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Pathology

Adrenal cystic lymphangioma with radiological, clinical and histopathological findings: Case report

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ABSTRACT

Cystic lymphangiomas are benign lesions originating from lymphatic endothelial cells. It occurs due to developmental anomalies of lymphatic vessels. They are usually localized in the head and neck region. Cystic lymphangiomas of adrenal origin are very rare. This presentation aims to report a case of left-sided adrenal cystic lymphangioma detected incidentally on radiological examination due to abdominal pain, with clinical, radiological and pathological findings. A 65-year-old female patient was admitted to our clinic with abdominal pain. In the abdominal examination, the pain was detected in the epigastric region and left the upper quadrant with palpation. No pathology was observed in the complete blood count and biochemical parameters, except for a CRP elevation of 10.2 mg/dL. In examination with ultrasonography (US), in the left upper quadrant of the abdomen, in the localisation of the adrenal gland, a multilocular cystic lesion with partially dense contents, which is not vascularised by Doppler US, containing thin echogenic septa was detected. Enhanced contrast multidetector computed tomography was performed to determine the nature and characterisation of the mass. A 60x57 mm cystic lesion with multi-lobulated contour and fluid density was defined in the left adrenal gland. The patient was diagnosed with cystic lymphangioma radiologically and was operated upon due to symptoms and size. Pathological diagnosis was reported as cystic lymphangioma. Preoperative clinical and radiological correct diagnosis is critical because the treatment approach and prognosis may differ from other adrenal tumours or cysts.

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INTRODUCTION

Cystic lesions of adrenal origin are rare, and their incidence has been reported as 0.06% in the literature. Cystic lymphangiomas of adrenal origin, also known as adrenal lymphangiomas, are extremely rare tumours. Due to their rarity, specific incidence rates for adrenal lymphangiomas are not well-established in the medical literature.[1] Cystic adrenal gland lesions are classified into four main histopathological groups: endothelial cysts, pseudocysts, epithelial cysts and parasitic cysts. Endothelial-derived cysts are further subdivided into lymphatic and angiomatous cysts based on the histological origin of the endothelium. Lymphatic cysts are also called adrenal lymphangiomas and adrenal cyst of lymphangiomatous origin (ACLO) according to their single or multi-local status.1 Cystic lymphangiomas originate from lymphatic endothelial cells. It occurs due to the developmental abnormality of the lymphatic channels. They are usually localised in the head and neck but can develop in different body parts, including the adrenal glands. Although they may occur at any age, it has been reported that they are more common in decades 3-6, in females and on the right side.[2]

Adrenal gland cystic lymphangiomas are typically discovered incidentally during imaging studies such as ultrasonography (US), computed tomography scans, or magnetic resonance imaging performed for unrelated reasons or intra-abdominal operations.[3-5] An adrenal gland cystic lymphangioma is characterised by forming cystic spaces filled with lymphatic fluid. They are usually asymptomatic. In some cases, however, larger cystic lymphangiomas can cause symptoms such as abdominal pain, a palpable mass, or hormonal imbalances if they interfere with the normal functioning of the adrenal gland. There is limited information available about their specific characteristics and management. Treatment is surgical if necessary. When deciding on surgery for adrenal masses, their size, hormonal activity, imaging findings suggestive of malignancy, and growth rates on follow-up examinations should be considered.[6] This presentation aims to report a case of cystic lymphangioma of adrenal origin, which was detected incidentally in the radiological examination due to abdominal pain, with clinical, radiological and pathological findings.

CASE REPORT

A 65-year-old female patient was admitted to our clinic with a complaint of abdominal pain for three weeks. The abdominal examination detected pain in the epigastric region and the left upper quadrant with palpation. No pathology was observed in complete blood count and biochemical parameters, except for a 10.2 mg/dL increase in CRP. In the US examination of the entire abdomen, In the left upper quadrant of the abdomen, in the localisation of the adrenal gland, a multilocular cystic lesion with partially dense contents, which is not vascularised by Doppler US, containing thin echogenic septa was detected (Figures 1 and 2).

In intravenous contrast-enhanced multidetector computed tomography (MDCT) performed for the nature and characterisation of the mass, a 60 × 57 mm cystic lesion with multi-lobulated contour and fluid density was defined in the left adrenal gland (Figures 3 and 4). The patient was diagnosed with cystic lymphangioma radiologically and was operated upon due to its symptoms and size. A cystic lesion on the cross-sectional surface measuring 6 × 5 × 2 cm was observed in the macroscopic evaluation. Histopatho-

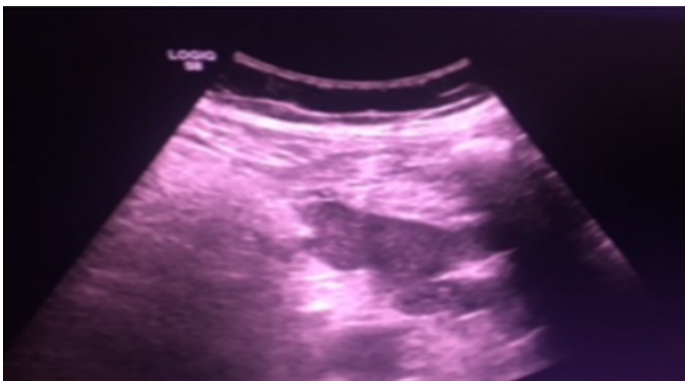

Figure 1. Multilobular, dense, cystic lesion with thin echogenic septa on abdominal ultrasonography examination.

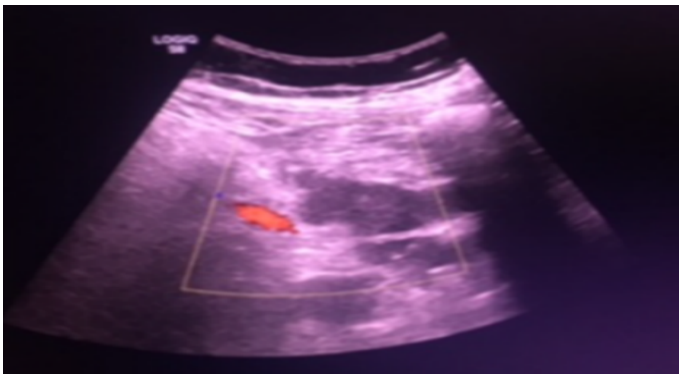

Figure 2. Non-vascularized, slightly dense, multiloculated cystic lesion on Doppler ultrasonography examination.

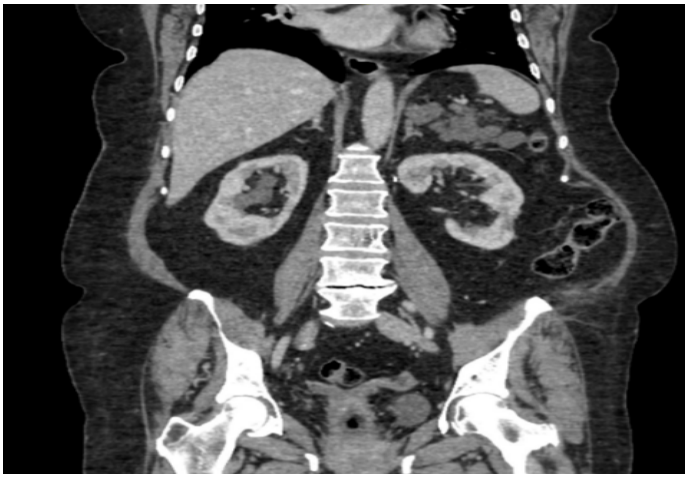

Figure 3. Coronal computed tomography sections: non-enhancing multilobulated cystic lesion in the left adrenal gland.

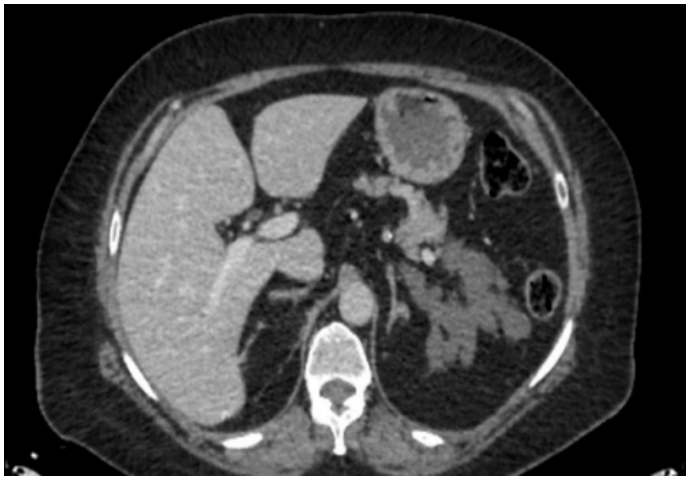

Figure 4. Axial computed tomography sections: multilobulated, non-enhancing cystic lesion in the left adrenal gland.

logical examination revealed that the lesion consisted of cystic structures lined with endothelium, and the case was reported as cystic lymphangioma (Figure 5).

Radiological imaging was performed with (GE brand Logic S7 Expert model) ultrasound and 64-section MDCT (Optima CT 660, General Electric Medical Systems, Milwaukee, Wisconsin, USA) devices. Water-soluble nonionic intravenous contrast agent (350 mgI/mL) was administered with an automatic double-injector system at a dose of 1 mL/kg at a rate of 4-5 mL/s through an 18-gauge cannula inserted into the antecubital vein. Intravenous contrast agent timing was performed with a bolus monitoring technique.

DISCUSSION

Adrenal lymphangioma typically presents as painless masses and may grow slowly over time. Symptoms, if present, can vary depending on the size and location of the cystic lymphangioma. They may include abdominal pain or discomfort, palpable mass in the abdomen, hormonal disturbances if the tumour affects the production of adrenal hormones, or compressive symptoms if it presses on nearby structures. Mostly, adrenal gland lymphangiomas may be discovered incidentally during imaging studies.[7] Extensive lesions may produce symptoms related to rupture or be detected incidentally.6 Our case was admitted to

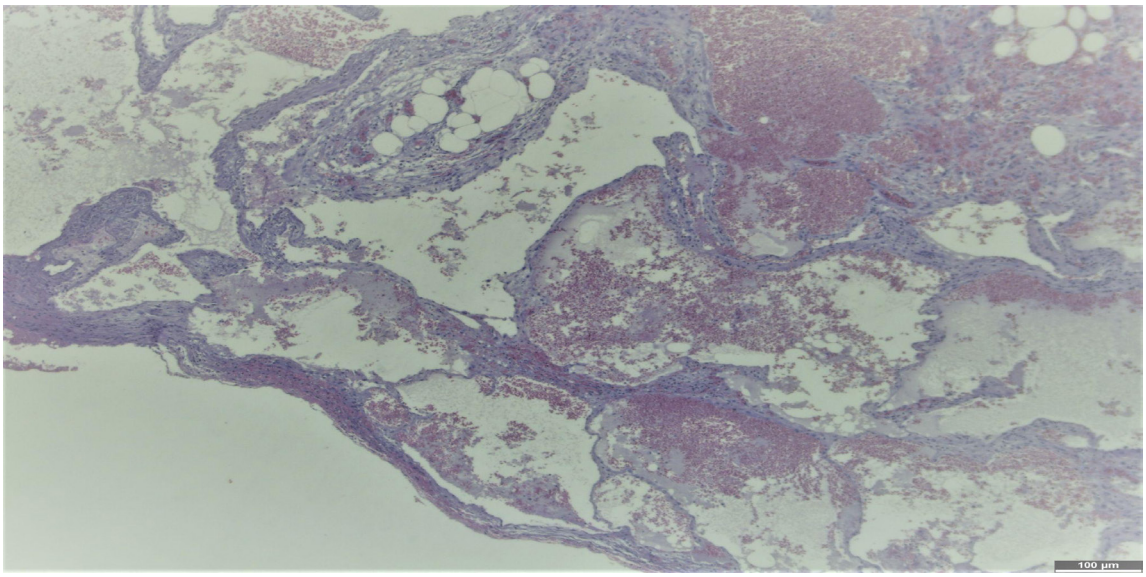

Figure 5. Cystic structures lined with endothelium (hematoxylin and eosin x40).

our clinic with abdominal pain for three weeks.

An adrenal gland cystic lymphangioma is usually diagnosed through imaging studies. These imaging techniques can help visualise the tumour and its characteristics, such as size, location, and cystic nature.[3-5] The technical developments in radiology and the availability of devices such as US, computed tomography, and magnetic resonance examination in many centres have increased the rate of diagnosing adrenal cystic lymphangioma cases. Radiologically, they appear as multilocular cystic lesions, which are approximately 3-6 cm in size, thin-walled, may contain septa, rarely show contrast enhancement in the walls and septa, and may contain millimetric calcifications at a low rate.[2,8] Although rare, it can grow rapidly and reach gigantic sizes.[9] During the abdominal examination performed with the US in our case, Doppler US revealed a multilocular cystic lesion with no vascularity, partially dense content, and thin echogenic septa in places.

Treatment options for adrenal gland lymphangiomas may include surgical removal of the tumour if it is causing symptoms or if there is a concern for malignancy. In some cases, a partial or complete removal of the affected adrenal gland may be necessary. However, a laparoscopic minimally invasive method can also be applied.1 There is no consensus on the surgical indication's mass size. Some authors recommended surgery if it is larger than 6 cm and surgery if it is smaller than 6 cm and has a risk of malignancy. Some authors have recommended surgery for masses larger than 3 cm.6,10 However, due to the rarity of these tumours, there is limited consensus on the optimal management approach, and decisions are often made on a case-by-case basis. Our patient was diagnosed with cystic lymphangioma radiologically and was operated upon due to its symptoms and size.

In the radiological differential diagnosis, adrenal cyst, adrenal hemangioma, retroperitoneal teratoma, cystic pheochromocytoma, cystic schwannoma, adrenal hydatid cyst and cystic metastasis can be considered.[11-13] However, the multilobular lesion, containing septa and a dense cystic appearance, suggests cystic lymphangioma. If contrast enhancement occurs in radiological images, it may be confused with adrenal carcinoma.[9] In our case, a 60 × 57 mm cystic lesion with multilobular contour and fluid density was observed in the left adrenal gland on IV contrast-enhanced MDCT.

Although there may be diagnostic difficulties in some cases before the operation, the pathological diagnosis of the surgically removed cyst is not difficult. Some of the radiologically diagnosed cysts are not included in the differential diagnosis because of their histologically distinctive histological features. In case of difficulty in diagnosis, differential diagnosis is made using an antibody panel such as CD31, CD34, pan cytokeratin AE-1/AE-3, D2-40, and factor VII from histopathologically similar cystic lesions.[2] Since there was no difficulty in the differential diagnosis in our case, an immunohistochemical examination was not performed.

CONCLUSIONS

It is important to consult a qualified healthcare professional, such as an endocrinologist or a surgeon specialising in adrenal gland disorders, for an accurate diagnosis and appropriate treatment plan.

Conflict of Interest

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Authors' Contribution

Study Conception: KG; Study Design: KG, ANK; Literature Review: ASA, AM; Critical Review: ANK; Data Collection and/or Processing: ASA, AM,; Analysis and/or Data Interpretation: ANK, AM; Manuscript preparing: ASA, AM, KG.

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