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Case report

Rhabdoid collecting duct carcinoma with lymphangitic carcinomatosis causing acute lethal chylopericardium

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ABSTRACT

Collecting duct carcinoma is a rare neoplasm of the kidney, accounting for only 1–2% of renal tumors. These tumors arise from the principal cells of the renal collecting ducts of Bellini. The majority of patients have lymph node involvement and metastases to lungs, liver, bone, adrenal glands, and brain. We present a case of a 48 year old woman who came to the hospital with a clinical presentation suspicious for pneumonia. One week later her symptoms aggravated. A CT chest and abdominal imaging showed bilateral pulmonary infiltrates, retroperitoneal lymphadenopathy, and left hydroureteronephrosis. She expired after developing acute respiratory failure. An autopsy was performed which revealed chylopericadium of 150 cc; bilateral reticular pattern on the surfaces of the lungs; neck, mediastinal and retroperitoneal lymphadenopathy, and a 5.1 cm left kidney mass located in the mid portion medulla. The kidney tumor was a rhabdoid collecting duct carcinoma. The lungs showed diffuse subpleural lymphangitic spread of the carcinoma. We report a rare case of chylopericardium due to lymphangitic carcinomatosis from a 5.1 cm rhabdoid collecting duct carcinoma not suspected clinically or radiologically. This case highlights the importance of performing autopsies in an era when clinicians heavily rely on high-tech imaging diagnostic modalities.

1. Introduction

Malignant kidney tumors account for 4.1% of cancer incidence and 2.4% mortality in the United States, and studies show an increased incidence worldwide [1]. Collecting duct carcinoma (CDC) is defined as a malignant epithelial tumor arising from the principal cells of distal segment of the collecting ducts of Bellini in the renal medulla [2]. It was first recognized by Fleming and Lewi [3] in 1986 as a distinct pathologic subtype of Renal Cell Carcinoma (RCC). CDC is a rare tumor, accounting for only 1%–2% of renal tumors. Clinically, it is characterized as an extremely aggressive type of RCC with an advanced stage at presentation and thus has a very poor prognosis [1,4]. This tumor is more common in men, has a predilection for the right kidney, and a median patient age of 55 years (range, 43 to 63 y) [1,5]. Common clinical symptoms of this tumor include gross hematuria, abdominal and/or back pain, fatigue, weight loss, and the presence of abdominal masses [1,4]. The majority of patients have extrarenal spread at presentation and approximately 80% of patients eventually develop metastasis to the lymph nodes, lungs, liver, brain, and bones [1,6]. Bone metastases have a tendency to be osteoblastic and painful. Spread beyond the lymph nodes may occur through the lymphatic system, resulting in lymphangitic carcinomatosis. Lymphangitic carcinomatosis of the lungs is distinct from pulmonary metastases as the tumor emboli do not proliferate or spread locally. They are often seen in collections measuring less than 10 μm and frequently trigger the coagulation cascade and obstruct the flow of blood in pulmonary capillaries. This may result in severe respiratory distress and often becomes the direct cause of death [7]. This case report details the presentation of a patient with rhabdoid collecting duct carcinoma who was found to have lymphangitic carcinomatosis and chylopericardium which rapidly progressed, resulting in her demise.

2. Case presentation

The patient is a 48 year old woman with a medical history of obesity, hypertension, hyperlipidemia, diabetes mellitus, upper extremity

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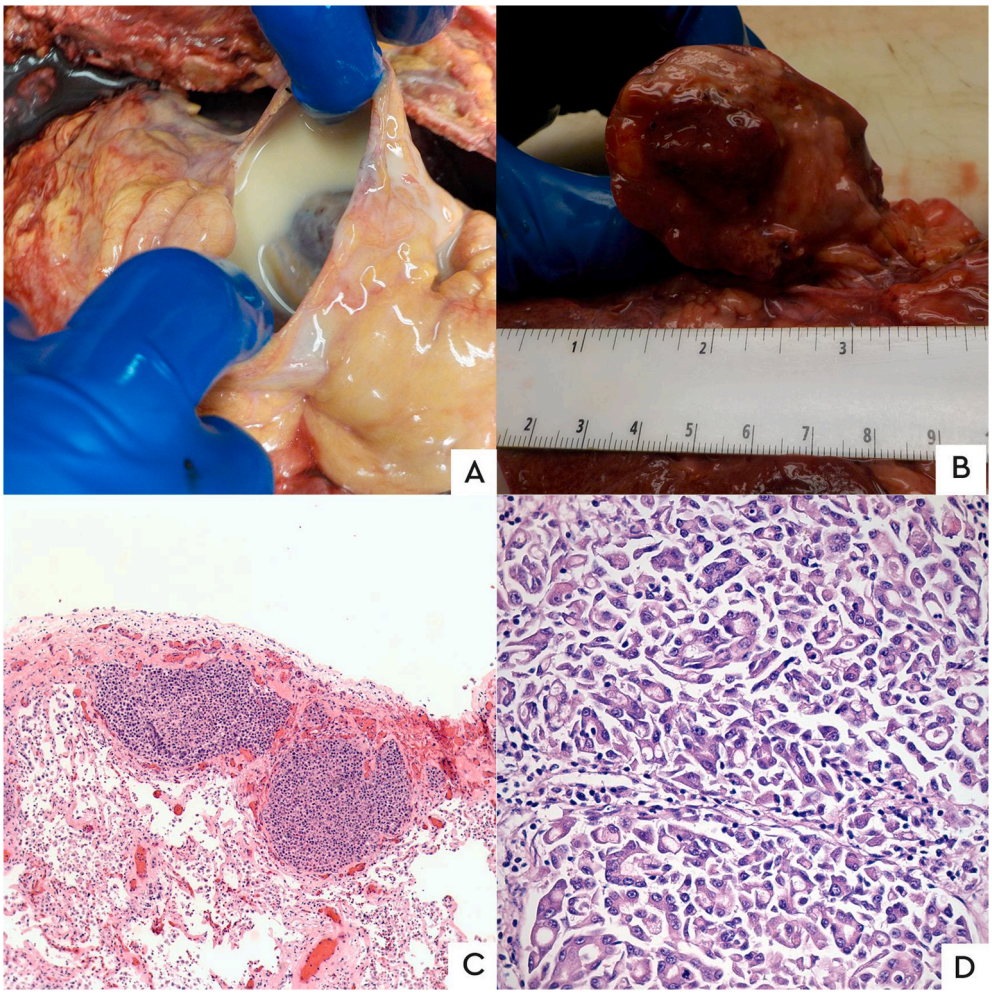

Fig. 1. (A) Gross image of chylous pericardial effusion. (B) Cross section of 5.1 cm tan, hemorrhagic left renal mass (C) Lymphangitic carcinomatosis in lungs (Hematoxylin and eosin stain 100X) (D) Collecting Duct Carcinoma with tubular morphology and rhabdoid features (Hematoxylin and eosin stain 400X).

thrombosis, and pulmonary embolism. Her surgical history is of abdominoplasty, breast reduction surgery, hysterectomy with bilateral oophorectomy, and gastric sleeve. She presented to an outside hospital with dyspnea. A chest computer tomography (CT) scan revealed bilateral pulmonary infiltrates and a neck CT showed bilateral lymphadenopathy. She refused a lymph node biopsy and was discharged on antibiotic therapy. A week later the patient returned to the hospital with worsening dyspnea and a chest CT scan report was unremarkable. An abdominal and pelvic CT scan showed retroperitoneal lymphadenopathy of the left para-aortic and common iliac nodes and left hydroureteronephrosis. Urinalysis was positive for hematuria. The patient was intubated due to respiratory failure. The next day she went into cardiac arrest and despite resuscitation efforts she expired.

3. Pathologic findings

At autopsy, the heart showed a chylous pericardial effusion of 150 cc (Fig. 1A.). The lungs displayed bilateral reticular pattern on the pleural surfaces. Multiple enlarged lymph nodes were identified at the neck and retroperitoneum. The left kidney showed a mass located in the mid portion medulla. Cut section revealed a 5.1 × 4.3 × 3.6 cm circumscribed, tan white mass with a red, beefy, hemorrhagic central area (Fig. 1B). Microscopically, the mass was composed of a tubular proliferation with extensive rhabdoid transformation (Fig. 1D). The cells showed abundant eosinophilic cytoplasm, occasional mucin vacuoles, nuclear pleomorphism, and hyperchromasia. The immunoprofile showed positivity for Pax-8 and CK-7, while CD-10 was negative. The mucin vacuoles were highlighted by positive staining with alcian blue and mucicarmine. These findings supported the diagnosis of a rhabdoid collecting duct carcinoma. The lungs showed diffuse subpleural lymphangitic spread of the carcinoma (Fig. 1C). The enlarged neck and retroperitoneal lymph nodes showed metastatic carcinoma.

4. Discussion

CDC is a rare type of a highly aggressive renal cell carcinoma with poor prognosis. Because of the rarity and diagnostic uncertainty, data on the molecular pathology has not been fully elucidated. The recognition of CDC as a distinct entity is crucial owing to differences of prognosis and therapeutic options. Therefore, to encourage uniformity of diagnosis, the International Society of Urological Pathology (ISUP 2013), and in the 2016 WHO Classifications of Tumors of the Urinary System and Male Genital Organs, emphasize the diagnostic criteria as follows: 1. At least a certain portion of the tumor involves the medullary region; 2. A predominant formation of tubules; 3. A desmoplastic stromal reaction; 4. Cytological high-grade features; 5. Infiltrative growth patterns; 6. And no other RCC subtypes or urothelial carcinoma [8].

The initial presentation of this tumor can often be generalized inflammatory symptoms, that is, fever or general malaise, as well as the classical triad associated with renal malignancy, that is, lumbago, abdominal tumor, and hematuria. Radiologically and grossly, CDC tumors occupy a deep medullary location and present with an infiltrative

lesion. The tumors are usually white, firm with areas of hemorrhage and necrosis. There is an irregular, poorly defined tumor margin with extension into the cortex and often beyond the kidney. They are often large and can be up to 15cm in diameter. The deep medullary location leads to distortion of the renal pelvi-calyceal system. Satellite nodules, particularly in a subcapsular location, are common and it has been argued this is due to intratubular spread [9].

As mentioned before, there should be a predominant tubule formation with papillo-tubular, microcystic, or solid patterns with marked cellular atypia. These lesions are associated with a desmoplastic stroma, which might be caused by the effects of tumor-derived cytokines [10]. Coagulative necrosis is commonly found and a sarcomatoid or rhabdoid transformation can also be present, as in our case. In particular, sarcomatoid or rhabdoid transformation is a morphologic feature bearing poor prognosis [11]. Small vessel invasion is frequently found and renal vein involvement is obvious in approximately 20% of the tumors. It has been reported that CDC consistently expresses high–molecular-weight cytokeratins (CK19 and 34bE12) and cytokeratin 7 and it may show co-expression of vimentin [2,11]. Furthermore, the majority of CDCs display nuclear expression of paired-box gene 2 (PAX2) and gene 8 (PAX8), and SMARCB1 (also called INI-1) as a transcriptional factor or regulator. Especially, PAX8 is consistently positive in CDC, normal collecting ducts, and differentiating nephrons. However, one must bear in mind the fact that PAX8 can also show nuclear positivity in up to 20% of urothelial carcinomas. It is uncommon for collecting duct carcinomas to express the RCC markers CD10, CD117, and kidney specific cadherin [12].

Another clinically unexpected finding in this case is the lymphangitic spread, with the lethal chylopericardium that was seen at autopsy. Multiple foci of the CDC were occupying the lumen of subpleural lymphatic vessels playing a role in the development of a chylous pericardial effusion.

The pathogenesis of pulmonary lymphangitic carcinomatosis (PLC) can be explained by two theories; the first one hematogenously by extravasation of the tumor cells through vessel walls into the perivascular lymphatics. This occurs in 22%–30% of cases and is associated with worse outcomes. The second theory is diffuse retrograde permeation and embolization of lymphatics occurs after involvement of the pulmonary hilar lymph nodes. Pulmonary vessels tend not to be involved. Therefore, pulmonary hypertension is a less common presentation in these cases [13]. Patients are rarely asymptomatic. Progressive subacute dyspnea is the most common presentation in 60% of cases as seen in our patient. Pleuritic pain can also occur when obstructed pleural lymphatics are involved with tumor. Other symptoms including cough, hemoptysis, fatigue, and weight loss are less likely.

What is unique in this scenario is the chylous pericardial effusion as the mechanism of death. Chylopericardium is a rare cause of pericardial effusion. It usually occurs secondarily to trauma, cardiothoracic surgery, radiation therapy, or neoplasm of the mediastinum [14]. Primary chylous pericardial effusions result from retrograde flow through abnormal lymphatics into the pericardial plexus. Such abnormal lymphatic channels may represent lymphangiomas or they may be a part of larger lymphatic tumors [15]. In our case the CDC was diffusely obstructing the lymphatic channels of the lungs causing a retrograde flow into the pericardial plexus and leading to the pericardial effusion.

5. Conclusion

The autopsy in our case provides a clear understanding of the mechanism of death in this patient presenting with a rare entity as is CDC. Neither the renal mass nor the pericardial effusion were seen radiologically. The rapid progression can be challenging to clinicians if the diagnosis is unknown. We highlight the importance of performing complete medical autopsies as an invaluable tool to expand our knowledge.

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Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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