Case Report

Adrenal Lymphangiomatous CYST with papillary and pseudopapillary endothelial proliferation and sinaptophysin positivity: A Case Report

Papiller ve psödopapiller endotelyal proliferasyon ve sinaptofizin pozitifliği gösteren adrenal lenfanjiyomatöz kist: Olgu Sunumu

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ABSTRACT

Benign adrenal cysts are rare lesions. Although they can cause some clinical symptoms such as abdominal distension and pain, they are mostly encountered incidentally. The most common are pseudocysts, endothelial, epithelial, and parasitic cysts. Endothelial cysts may be of vascular or lymphatic origin. In this case report, a unilocular lymphangiamotous adrenal cyst showing marked papillary and micropapillary proliferation and synaptophysin positivity is discussed in the light of current literature.

Keywords: Adrenal cyst, endothelial proliferation, lymphangiomatous cyst, synaptophysin

ÖZ

Benign adrenal kistler nadir görülen lezyonlardır. Karında şişkinlik, ağrı gibi klinik belirtilere neden olabilse de çoğunlukla insidental olarak karşılaşılmaktadır. Psödokistler, endotelyal kistler, epitelyal kistler ve parazitik kistler en sık görülenlerdir. Endotelyal kistler vasküler veya lenfatik kökenli olabilir. Bu olgu sunumunda belirgin papiller ve mikropapiller proliferasyon ve sinaptofizin pozitifliği gösteren uniloküler lenfanjiamotöz adrenal kist güncel literatür ışığında tartışılmıştır.

Anahtar Kelimeler: Adrenal kist, endotel proliferasyonu, lenfanjiomatöz kist, sinaptofizin

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A 20-year-old female patient was admitted to the hospital due to abdominal pain. Abdominal computed tomography revealed a unilocular cyst with a diameter of 10.5 cm in the right adrenal gland. The patient was operated by open excision.

In the macroscopic examination of the mass, a unilocular cystic lesion with a diameter of 10.5 cm was observed adjacent to normal adrenal and fatty tissue (Figure 1).

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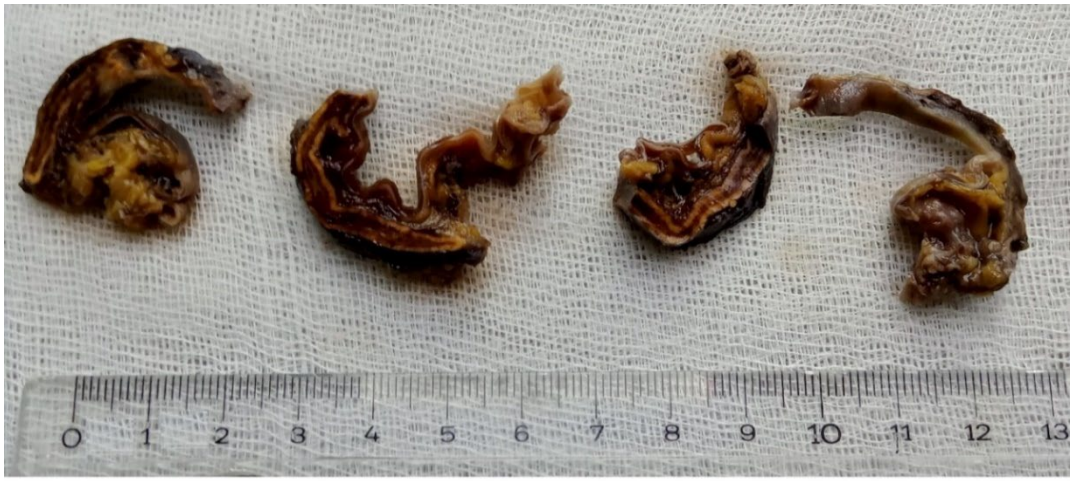

Figure 1. Macroscopic apperence of the cyst and adrenal tissue

Histological examination revealed a cystic lesion lined with endothelial cells next to the adrenal gland. Papillary and pseudopapillary proliferation was observed throughout the cystic lesion. The cells of the lining epithelium were generally columnar in shape, with an increased nucleus-cytoplasm ratio, and some consisted of hobnail-like cells. No mitotic activity, necrosis, or hyperchromasia was observed. Hyalinized connective tissue containing many dilated vessels was observed at the base of the papillary structures (Figure 2-3).

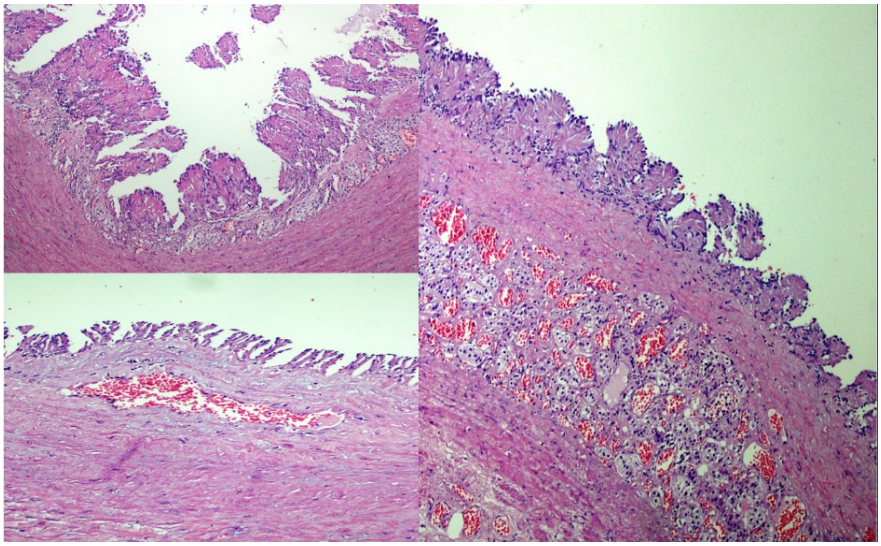

Figure 2. Papillary and micropapillary structure of the cysts H&E X100 and H&E X200.

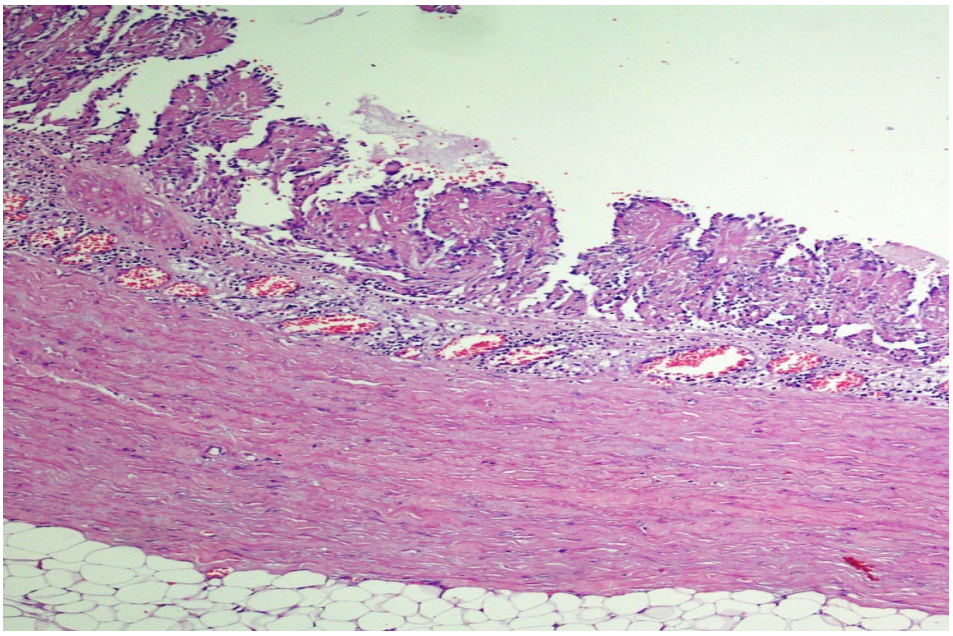

Figure 3. Papillary structure H&E X 400.

Immunohistochemical examinations were performed for the differential diagnosis of adrenal tumors and renal tumors. Negative staining was obtained with CAIX, CK7, PAX8, Melan-A, inhibin, CD10, RCC, HMB45, and chromogranin. CD 31, FLI-1, factor VIII, podoplanin, synaptophysin, and ERG were shown to have immunopositivity (Figure 4).

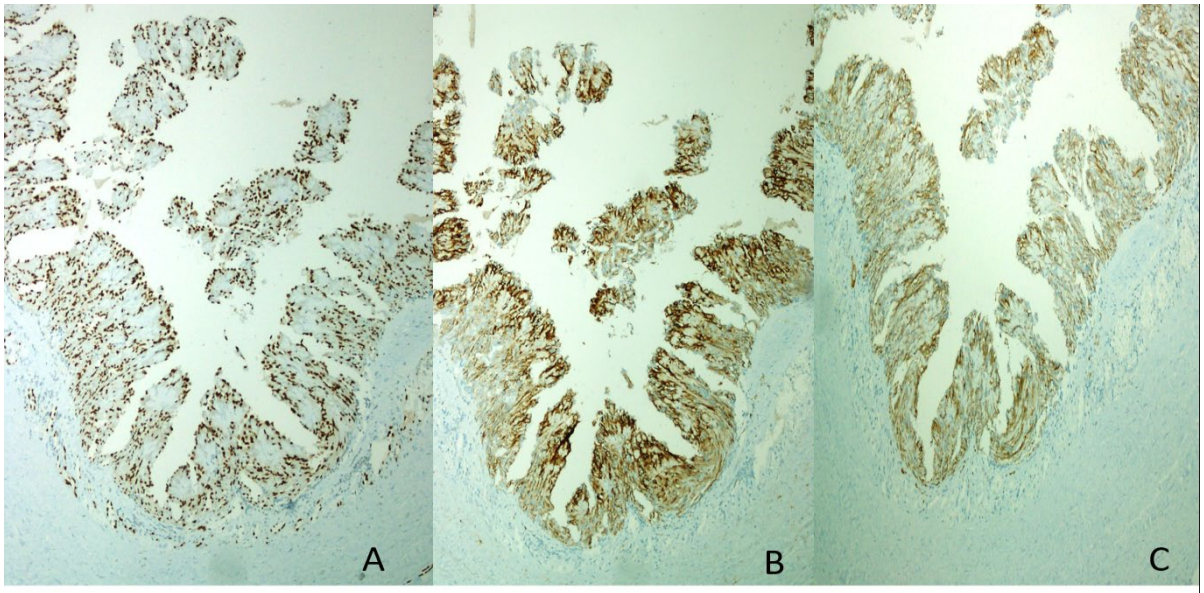

Figure 4. Immunohistochemistrial staining A: Podoplanin, B: ERG C: Synaptophysin.

Based on histological and immunohistochemical findings, the case was diagnosed with a lymphangiomatous cyst showing papillary proliferation.

DISCUSSION

Benign adrenal cysts are rare lesions. These cysts can vary in size and may be multiloculated or unilocular. Although there are differences in autopsy series and surgical studies, pseudocysts and endothelial cysts constitute the majority of these cysts (1). Pseudocysts are not lined by epithelium. Epithelial cysts are subdivided into glandular or retention cysts, cystic adenomas, and embryonal cysts. Hamartomatous, angiomatous, and lymphangiomatous cysts are subtypes of endothelial adrenal cysts.

Sometimes, benign adrenal cysts need to be surgically removed so that a definitive diagnosis can be made through pathological examination. Surgical indications for adrenal cysts are generally large size, complications, and suspicion of malignancy (2-5). Cystic metastases and cystic pheochromocytoma may be confused with benign adrenal cysts and pathological examination may be required (6).

The differential diagnosis of these lesions is based on histological and immunohistochemical features (7-10). Lymphatic cysts are usually lined with a flattened epithelium and contain serous fluid. Koperski et al. classified lymphatic cysts histologically as ‘typical multicystic lymphatic malformation’, ‘unilocular lymphangiomatous cyst’, and ‘lymphangiomatous cyst with papillary endothelial proliferation’. In this review, 4 cases showed diffuse papillary or pseudopapillary proliferation (9). All of these cases were unilocular cysts, similar to our case, and in this study, no radiological and clinical differences were observed, except that the cysts showing papillary proliferation were larger in size than other lymphatic cysts. Immunohistochemistry can be used to distinguish tumours with this histological appearance from tumours presenting with papillary structures. While lymphatic cysts are expected to stain positively with D2-40, CD31, and ERG, they stain negatively with pankeratin and CD34. However, although its importance is unknown, in our case, synaptophysin positivity was accompanied by the markers indicating lymphatic origin. Although synaptophysin positivity has been reported in vascular lesions, no satisfactory information has been found in the literature regarding synaptophysin positivity in lymphatic cysts (11).

CONCLUSIONS

Benign adrenal cysts can be operated if the lesions are large, cause complications, or to rule out malignancy. Papillary and pseudopapillary structures can be seen in cysts of lymphatic origin, and the differential diagnosis of these lesions can be made by pathological examination accompanied by macroscopic, microscopic, and immunohistochemical findings.

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