Sclerosing Extramedullary Hematopoietic Tumor
Sklerozan Ekstramedüller Hematopoetik Tümör

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Dear Editor;

Sclerosing extramedullary hematopoietic tumor (SEMHT) is a very rare disease. It is first described by Remstein et al. [1] in 2000 and few cases were reported since then. It is associated with chronic myeloproliferative disorders. We describe a patient of SEMHT with bilateral renal involvement.

A 72-year-old man was admitted to the emergency clinic with dyspnea and abdominal pain. Physical examination revealed decreased breath sound over the right and left lower lung areas and severe edema in both lower extremities. He was clinically overhydrated. The rest of the physical examination was normal. Routine laboratory tests showed the following: serum levels of hemoglobin: 9.8 g/dL, white blood cell count: 31x10^9/L, platelet count: 100x10^9/L, blood urea nitrogen: 36mg/dL, serum creatinine: 1.9 mg/dL, serum albumin: 2.2g/dL. The peripheral blood smear was normal. Bone marrow biopsy was not performed. Abdominal ultrasound showed perirenal hypoechoic mass compressing bilateral kidneys (Fig. 1a). Magnetic resonance imaging revealed renal capsular heterogenous involvement with maximum thickness of 30mm (Fig. 1b). Renal tru-cut biopsies were performed from both kidneys. Microscopically both lesions showed myxoid and sclerotic stroma intermixed with large atypical cells (Fig. 1c,1d). Immunohistochemistry displayed positivity for factor 8 (Fig. 1e) and CD41 in atypical cells. (Fig. 1f) Scattered mature myeloid cells
were positive for myeloperoxidase. CD34, CD117, S100, CD3, CD30, CD20, glycoporphin, MDM2, keratin, EMA, desmin, myogenin were all negative. The presence of CD41 positive atypical megakaryocytes within the tumor suggested the diagnosis of SEMHT. His previous history of primary myelofibrosis and splenectomy was learned after the histologic diagnosis of the renal tumor.

SEMHT is an uncommon lesion formerly known as fibrous hematopoietic tumor or myelosclerosis [1]. It is associated with chronic myeloproliferative disorders mainly chronic idiopathic myelofibrosis in older age group. SEMHT has a predilection for mesentery and reproperitoneum. However, tumors involving skin, liver, kidney and lacrimal glands were described as single reports [1-3]. SEMHT usually presented as multiple nodules with varying size. Our case showed diffuse infiltrative nature of the tumor compressing the bilateral kidneys. Bilateral renal involvement is an unreported radiologic finding. Microscopically these tumors were characterized by myxoid to sclerotic stroma with thick collagen bundles, intermixed with large atypical megakaryocytes. Occasional foci of mature hematopoetic cells were encountered [1,2]. It is believed that sclerosis within the tumor was produced by fibroblasts, which is induced by cytokines released from clonal megakaryocytes. The presence of JAK2 V617F mutation may also suggest the clonal nature of the lesion [4].

The differential diagnosis includes sclerosing liposarcoma, malignant fibrous histiocytoma/pleomorphic sarcoma, sarcomatoid/anaplastic carcinoma and Hodgkin lymphoma [1,3]. The presence of dysplastic megakaryocytes with “ink blot-like” nuclei and eosinophilic cytoplasm, is in favor of SEMHT. Factor-8, CD41 and CD61 are helpful markers for the confirmation of the diagnosis. Pathologist should keep in mind this rare entity for the differential diagnosis of tumors with anaplastic morphology. High cellular pleomorphism may lead to inaccurate diagnosis of sarcoma or carcinoma and a subsequent unnecessary surgery.

REFERENCES

FIGURE LEGEND:
Figure 1: (a) Abdominal ultrasonographic image and (b) magnetic resonance image showing renal capsular heterogenous mass. (c) Photomicrograph showing large atypical megakaryocytes in a myxoid to collagenous background (H&E, x100). (d) Megakaryocytes at high power view (H&E, x400) (e) showing factor 8 positivity and (x400) (f) CD41 positivity (x400).