblasts are not seen in this view, however (Fig 4). The tumor was immunoreactive for common muscle actin and desmin, was negative for smooth muscle actin and keratin (CAM 5.2 and AE1/AE3), but demonstrated convincing nuclear positivity for myogen. Final diagnosis was a primary anterior mediastinal rhabdomyosarcoma, anatomically related to, and likely arising from, the thymus.

Primary mediastinal rhabdomyosarcomas unassociated with germ cell, teratomatous, or malignant epithelial components are extremely rare. They are thought to arise from the thymus; the ability of thymic tissue to differentiate toward myoid tissue has been reported previously. These tumors tend to occur in the anterior mediastinum of young adults and demonstrate large size and local invasion at the time of diagnosis. Their biologic behavior is aggressive, usually characterized by rapid recurrence and dissemination, even following resection. Because of the limited information regarding treatment of these unusual tumors, therapy must be extrapolated from the management of pediatric rhabdomyosarcomas and applied to these patients. This patient is currently receiving combination chemotherapy.
consisting of cyclophosphamide, doxorubicin, and vincristine alternating with ifosfamide and etoposide. Mediastinal and left hemithoracic external-beam radiation therapy will also be administered.

D. Bruce Panasuk, Thomas L. Bauer, Allen L. Davies, Charles Schneider, and Cynthia Flynn
Helen F. Graham Cancer Center, Christiana Care Health Services, Newark, DE

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CASE 2. MANTLE-CELL LYMPHOMA OF THE PROSTATE GLAND

A 73-year-old man with a 6-month history of prostatism (urgency, frequency, hesitancy, and poor urine stream), presented to the emergency department with acute urinary retention. There were no systemic symptoms. Physical examination revealed generalized lymphadenopathy. Groin lymph node biopsy showed effacement of the lymph node by nodular and diffuse infiltration of medium-sized lymphoid cells, which were immunoreactive for CD20, CD5, and cyclin D1, but negative for CD3, CD23, and bcl2. Transrectal prostatic biopsy showed similar lymphomatous infiltrate (Fig 1A and 1B) with the same immunophenotypic profile (Fig 1C [cyclin D1] and 1D [CD5 positivity]). Computed tomography showed a diffusely enlarged prostate gland (Fig 2A [before contrast administration] and 2B [after contrast administration]). Complete staging showed Ann Arbor stage IVA mantle-cell lymphoma (MCL). He is currently in complete remission with complete resolution of obstructive symptoms after six courses of combination chemotherapy (cyclophosphamide, vincristine, procarbazine, and prednisolone).

Lymphoma of the prostate, both primary or secondary, is rare. The rarity of secondary prostatic lymphoma is illustrated in an autopsy study of 6,000 male patients dying of malignancy over a 25-year period: only 185 (3.1%) patients had metastatic prostatic malignancy, of which 49 (0.82% of all autopsied patients) had non-Hodgkin’s lymphoma. This is further illustrated in another study of 1,474 patients with malignant disease of the prostate at the M.D. Anderson Cancer Center (Houston, TX) over a 20-year period, in which 18 nonleukemic secondary prostatic malignancies were identified. In this study, only three were diagnosed during life, and in the remaining 15 patient cases (which represented 0.5% of the 3,663 male patients dying of cancer) prostatic involvement was an incidental finding at autopsy. Moreover, in another study of 1,068 patients for urogenital involvement by lymphoma with evidence either by radiologic study or at operation or autopsy, only two presented with a perineal mass, which might possibly be prostatic involvement. Our patient presented with obstructive symptoms of the urinary tract but he had stage IV disease at diagnosis. According to the criteria of Bostwick and Mann, primary prostatic lymphoma will be diagnosed only if the following criteria...
are fulfilled: primary symptoms are attributable to prostatic enlargement; the major bulk of disease is localized to the prostate; and lymph nodes, liver, or spleen are not involved within 1 month of diagnosis. Therefore, even though our patient presented with symptoms attributable to prostatic enlargement, he should be classified as having secondary prostatic lymphoma.

MCL is one form of small B-cell lymphoma characterized by t(11;14) with dysregulation of the cell cycle by upregulation of the cyclin D1.6,7 Clinically, the majority of patients are elderly with advanced-stage disease. Extranodal involvement of bone marrow, liver, spleen, Waldeyer’s ring, and gastrointestinal tract (in the form of multiple lymphomatous polyposis) are frequent.7 The diagnosis of MCL in our patient was based on the demonstration of small B-cell morphology, and immunoreactivity with cyclin D1, CD5, and CD20. The lack of CD23 expression excluded chronic lymphocytic leukemia, in which leukemic cells are positive for both CD5 and CD23. Regarding the histology of secondary prostatic lymphoma, the majority have non-Hodgkin’s lymphoma.1 In one report of 62 patients with prostatic lymphoma,1 half were secondary, and the majority of patient cases had diffuse large B-cell lymphoma, followed by B-chronic lymphocytic lymphoma or small lymphocytic lymphoma, and then follicular lymphoma.1 In contrast, according to our knowledge, MCL involving the prostate gland is hitherto unreported, and this is the first report in the literature. Urgency and frequency are the most common presentations of lymphoma, with systemic symptoms such as fever, night sweating, and weight loss much less frequent.1 Obstructive symptoms are common in the elderly male population, and are frequently attributed to nodular hyperplasia or prostatic carcinoma. Our patient illustrated that secondary lymphoma of the prostate is a possible cause of urinary outflow tract obstruction. The obstructive symptoms might resolve completely with combination chemotherapy. Moreover, MCL can be added to the list of histologic subtypes in prostatic lymphoma.

C.S. Chim, F. Loong, T. Yau, G.C. Ooi, and R. Liang

*University Departments of Medicine, Pathology, and Radiology, Queen Mary Hospital, University of Hong Kong, Hong Kong.*

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REFERENCES


CASE 3. EPIGASTRIC DISTRESS CAUSED BY A DUODENAL POLYP: A RARE PRESENTATION OF ACUTE LEUKEMIA

A 27-year-old man who was previously healthy presented with a 2-month history of upper abdominal pain, nausea and vomiting, and a weight loss of 7 kg. Physical examination was normal and chemical laboratory tests were normal. The patient’s hemoglobin level was 14.2 g/dL, the platelet count was 152 × 10^9/L, and the leukocyte count was 4.1 × 10^9/L, with a normal automated differential cell count. A gastroscopic examination showed a polypoid mass in the duodenum (Fig 1). Histologic evaluation revealed infiltration by myeloid blast cells (Fig 2A), which were positive for myeloperoxidase (Fig 2B). A bone marrow examination performed after the results of the biopsy were available showed trilineage dysplasia with 30% blasts. Other studies revealed chromosome 16 inversion and a manual differential count of 16% blasts on a peripheral blood smear. The patient received induction therapy with idarubicin and cytarabine. After an uneventful remission a repeat gastroscopic examination was macroscopically normal although biopsy of the duodenal wall still showed some blasts. He was given consolidation therapy with high-dose cytarabine and then underwent allogenic bone marrow transplantation from a matched unrelated donor and has remained without relapse for 2 years.

An extramedullary myeloblastoma (also called a granulocytic sarcoma) is a localized tumor composed of immature myeloid cells that has been associated with myeloid leukemia. It has been reported during the course of acute or chronic leukemia, preceding the diagnosis,
and as a presentation of leukemia, although there have been patients without any bone marrow disease. The common sites of involvement are the skin, bone, periosteum, and lymph nodes, but other tissues may be involved as well. Localization to the intestine was reported as rare in one series but in another it was 17%. Symptoms of granulocytic sarcoma of the small intestine were usually of abdominal pain or intestinal obstruction; bleeding was less common. In most patients, when the presentation was of obstruction, the patients had surgery; in patients in whom resection was not urgently required, the lesions regressed with chemotherapy.

This case report demonstrates the occurrence of granulocytic sarcoma as a presentation of acute leukemia, the importance of performing endoscopic examinations for epigastric distress in otherwise healthy patients in the presence of important symptoms such as weight loss, and the importance of performing manual differential counts rather than relying on automated blood cell counts.

Odelia Goor, Yoav Goor, Fred Konikoff, Leonor Trejo, and Ella Naparstak
Departments of Hematology, Internal Medicine 6, Gastroenterology, and Pathology, Tel Aviv Sourasky Medical Center
Tel Aviv, Israel

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