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Case Report

Inflammatory Myofibroblastic Tumor in the Adrenal Gland: A Case Report

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Myofibroblastic tumor · Adrenal gland · Inflammatory tumor

Abstract
Inflammatory myofibroblastic tumors (IMTs) were first described by Harold Brunn in 1939. IMTs are mainly found in the lungs and other sites of the body; hence, its occurrence in the adrenal gland is exceptional. In the literature, less than 10 cases of IMTs in the adrenal gland have been reported. The etiology of IMT remains unknown, with post-inflammatory changes and a neoplastic origin being proposed.

We present a case of a 19-year-old woman and adrenal gland IMT. The patient presented with abdominal pain and low cardiac output without hypovolemic shock. Computed tomography revealed a tumor in the adrenal gland measuring 11.4 cm with extravasation of contrast medium within the tumor. Treatment included conservative management with selective embolization due to minimal invasion of the inferior artery of the adrenal gland. The patient was then discharged with possibility of future elective surgery. Four months later, the size of the tumor decreased to 6.3 cm, and her Eastern Cooperative Oncology Group physical status was 0. The Multidisciplinary Tumor Board suggested surgical management. The final histopathology report was compatible with an IMT of the adrenal gland, with the immunohistochemical report showing positivity for anti-actin muscle-specific and anti-actin smooth muscle and negativity for anaplastic lymphoma kinase. IMTs of the adrenal gland may be treated electively through multidisciplinary management together with interventional radiology and surgery, achieving a favorable outcome for the patient.

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Introduction

Inflammatory myofibroblastic tumors (IMTs) are rare and infrequent. IMTs were first described by Harold Brunn in 1939 [1]. IMTs mainly occur in the lungs, but it can also be found in the gastrointestinal and genitourinary tracts; hence, its presentation in the adrenal gland is unusual. In the literature, only 8 cases of adrenal gland IMTs have been reported. The etiology of IMTs remains unknown, but there are two theories: the post-inflammatory theory, wherein tumors appear after trauma or surgery, and the neoplastic origin theory, wherein clonal alterations of up to 50% are observed in chromosome 2p23 [2]. Recently, ROS1 and PDGFRB have been implicated in the pathogenesis of IMTs. IMTs are composed of myofibroblastic and fibroblastic spindle cells [2], and they express actin, desmin, and keratin; anaplastic lymphoma kinase (ALK)-negative IMTs have a good prognosis. IMTs usually are usually 1–20 cm, with an average size of 6 cm [3, 4]. IMTs tend to be asymptomatic, but they may present with constitutional symptoms in 10–20% of cases. Pyrexia and weight loss are the primary manifestations, as well as febrile symptoms of unknown origin, requiring cabinet studies where it is found as an incidentaloma [4]. IMTs have an indeterminate biological behavior, and definitive treatment is total resection. Distant metastases are rare but mainly occur in the lungs and brain followed by liver and bone. If complete resection is not performed, 10–25% of cases will have local recurrence, and <5% will have distant metastasis, which may occur between 1 and 9 years after initial presentation [5]. IMTs of the adrenal gland are usually discovered incidentally, but it may also be accompanied by fever of unknown origin and abdominal pain.

Herein, we present a case of IMT in the adrenal gland that was treated by surgery. The CARE Checklist for this case report, which is attached as online supplementary material (for all online suppl. material, see https://doi.org/10.1159/000534129).

Case Report

A 19-year-old woman with a history of atopic dermatitis presented with a 3-month history of pain in the right dorsal region. She had no history of trauma or surgery. One week prior to presentation, pain was described as incapacitating, which prompted her to seek consultation with a doctor. On the visual analog scale, pain was scored 7/10 and had no alleviating or aggravating factors. Ultrasound was performed and revealed a retroperitoneal tumor. She was prescribed with paracetamol/tramadol 37.5 mg/325 mg for pain. As pain did not subside, she then visited our clinic. Upon admission, she was in severe pain, tachycardic, and diaphoretic. Her mean arterial pressure was within normal range. Physical examination revealed an Eastern Corporate Oncology Group physical status of 1. She was also dehydrated, had pale skin and integuments, and had a palpable tumor at the right hypochondrium. There was tenderness on palpation at the right hemiabdomen and a positive Giordano's sign. Blood examination revealed a hemoglobin of 6.5. Hence, blood transfusion was performed. As her vital signs stabilized after transfusion with two packs of erythrocytes, computed tomography (CT) of the abdomen and pelvis (Fig. 1) was performed, revealing loss of morphology of the right adrenal gland due to an ovoid lesion measuring 11.4 × 10.2 × 10.3 cm. The lesion had a volume of 626 mL and exerted a volume effect, anteriorly displacing segments VI and VII of the liver, partially collapsing the inferior vena cava, and anteriorly and caudally displacing the right kidney. There was evidence of active bleeding in its anterior wall, which caused hemoperitoneum. Based on these findings, a retroperitoneal hematoma and probable adrenal tumor (angiomyolipoma) were suspected.

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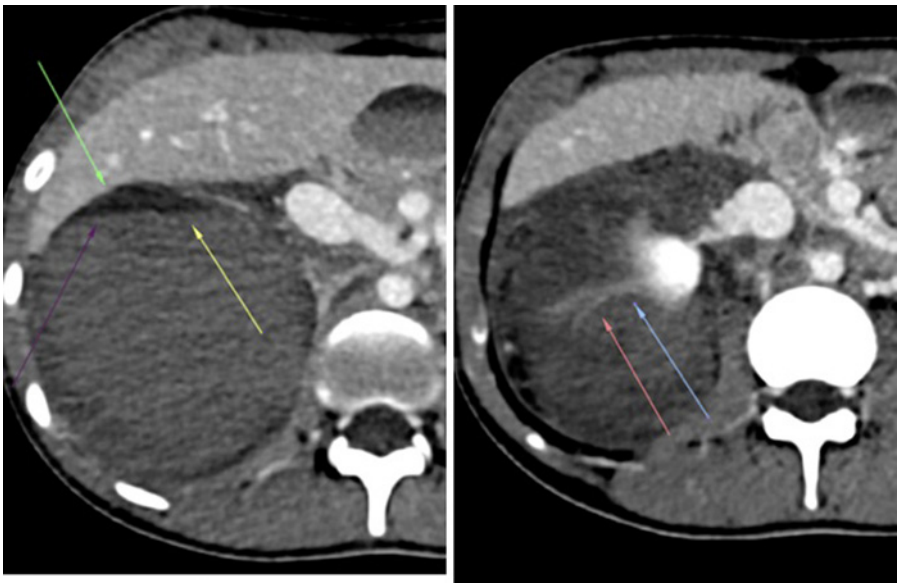

Fig. 1. Axial computed tomography images without (left) and with (right) contrast, with extravasation of contrast causing hematoma.

Due to the origin of the bleeding, instead of performing laparotomy, it was decided to perform interventional radiology with angiography of the right renal artery, angiography and embolization of the right inferior adrenal artery, and angiography of the right upper and middle adrenal arteries, resulting in the discovery of bleeding from the right inferior adrenal artery and an anatomical variant of the superior polar renal artery (Fig. 2). After the patient's status improved, she was the discharged with instructions to follow-up to monitor hematoma formation and to perform elective surgery on the adrenal gland.

On follow-up, the patient remained asymptomatic, with vital signs within normal range. Blood examination revealed a cortisol level of 15.2 μg/dL (normal: 5–25 μg/dL), urine metanephrine level of 338 μg/24 h (normal: <400 mg/24 h), and vanillylmandelic acid level of 7.6 mg (normal: <8 mg), suggesting a nonfunctional tumor. Four months after angioembolization, CT scan of the thorax, abdomen, and pelvis revealed a delimited tumor in the right adrenal gland measuring 6.3 × 6.1 × 5.5 cm, with a volume of 110 mL, and without evidence of active hemorrhage. The tumor still caused anterior displacement of segment V toward the medial anterior to the inferior vena cava and caudal displacement of the right kidney through its upper pole (Fig. 3).

An evaluation was carried out by the Multidisciplinary Tumor Board, and after considering differential diagnoses, surgery through open right supra-adrenalectomy was considered. Surgery was uneventful, and a lipomatous lesion measuring 6 × 8 × 10 cm encapsulated in the right adrenal gland within an area of hematoma and necrosis was excised. The right kidney was spared and preserved.

On macroscopic examination, the tumor measured 10 cm at its maximum diameter, weighed 109 g, was soft and depressible, oval in shape, with a dull surface, and heterogeneous in color with some dark brown and light gray areas (Fig. 4a, b). Microscopic examination revealed a neoplasm composed of bundles of randomly arranged plump spindle cells (Fig. 4c). On immunohistochemical examination, tumor cells were positive for anti-actin specific muscle (AAME) and anti-actin smooth muscle (AAML) and were negative for anaplastic lymphoma kinase-1 (Fig. 4d). Molecular tests were not performed because they were not available in our hospital. On follow-up 8 months postoperatively, the patient remained asymptomatic with no evidence of hypotension of recurrence.

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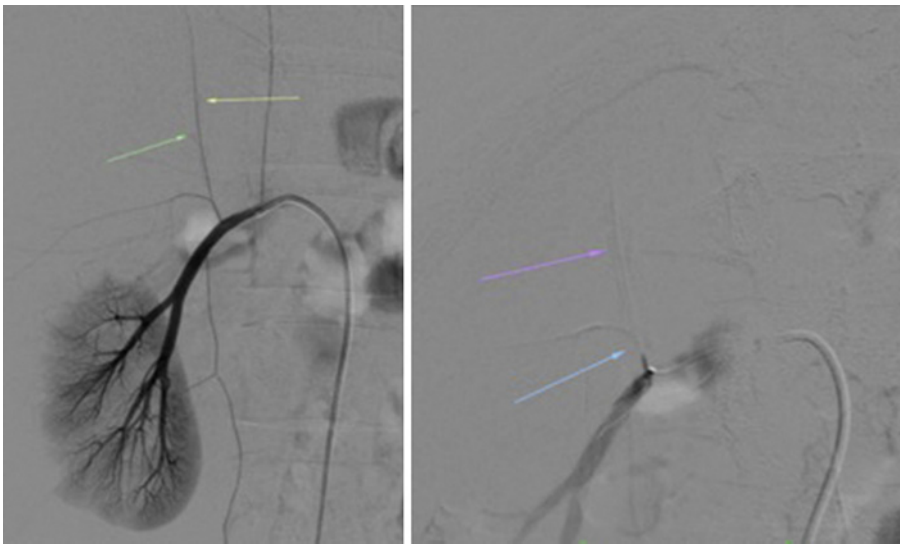

Fig. 2. Green and yellow arrows: extravasation of contrast medium in the distal portion of the right inferior adrenal artery. Purple and blue arrows: angiography and embolization of the right inferior adrenal artery.

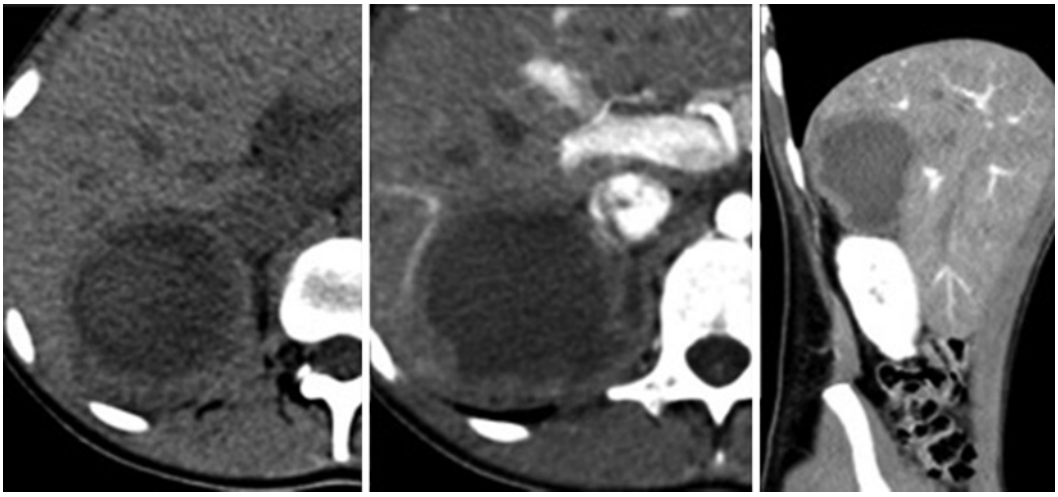

Fig. 3. Tumor encapsulated in the right adrenal gland with a decrease in size.

Discussion

IMTs in the adrenal gland are rare. Previously, IMTs were considered inflammatory tumors; however, as IMTs have different histological characteristics from those of myofibroblasts or proliferating fibroblasts with inflammatory infiltration, they were considered as being different from inflammatory tumors. Herein, we report a case of a patient with an IMT in the adrenal gland who presented with low cardiac output. Our patient's presentation was unusual in that the typical manifestation of adrenal gland IMTs is pain without hypotension, which may make our case the first such presentation in the literature. In our patient, the tumor had an idiopathic origin as we were unable to rule out a molecular origin due to lack of

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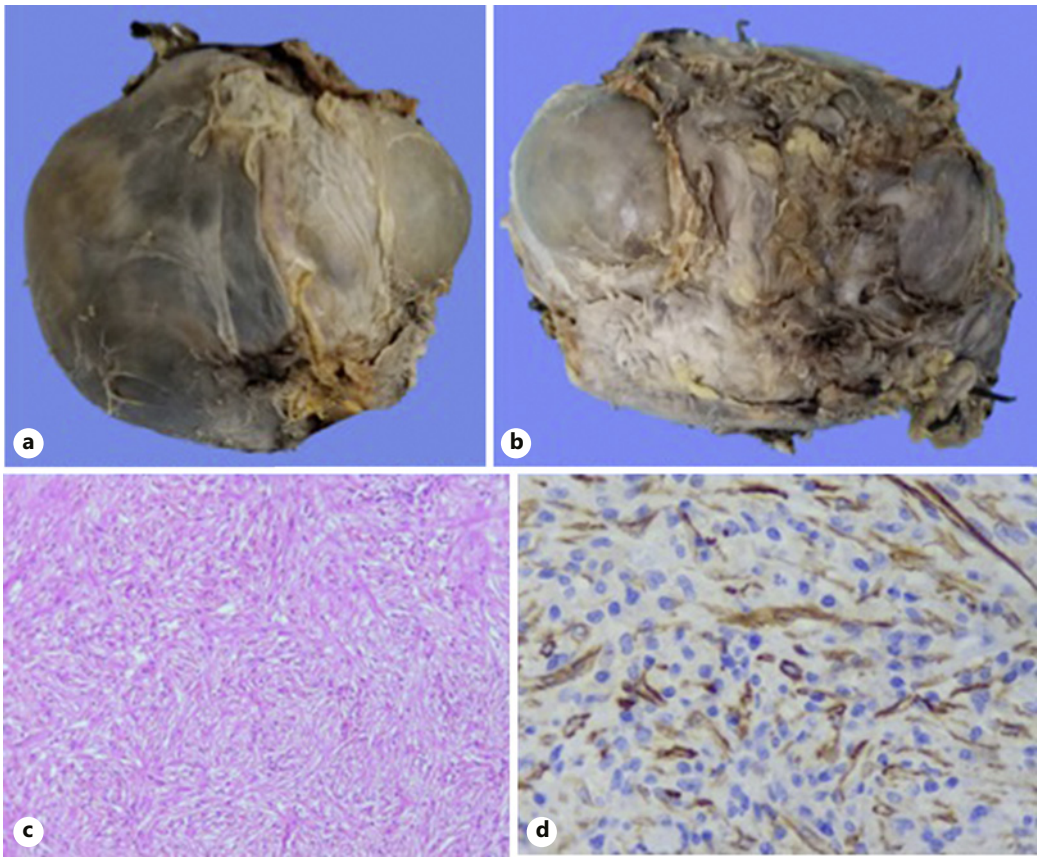

Fig. 4. a, **b** Macroscopic view of the tumor. **a** Outer surface. **b** Cut surface. **c** Low-power view of the tumor showing randomly arranged bundles of plump spindle cells. **d** Smooth muscle actin positivity.

resources to carry out the required tests. Despite having anemia and active bleeding, the hematoma was contained. Therefore, interventional radiology was considered before performing adrenal gland resection and possibly the loss of the kidney due to angioembolization. Once the patient was stable, proper evaluation and assessment of the adrenal tumor was performed, resulting in an appropriate pre-surgical protocol that involved selective surgery of the adrenal gland. Based on laboratory results, a functional tumor was ruled out. Metastases were also ruled out by imaging, and a pheochromocytoma was unlikely because of her stable vital signs and hypotension. Meanwhile, an adenoma was ruled out due to the patient's age. Hence, the primary consideration was a nonfunctioning adrenal carcinoma as well as a benign tumor such as an IMT; however, IHC was required to confirm the diagnosis. Due to the unavailability of IHC in our institution, it was decided to perform surgery instead. Eventually, surgical intervention with complete resection of the tumor was performed to reduce the risk of recurrence; this was achieved as the approach was electively selected, and the tumor was evaluated adequately. Notably, a negative ALK results in a better prognosis, in accordance with what is mentioned in the literature. Due to the location of the tumor, its rarity, sparse bibliographic evidence, and clinical manifestations, diagnosing this tumor is complicated, with histopathology playing an important role. Selective surgery of the adrenal gland should be considered in this case. The strength of this case is its clinical presentation and unusual treatment.

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The authors are grateful to the patient for participating in this case report.

Statement of Ethics

Written informed consent was obtained from the patient for publication of this case report and any accompanying image. Ethical approval was not required for this study in accordance with local or national guidelines.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Reiko M. Rodriguez-Nakamura contributed to the design of the report and drafted the manuscript. Martin Sanchez-Garcia contributed to the design of the report. Leonora Chavez Mercado contributed at the pathology revision. Arturo Del-Valle Rivera, Elizabeth Mota-Garduño, Sergio Adolfo Espinoza Guibarra, and Omar Vazquez-Gomez critically revised the manuscript.

Data Availability Statement

All data generated or analyzed during this study are included in this article and its online supplementary material. Further inquiries can be directed to the corresponding author.

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