

Small round tumour cells (CD38, CD 79a positive) in the adrenal gland



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1. Introduction

The incidence of primary adrenal lymphoma is a very rare. Primary adrenal lymphoma is usually suspected in a patient with bilateral adrenal masses, with/without lymphadenopathy and with/without any signs of adrenal insufficiency.^{1,2} The most common symptom observed in nearly 70% patients is presence of bilateral adrenal mass. Nearly 50% of the patients present with syndrome of adrenal insufficiency. However histologically most of the cases reported in literature are of large B-cell type; centroblastic or immunoblastic.³ We here report an interesting case of primary adrenal lymphoma which on subsequent histological examination was found to be infiltrating the pancreas.

2. Case report

A 45-year-old man, with insignificant past medical history was referred with fever and fatigue for 3 months, with blood pressure of 130/82 mmHg, pulse rate of 90 bpm and body temperature of 98.6 °F. Systemic examination was unremarkable without evidence

of any lymphadenopathy or skin pigmentation. Per abdomen examination revealed a non-tender, firm abdominal lump, approximately measuring 12 × 9 cm. Hematological investigations revealed hemoglobin of 13.2 gm/dL, white blood count $13.48 \times 10^3/\mu\text{L}$ with mild predominance of neutrophils, platelet count of $4.82 \times 10^6/\mu\text{L}$. Coagulation profile was unremarkable. ESR was 88 mm at the end of 1 h. Serum electrolyte estimation revealed sodium concentration of 128.9 mEq/L (normal range: 135–145 mEq/L) and potassium concentration of 4.2 mEq/L (normal range: 4.5–5.5 mEq/L). Serum phosphate level was 3.5 mg/dL (normal range: 2.5–4.5 mg/dL). Liver and renal function tests were within normal range. Serum lactate dehydrogenase (LDH) level was 572 IU/L (normal range, 110–200 IU/L), serum cortisol concentration at 8 AM was 11.8 µg/dL (normal range, 5–23 µg/dL). The plasma free metanephrine level was 32.6 pg/mL (normal range, <90 pg/mL).

Chest radiography was unremarkable for any mediastinal. Ultrasonography and computed tomography imaging (CT) of the abdomen revealed bilateral hypoechoic adrenal masses; $12.5 \times 9.2 \times 4.5$ ccm (right) and $15.5 \times 10.6 \times 5.5$ ccm (left), infiltrating the pancreatic tail (Fig. 1). The patient underwent surgical laparotomy and the specimen of left adrenal mass with spleen, left kidney and pancreatic tail was sent for histopathological evaluation which weighed 710 g. The left adrenal mass weighed 580 g, measuring $17.5 \times 9.5 \times 8.8$ cms, grey-tan in colour with multiple attached lymph nodes. Cut surface appeared grey-tan. Cut section of left kidney, spleen appeared unremarkable but pancreas was found to be adherent to the adrenal mass at the inferior border. Microscopic examination revealed sheets of monomorphic small round cells having round to oval, occasional cleaved nuclei, with prominent nucleoli and scanty basophilic cytoplasm with mitotic figures (Fig. 2a), separated by delicate fibrous bands containing also infiltrating the pancreatic tissue (Fig. 2b). Sections from kidney, spleen and attached lymph nodes were unremarkable. Immunohistochemical stains were performed for cytokeratin-7, CD3, CD5, CD20, CD38, CD68, CD79a, BCL-2, cyclin D1, leucocyte common antigen (LCA), vimentin, κ light chain, λ light chain, neuron-specific enolase (NSE), S100, synaptophysin and chromogranin (Dako, USA). The monomorphic tumour cells displayed positivity for CD20/38/79a, BCL-2, LCA, vimentin and κ light chain (Fig. 3) and were negative for CD3/5/68, λ light chain, NSE, synaptophysin,

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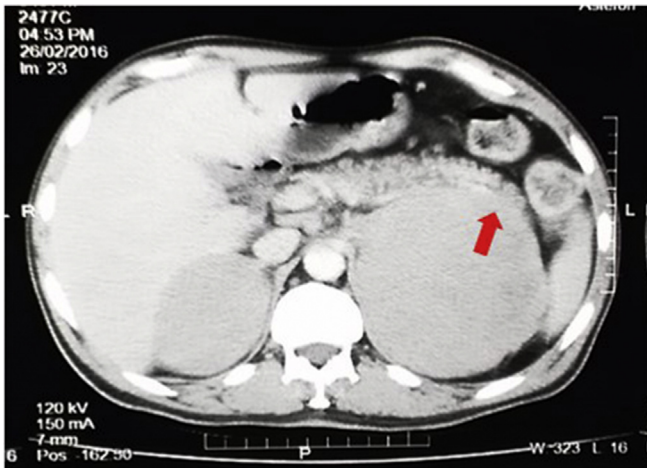


Fig. 1. Computed tomography scan image showing bilateral hypoechoic adrenal masses infiltrating the pancreas on the left side (marked by red arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

chromogranin and S-100. Ki-67 labelling index was 70%. The lymphoma was typed as diffuse large B-cell lymphoma (subtype B-cell) type according to World Health Organization classification. The bone marrow examination and the trephine biopsy showed no evidence of infiltration by lymphoma cells. The patient was treated by CHOP chemotherapy regimen which included Vincristine (1.4 mg/m²) and cyclophosphamide (750 mg/m²), Prednisolone (100 mg/day). He is doing well on the last follow-up 1 year after therapy.

3. Discussion

Adrenomegaly, non-neoplastic as well as neoplastic is one of the differential diagnosis for presence of an abdominal mass. Differential diagnosis of adrenal gland masses include various benign conditions like traumatic hemorrhage, pheochromocytomas, infections, inactive and active adrenal adenomas, adrenocortical hyperplasia, carcinomas, myelolipoma and metastasis from other organs.^{1,4} Males are twice affected than females with median age of affection being 68 years (range: 39–89 years). Bilateral cases comprise nearly 70% of the total adrenal lymphomas. The exact pathogenic mechanism by which lymphoma develops in the

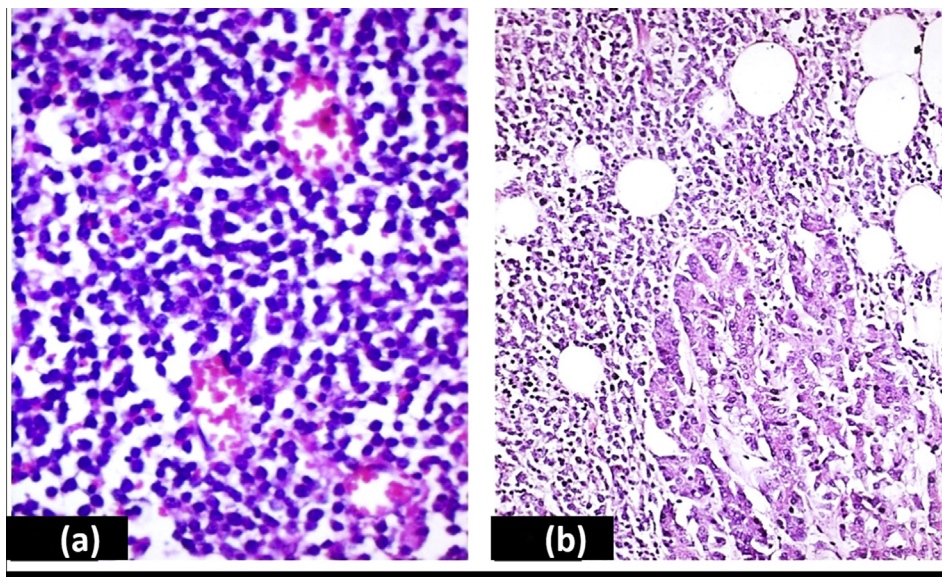


Fig. 2. Monomorphic small round cells with round to oval, occasional cleaved nuclei, with prominent nucleoli and scanty basophilic cytoplasm [Figure 2 (a) Hematoxylin and Eosin, X 400]; tumour cells infiltrating the pancreas [Figure 2 (b) Hematoxylin and Eosin, X 100].

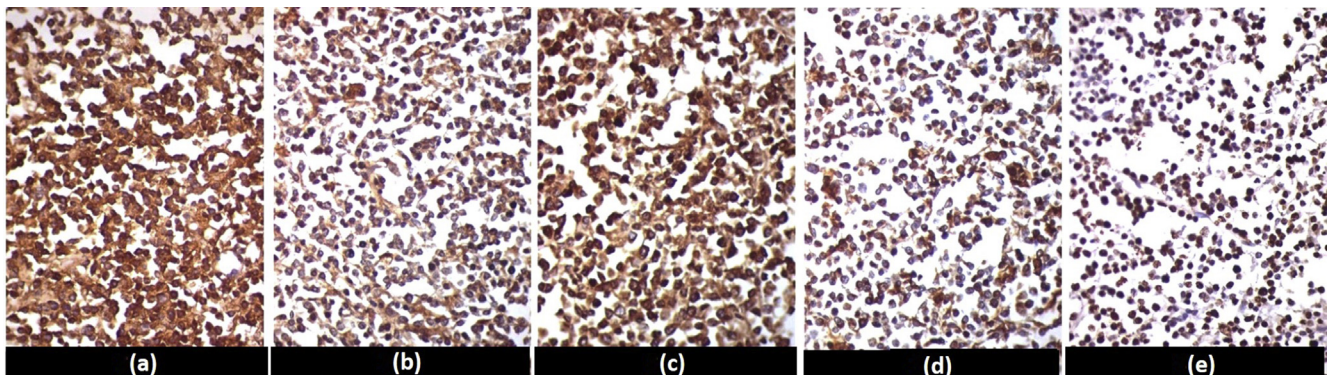


Fig. 3. Immunohistochemistry panel of positive markers (IHC marker, X 200): Vimentin (3a); kappa (3b); CD20 (3c); CD79a (3d) and Ki-67 (3e).

adrenal gland is largely unknown. According to the observations of Ozimek et al., it was proposed that PAL arises from previous autoimmune adrenalitis.⁵ Ellis RD et al. have suggested that these tumors may arise from hematopoietic tissue inherent to adrenal gland and an immune dysfunction could predispose to PAL.⁶

The patients having this tumour clinically present with abdominal pain (26%), fever (46%) and fatigue with weight loss (24%). Few patients may present with symptoms of adrenal insufficiency. Hepatosplenomegaly, lymphadenopathy, concurrent or prior immune dysregulation, and bone marrow involvement are uncommon.⁷ Histologically, most of the tumors (70–90%) of primary adrenal lymphomas are of diffuse large B-cell type. Ludvik et al. reported a case of primary centroblastic adrenal lymphoma in a 76 years old man, which was positive for vimentin, cytokeratin, CD20/38/79a, Bcl-2, LCA and κ light chain but negative for CD3/30/CD45/45RO/68, λ light chain, EMA, chromogranin A, S-100, synaptophysin, HMB-45 and NSE.

Various therapeutic modalities available for these patients include surgical excision, chemotherapy, surgery followed by chemotherapy and/or radiotherapy and corticosteroid replacement. R (Rituximab)-CHOP chemotherapy regimen is found to be useful in many of the cases. However most of the patients have a fatal outcome.³

The most unique feature of our case is the presentation as bilateral adrenal mass without evidence of primary adrenal insufficiency infiltrating the pancreas which may contribute to unfavourable prognosis.

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