Granulocytic sarcoma of the adrenal gland

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We report a case of primary granulocytic sarcoma (GS) of the left adrenal gland, with no evidence of hematologic involvement. To our knowledge, this is the first case of granulocytic sarcoma of the adrenal gland.

Key Words: adrenal gland, granulocytic sarcoma, retroperitoneal hemorrhage

Introduction

Granulocytic sarcoma (GS) is a rare solid tumor composed of neoplastic myeloid cells and was first linked to myeloid neoplasms in 1893 by Dock.[1] This tumor has been referred to by a variety of names including myeloblastoma, chloroma (due to its characteristic greenish hue), or chloroleukemia. GS is a rare occurrence with an estimated incidence of 0.7 per million in children and 2 per million in adults. This tumor occurs in about 5% of myeloid leukemias in adults and 13% of children with myeloid leukemias.[2,3] In autopsy series, granulocytic sarcoma occurs in 2% to 8% of patients with acute (AML) and chronic myelogenous leukemia (CML).[3,4]

Case presentation

A 54-year-old obese white male presented with acute onset of left flank pain, hypertension, tachycardia, and anemia. Cat scan of the abdomen revealed a large retroperitoneal hemorrhage associated with a large left adrenal mass, Figure 1. Emergency angiography was negative for arterial bleeding. Once the patient stabilized, work up for an underlying pheochromocytoma was performed. The studies were equivocal in diagnosing pheochromocytoma, he was empirically started on alpha and beta blockade. An interval left adrenalectomy was performed. The tumor measured 14 cm and weighed 1091 grams. No adrenal gland was identified on gross examination; however, on histologic examination, necrotic adrenal was identified in association with tumor, Figure 2b. The tumor was of high-nuclear grade with a "starry-sky" pattern, Figure 2a. The initial impression on hematoxylin and eosin stained sections was that of large cell lymphoma. The tumor cells stained positively for CD45 (LCA), CD43, CD60, BA2 & CD117, focally positive for UCHL1 and positive with a Leder stain (chloracetate esterase). The tumor was negative for CD34, CD20, CD27a, CD3, CD30, Alk1, light chains and myeloperoxidase. The findings were consistent with a diagnosis of granulocytic sarcoma with myelomonocytic differentiation.

The patient also underwent bone marrow biopsy aspirate to rule out the presence of a simultaneous hematological disorder. The work up was negative, which included examining cell morphology, cell cytogenetics, and fluorescent insitu hybridization.

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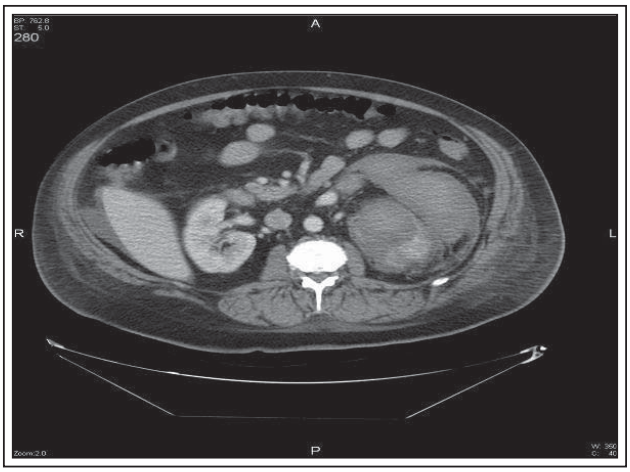

Figure 1. Cat scan of abdomen demonstrating adrenal mass with retroperitoneal hemorrhage.

Discussion

Granulocytic sarcoma is a rare solid tumor and is typically misdiagnosed in patients with no hematological malignancies. The distinction from other tumors, especially other lymphomas, can be critical since it can have profound therapeutic implications. Delay can interfere with the institution of appropriate chemo- and/or radiotherapy.

Immunohistology, whether by flow cytometry or performed immunohistochemically on tissue sections, is always necessary to confirm the diagnosis of granulocytic sarcoma. It had been demonstrated that 100% of the well differentiated and 89% of the poorly differentiated/blastic tumors were CD43 positive.[7]

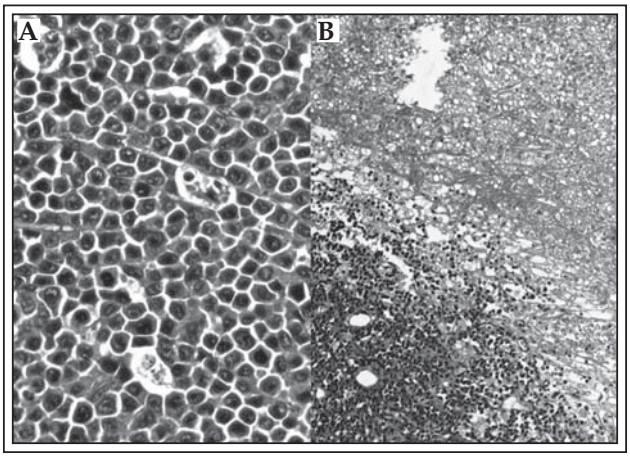

Figure 2. A) Tumor cells and tingible body macropages making a starry sky pattern. 40x magnification. B) Tumor and necrotic adrenal. 10x magnification.

However, CD43 cannot be used singly to make the diagnosis of GS. CD43 positivity in the absence of other T cell lineage markers should raise the possibility of a GS. In a series of 26 GS tumors, all were CD3, CD20, CD30, and CD79a negative.[8] In our case, the tumor was poorly differentiated and was positive for LCA, CD43, and negative for myeloid marker lysozyme and for B and T cell lineage markers. Demonstration of myeloid differentiation and absence of cells marking with B and T cell markers as was seen in our case is required to confirm the diagnosis.[8,9]

In this case, this patient had no previous hematologic abnormalities. The primary site of the tumor was the adrenal gland with no involvement of the retroperitoneal/mesenteric lymph nodes.

Most cases of GS that occur in nonleukemic patients will progress to AML within 1 year.[10,11] Bone marrow biopsy in our patient showed no evidence of malignancy. The patient was treated with a chemotherapy regimen of cytarabine 10 mg/m^2 and idarubacin 13 mg/m^2. Six months postchemotherapy, he has been free from recurrence of GS or AML/CML. □

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