RENAL CELL CARCINOMA IN A 22 YEAR OLD PATIENT WITH SINUS HISTIOCYTOSIS

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Sinus histiocytosis (SH) or Rosai-Dorfman disease is a rare idiopathic entity characterized by massive lymphadenopathy (LAD). Extranodal disease occurs in up to 43% of the cases and can involve the kidneys. SH is usually benign with resolution spontaneously or after low dose steroids. An association with different types of malignancy has been described but none with renal cell carcinoma (RCC). We describe a case of SH associated with a renal cell carcinoma.

A 22-year-old female previously healthy developed a nephrotic syndrome and was diagnosed with primary FSGS. Few months later, she developed bilateral inguinal lymphadenopathy and a CT scan of the abdomen and pelvis revealed a 2.8 cm left renal mass and prominent mesenteric lymph nodes. Except for recent fatigue, she denied other constitutional symptoms. Her exam was remarkable for bilateral inguinal nodes and pitting edema in her lower extremities. She did not have any family history of renal disease. Laboratory work up was unrevealing. T & B cell flow cytometry did not show any monoclonal cell population to suggest a lymphoma. Excision of a right inguinal lymph node revealed follicular lymphoid hyperplasia and sinus histiocytosis. Because SH can presents as a kidney mass no further investigation was planned. Eventually, a fine needle aspiration of the kidney mass was done and suggested a low-grade chromophobe cell carcinoma. A left partial nephrectomy was performed and pathology confirmed the RCC (chromophobe cell type) confined to the kidney. The patient remained free of recurrence 3 years later and her LAD resolved spontaneously.

This case illustrates that a renal mass should be fully investigated even in a young patient despite the presence of sinus histiocytosis, a usually benign condition that was presumed to be causing the kidney mass.