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Oncology

Giant cystic lymphangioma of adrenal gland: A case report and review of the literature

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Introduction

Cystic Lymphangiomas are benign malformations of lymphatic vessels. Location of this pathology can be diverse but most commonly occurs in the neck or axilla. Less than 60 cases of adrenal lymphangiomas have been reported in the literature. We present the case of a 74 -year-old woman who presented to the hospital with a 10 cm cystic lesion located in the left adrenal gland. Left adrenalectomy was performed. Histopathological examination analysis of the cystic lesion was consistent with a lymphangioma. Diagnostic features of adrenal cysts including lymphangiomas are discussed.

Case

A 78-year-old woman with a past medical history of systemic hypertensive disease for 5 years presented to the hospital with constant lower back pain during the last year. On examination, the patient showed no abnormality. A computed tomography (CT) scan revealed 12*10*9 cm hypodense non-enhancing lesion on the upper pole o f the left kidney, which was suspected to be an adrenal cyst(Fig. 1A and B). The patient with adrenal masses were evaluated and screened for hyperaldosteronism, pheochromocytoma, and hypercortiolism. Laboratory data showed no appreciable abnormality of biochemical test. Due to the size and position of the cyst, and to avoid a rupture of the cyst during laparoscopy, a left subcostal laparotomy was performed. During surgery, the adrenal origin of the cyst was confirmed, and a left adrenalectomy was performed without rupture of the cyst. The postoperative course was uneventful. On pathologic examination the adrenalectomy specimen measured 6 × 2.5 × 8.5 cm. The cyst was filled with clear, non viscous, brown colored fluid. Microscopic examination revealed a multi-cystic lesion laid with flat endothelial cells (Fig. 2A). Immunohistochemical examination revealed an endothelial lining positive for factor VIII-related antigen (Fig. 2B) and CD34 (Fig. 2C). No significant endothelial atypia was noted. The diagnosis of adrenal lymphangioma was made. One year after surgery, the patient remains free of symptoms.

Discussion

Lymphangiomas are benign malformations of vessels. They are most commonly located in the neck, axillary region and mediastinum (95%). The remaining 5% are found in the abdominal cavity.[1] They have extremely rarely been documented in the adrenal gland, with less than 60 cases reported in literature and account for < 1% of all abdominal lymphangiomas.[1] Adrenal cysts have been classified into four main groups: endothelial cysts (45%), pseudocysts (39%), epithelial (9%) and parasitic cysts (7%).[2] Endothelial cysts include hemangiomas, hamartomas, and lymphangiomas.[2] The exact pathogenesis of adrenal lymphangioma is not completely resolved. Some believe that these lesions stem from continued growth of ectopic or malformed lymphatic tissues or represent hyperplasic reaction to inflammation or blockage of draining lymphatics.[2] Adrenal lymphangiomas don't have a

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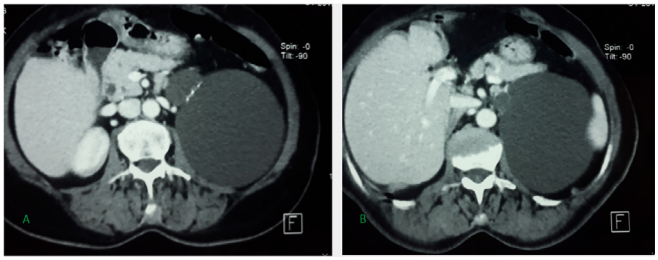

Fig. 1. A and 1B: Contrast enhanced CT scan in the axial plane showing a 12*10*9 cm hypodense non-enhancing lesion with septa on the upper pole o f the left kidney.

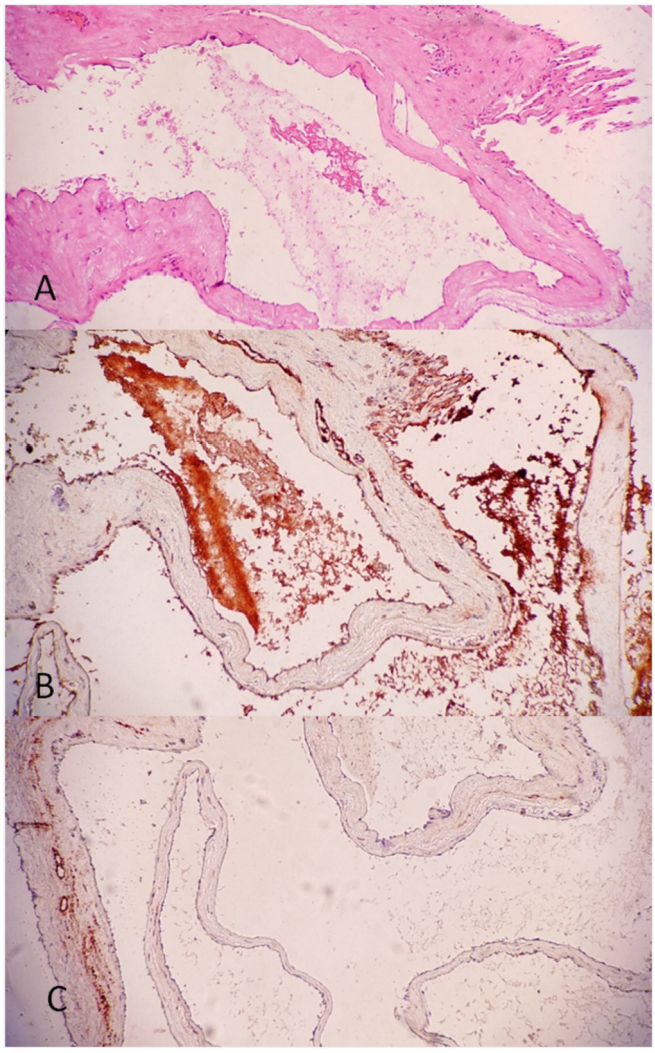

Fig. 2. A, 2B and 2C: Histological examination (HE X 20) showing a multicystic lesion laid with flat endothelial cells (A) and Immunohistochemical examination showing the endothelial lining stains positive for factor VIII-related antigen (B) and CD 34(C).

pathognomonic radiological presentation. Ultrasound often demonstrates a well-marginated lesion. An adrenal lymphangioma should be suspected on CT when a unilocular or multilocular, low-attenuation, non enhancing lesion is present in the region of the adrenal gland, particularly if the lesion contains either of the above-noted calcification patterns.[3] On MRI, most are low in signal intensity on T1-weighted images and high on T2-weighted images.[3]

Certain features raise the suspicion of malignancy within a cystic adrenal lesion, including a heterogeneous appearance on imaging and the presence of necrosis in the center of the mass accompanied by calcification, and the size of the adrenal mass [12,13]. Adrenal tumors > 5 cm on cross-sectional imaging are more likely to be malignant, and the risk of malignancy in adrenal pseudocysts > 5 cm is approximately 7%, rising with increased size.[3] On pathological examination adrenal lymphangiomas are multiloculated, thin-walled cystic lesions, filled with non viscous, clear, yellow-brown colored fluid. Microscopy demonstrates that the irregular dilated cystic channels spaces contain proteinaceous material and usually lack red blood cells. Immunohistochemical examination has to be made to prove the lymphatic origin of the cyst. The endothelial lining stains positive for factor VIII-related antigen, CD31 and CD34, while it demonstrates a lack of staining for cytokeratin confirm the lymphatic rather than the mesothelial nature of the lining. D2-40 which is a monoclonal antibody to the transmembrane mucoprotein is a more specific marker of lymphatic lineage. Some authors recommend aspiration of the contents of adrenal cysts both for diagnosis and management instead of surgical excision, if the suspicion of malignancy is low, or the lesion is non-functional and asymptomatic.[4] This method, though, is characterized by the high reaccumulation of the cyst fluid and probably the dispersion of malignant cells in the peritoneal cavity, while its ability to determine the histology of cyst is limited.[4] Surgery should be the appropriate treatment for large adrenal lymphangioma. Complete resection of the cyst is recommended, and associated adrenalectomy depends on the location of the cyst and on the intraoperative dissection.[5] Decision to undertake a laparotomy or a laparoscopy depends on the position, the size, and the risk of rupture of the cyst.

Conclusion

Adrenal lymphangioma is a rare pathology that should be included in the differential diagnosis of cystic lesions of the adrenal glands. Its diagnosis can be difficult and challenging. The diagnosis is established after surgery by pathological report. Surgery is the appropriate treatment of large symptomatic cyst.

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