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No potential conflicts of interest relevant to this article were reported.

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Primary adrenal T-cell lymphoma in a young adult presented with pseudo-hypopyon: a case report and literature review

TO THE EDITOR: A 26-year-old Indian man presented with weight loss for 6 months and blurred vision on his right eye for 2 weeks. He seemed to be pale, but had no lymphadenopathy or hepatosplenomegaly. Initial complete blood count showed a hemoglobin level of 10.7 g/dL, white blood cell count of $1.4 \times 10^9/L$ (neutrophils 44%, lymphocytes 43%, eosinophils 3%, and monocytes 10%), and platelet count of $40 \times 10^9/L$. Ophthalmic evaluation revealed a white meniscus in the anterior chamber of the right eye. The corresponding aqueous humor aspirate showed infiltration of atypical lymphoid cells (Fig. 1). Whole body fluorine-18-fluorodeoxyglucose (FDG) positron emission tomography/computed tomography (^{18}F -FDG PET/CT) did not re-

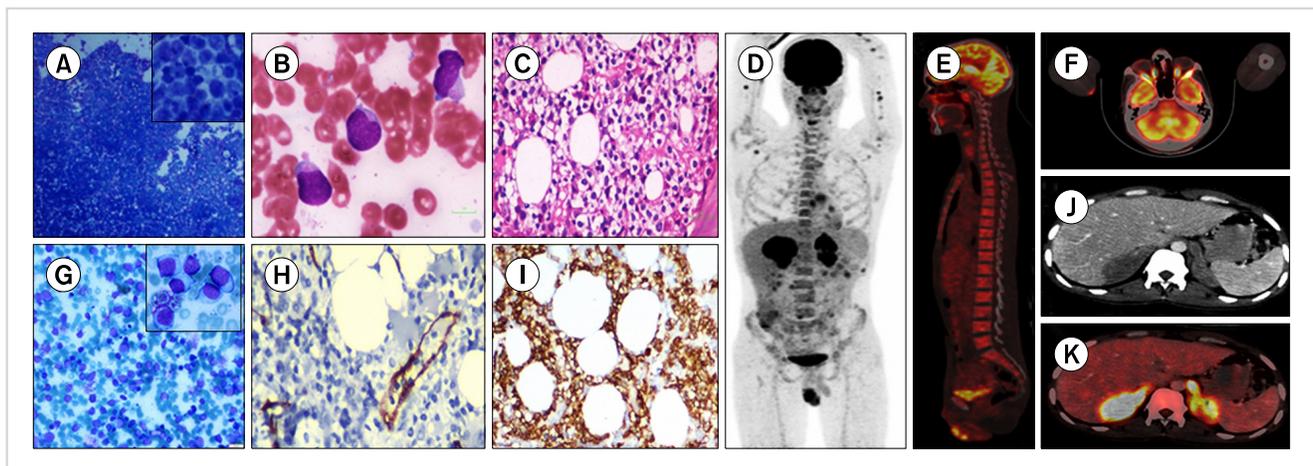


Fig. 1. Microscopic findings, immunohistochemical staining, and whole-body fluorine-18-fluorodeoxyglucose positron emission tomography/computed tomography (^{18}F -FDG PET/CT) of primary adrenal T-cell lymphoma. (A) Aqueous humor aspiration showed infiltration of atypical lymphoid cells. (B) Bone marrow (BM) aspiration showed infiltration of atypical lymphoid cells with irregular nuclear membrane, coarse chromatin, basophilic cytoplasm and fine azurophilic granules. (C) BM biopsy showed infiltration of lymphoma cells with "fried egg" pattern. (D) FDG avid lesions in the adrenal glands. (E) Diffuse FDG uptake in the BM. (F) Absence of FDG avid intra-orbital and intra-ocular mass lesions. (G) Adrenal aspiration showed lymphoma infiltration. (H) CD34 immunohistochemistry (IHC) of the BM biopsy highlighting intra-sinusoidal pattern. (I) CD8 positivity in the BM lymphoid infiltrate. (J, K) Diffuse and intense FDG uptake in both the adrenal glands. (A, B, & G: May-Grünwald Giemsa stain; C: Hematoxylin and eosin stain).

Table 1. Review of clinical and radiologic findings in patients with primary adrenal T-cell lymphoma.

	This case	Sampath <i>et al.</i> [4]	Pimentel <i>et al.</i> [7]	May <i>et al.</i> [8]	Sfasi <i>et al.</i> [9]
Age (yrs)	26	33	42	59	70
Gender	Male	Male	Male	Male	Male
Diagnosis	CD8 positive extranodal T-cell lymphoma	Not sub-classified	Large, cleaved T-cell lymphoma	Centroblastic T-cell lymphoma	Not sub-classified
Symptoms	Weight loss, blurred vision	Weight loss, fever, abdominal pain	Weight loss, fever, vomiting	Asymptomatic	Weight loss, fever
Adrenal insufficiency	No	No	Yes	No	Yes
Side	Bilateral	Bilateral	Bilateral	Right adrenal	Bilateral
Metastasis	Anterior chamber of the right eye	No	CNS (CSF positive) and rectus sheath of both eyes	No distant metastasis, but the mass was adherent to the IVC	No distal metastasis, but the left adrenal mass was adherent to the left renal pedicle
Treatment	Conservative	CHOP	CHOP with intra-theatrical methotrexate	Surgery and radiation	Surgery and chemotherapy
Outcome	Expired due to intra-abdominal hemorrhage	After 3 cycles of therapy, patient was asymptomatic and gained 5 kg	Expired 4 months later due to disease progression	Remission for 8 years	Expired due to multi-organ failure secondary to sepsis
Bone marrow infiltration	Yes	NA	No	No	No

Abbreviations: CHOP, Cyclophosphamide 750 mg/m² per day, vincristine 2 mg per cycle, doxorubicin 50 mg/m² per cycle, prednisone 60 mg per day; CNS, central nervous system; CSF, cerebrospinal fluid; IVC, Inferior vena cava; NA, not available.

veal any FDG avid intra-ocular/intra-orbital lesions. However, there were FDG avid soft tissue masses in both the adrenal glands (4.8×9.3×6.2 cm lesion with maximum standardized uptake value (SUVmax) of 22.7 in the right adrenal gland and 4.6×9.2×5.9 cm lesion with SUVmax of 17.6 in the left adrenal) suggesting a lymphomatous pathology (Fig. 1). CT guided fine needle aspiration from the adrenal glands revealed the infiltration of atypical lymphoid cells, which were CD3 positive and CD20 negative by immunocytochemistry. Bone marrow aspiration showed increased number (about 83%) of 15–20 µm sized atypical lymphocytes (Fig. 1). In flowcytometric analysis, these cells showed expression of CD45, CD2, CD3, and CD8, whereas no expression of CD1a, CD4, CD5, CD7, CD34, CD16, CD56, CD57, TCRαβ and TCRγδ. These lymphoid cells had clear cytoplasm, imparting a 'fried-egg' pattern of arrangement in bone marrow biopsy. On immunohistochemistry (IHC), these lymphoid cells were positive for CD8 and negative for CD3, CD5, CD4, and CD56. Negative staining for CD34 by IHC highlighted vascular proliferation and intra-sinusoidal infiltration (Fig. 1). The patient was diagnosed with CD8-positive extranodal T-cell lymphoma, but the patient died on 2 days of admission, prior to initiating therapy.

Primary adrenal lymphoma (PAL) is a rare and aggressive malignancy and involves bilateral adrenals in 60–75% of cases [1, 2]. The most common PALs are diffuse large B-cell lymphomas (~80%), followed by peripheral T cell lymphoma (7%) [2–4]. Since the majority of intraocular lymphomas (IOL) are of B-cell origin, T-cell type IOLs are very rare and often secondary to cutaneous or adult T-cell lymphoma [5]. Except for a single case report showing choroidal involve-

ment, intraocular metastasis by PAL is extremely rare [6].

To the best of our knowledge, only 4 cases of T-cell type PAL have been reported in literature, but none of the cases had bone marrow involvement (Table 1) [4, 7–9]. The median age at diagnosis was 40 years (range, 31–70 yrs). The most common presentation is weight loss and most of the cases have poor outcome in spite of intensive chemotherapy.

In conclusion, we describe the first case of primary adrenal T-cell lymphoma with ocular and bone marrow metastasis in a young adult. In the absence of clinical evidence to suspect a primary adrenal pathology, our case reinforces the importance of thorough radiologic workup, especially ¹⁸F-FDG PET/CT, in the evaluation of occult lymphomas. In addition, we also emphasize the role of morphological evaluation combined with immunophenotyping for definite diagnosis in lymphomas.

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Combined occurrence of Bernard-Soulier syndrome and prekallikrein deficiency

TO THE EDITOR: Bernard-Soulier Syndrome (BSS) and prekallikrein (PK) deficiency are two rare genetic disorders with autosomal recessive transmission patterns, and the combined occurrence of these two disorders is rare [1, 2]. In BSS, abnormalities are caused by glycoprotein (GP) Ib/IX/V complex defects, which constitute the Von Willebrand factor (VWF) receptor on the platelet surface [1, 3, 4]. This condition is clinically characterized by thrombocytopenia and prolonged bleeding time (BT) [5]. Common symptoms include easy bruising and gum and nose bleeding

episodes [6].

Coagulation factor XII (FXII), PK, and high-molecular weight kinogen (HMWK) are the three important plasma proteins of the kallikrein-kinin system. Deficiencies in any of these factors are rare and diagnosed when the results of routine coagulation tests show a prolonged activated partial thromboplastin time (aPTT) [2]. Most cases of PK deficiency are asymptomatic. However, there had been a few reports on the association between severe PK deficiency and thrombotic phenomena and recurrent pregnancy losses [7].

In this report, we describe the first combined occurrence of BSS and PK deficiency in a 3-year-old girl who presented with recurring epistaxis.

A 3-year-old girl, who was born out of consanguineous marriage through caesarean section with a birth weight of 2,650 g and gestational age of 38 weeks, was admitted to our hospital with a history of recurrent nose bleeding and body bruising within a one-year duration. She was hospitalized numerous times due to similar complaints. No significant history of trauma, jaundice, fever, or previous blood transfusions was recorded. The mother had a history of anemia and thrombocytopenia during pregnancy with a platelet count of 90,000/ μ L and did not have any known disease before pregnancy. In addition, she had no history of medication use. Her past medical history was negative for any thromboembolic phenomena. No family history of similar illness was obtained.

The results of the patient's ENT (ear, nose, and throat) examination were not significant. She had active nose bleeding, and petechiae, purpura, and ecchymosis were observed in the limbs. Her systemic examination was essentially normal with no organomegaly.

Blood test results revealed a hemoglobin level of 12.2 g/dL and white blood cell count of 5,600/ μ L, with normal differential counts. Her platelet count was 37×10^3 / μ L and platelet morphology showed numerous giant platelets. Furthermore, the following laboratory tests were also obtained: BT (16 sec; control: 3-7 sec), prothrombin time (13.1 sec; control: 13.2 sec), aPTT (>180 sec; control: 34.1 sec), mixed PTT (35.3 sec), VWF (Ag=87%; VWF Ab=73%). The aggregating agent results were the following: ADP: 2 μ mol/L; (63; normal, 50-150); ristocetin: 1.5 mg/mL (0; normal, 50-150); collagen: 2 μ mol/L; (69; normal, 50-150); and arachidonic acid: 0.5 mmol/L (53; normal, 50-150). The result of the platelet aggregation test confirmed the diagnosis of BSS, a condition wherein platelets do not aggregate in response to ristocetin and is characterized by thrombocytopenia and giant platelets.

Factor VIII level was 62 mg/dL (normal, 60-150 mg/dL), factor IX level was 62 mg/dL (normal, 60-150 mg/dL), factor XI level was 72 mg/dL (normal, 50-110 mg/dL), and factor XII was 69 mg/dL (normal, 50-120 mg/dL). In addition, the absolute fibrinogen level was 2.4 mg/dL (normal, 1.5-4.5 mg/dL). The pre-incubation of plasma with the surface activator, kaolin, caused a rapid shortening of the abnormal