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Case Report

Primary Intimal Sarcoma with Chondrosarcoma Differentiation of the Pulmonary Artery

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Abstract

We report a rare case and our experience of successfully treating intimal sarcoma (IS) with chondrosarcoma differentiation arising in the pulmonary artery. A 36-year-old man presented with a thrombosis-like mass in the pulmonary trunk. Anticoagulant therapy was initiated, but his clinical condition worsened despite adequate anticoagulation. Positron-emission tomography/computed tomography (CT) revealed a neoplastic lesion. The patient underwent endarterectomy and tumor resection. Microscopically, marked cartilaginous differentiation of the cancer cells admixed with pleomorphic, spindle, and round cells was noted. They were immunoreactive for S-100 protein and isocitrate dehydrogenase 1, focal and weak for desmin and murine double minute-2, but negative for ERG, CD34, and myogenin. A diagnosis of IS with chondrosarcoma differentiation was made. The patient received six cycles of adjuvant chemotherapy, and a follow-up chest CT did not show evidence of recurrence.

Keywords: Chondrosarcoma, intimal sarcoma, pulmonary artery neoplasm

Introduction

Intimal sarcomas (ISs) are rare malignant mesenchymal tumors arising from the tunica intima of large blood vessels of the systemic and pulmonary circulation. About two-third of all cases occur in the pulmonary arteries, and about 80% of these cases occur in the pulmonary trunk.[1] IS presents with various histological subtypes of differentiation, including fibrosarcoma, leiomyosarcoma, anaplastic sarcoma, osteosarcoma, and chondrosarcoma.[1] These patients are frequently misdiagnosed with pulmonary embolic disease and inappropriately treated with anticoagulation or thrombolysis for a period of time.[2] Even though the prognosis of most patients is poor due to aggressive tumor growth and progressive heart failure, a recent large observational analysis suggested that a rapid and accurate diagnosis followed by appropriate treatment can improve outcomes.[3] We report a case of IS with chondrosarcoma differentiation which was successfully

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treated. Ethical approval for this study was obtained from our hospital, and we informed both the patient and his family regarding this study and obtained their written informed consent (KMUHIRB-E (I)-20210214).

Case Report

A 36-year-old man was admitted to our hospital with a chief complaint of exertional dyspnea and chest tightness for 2 months. A physical examination revealed a systolic murmur. An ultrasound examination revealed moderate tricuspid regurgitation and possible pulmonary hypertension. Contrast-enhanced chest computed tomography (CT) showed a filling defect with fat content within the pulmonary trunk and right lower pulmonary arteries without any calcification [Figure 1a]. The differential diagnosis included pulmonary thromboembolism and a primary pulmonary artery tumor. The patient initially received anticoagulation for suspected pulmonary embolism; however, his clinical condition worsened despite adequate anticoagulant therapy. Therefore, a fluorodeoxyglucose-positron-emission tomography/CT (FDG-PET/CT) scan was ordered, and a mass with FDG accumulation in the pulmonary trunk was found with a maximum standard uptake value of 10.2. No other mass with increased FDG uptake was detected [Figure 1b]. Under the possible diagnosis of a neoplastic lesion, median sternotomy with the institution of cardiopulmonary bypass and endarterectomy of the pulmonary artery was performed, and the tumor was completely removed from the vessel lumen [Figure 1c].

Gross pathology showed ten grayish-tan soft tissue fragments [Figure 1d]. Microscopically, marked cartilaginous differentiation of the cancer cells admixed with pleomorphic, spindle, and round cells was noted. They were immunoreactive for S-100 protein and isocitrate dehydrogenase 1, focal and weak for desmin and murine double minute, but negative for ERG, CD34, and myogenin [Figure 2]. The features were consistent with high-grade intimal chondrosarcoma. He received six cycles of adjuvant chemotherapy with doxorubicin and ifosfamide and a follow-up contrast-enhanced chest CT scan 3 months later showed no evidence of tumor recurrence. The patient has currently been followed up for 1 year at an outpatient clinic without any known complications.

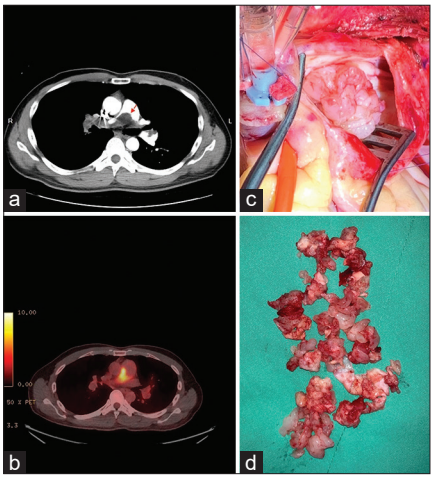

Figure 1: (a) Contrast-enhanced chest computed tomography showed a filling defect with fat content within the pulmonary trunk and right lower pulmonary arteries (red arrow). (b) A fluorodeoxyglucose-positron-emission tomography/computed tomography scan was ordered, and a mass with fluorodeoxyglucose accumulation in the pulmonary trunk was found. (c) Gross picture of the tumor during endarterectomy of the pulmonary artery. (d) Grayish-tan soft tissue fragments of the tumor

Discussion

IS with chondrosarcoma differentiation of pulmonary artery is a rare disease, with only a few case reports reported.[4,5] In the present case, the tumor showed features of sarcoma and involved only the pulmonary artery, findings consistent with a diagnosis of IS. The mean age at diagnosis is 48 years for pulmonary tumors and 62 years for aortic tumors, and the sex distribution is almost equal.[1] In a review of 180 primary sarcomas of large arteries, 61% were classified as ISs, and of these, 26% were differentiated.[6] Differentiated IS presents with a spectrum of histopathological morphologies with variable immunophenotypes. Commonly reported types include angiosarcoma, leiomyosarcoma, myxofibrosarcoma, epithelioid hemangioendothelioma, and osteosarcoma.

A correct diagnosis of IS of the pulmonary artery is challenging and is often misdiagnosed as pulmonary thromboembolism due to its rarity and insidious growth characteristics. The pulmonary artery lesion in our case showed resistance to anticoagulant therapy, leading to the suspicion of neoplasia on FDG-PET/CT and prompt surgery for the tumor. The mean survival in untreated patients is approximately 1.5 months, and more than half of the patients receiving surgical resection died within the 1st year.[7] Furthermore, the presence of distant metastasis has been associated with a more than twofold increase in mortality during follow-up.[3]

Regarding treatment for IS of the pulmonary artery, patients who undergo an attempt at curative resection have been reported to have a longer overall survival (OS) compared to those who undergo incomplete resection (median OS of 36.5 vs. 11 months).[8] Thus, the standard approach is an early aggressive surgery aiming for complete surgical resection with clear margins. As for medical management, due to its rarity, there is no clear consensus concerning the role of systemic therapy. A chemotherapeutic regimen combining doxorubicin and ifosfamide has been demonstrated to be effective regardless of the histological subtype of these tumors. Other agents such as carboplatin, epirubicin, cyclophosphamide, gemcitabine, dacarbazine, etoposide, and vinorelbine have shown a positive effect in some cases.[9] Neoadjuvant chemotherapy is usually recommended to improve the outcomes and facilitate surgery of locally advanced soft tissue sarcoma patients who are not amenable to optimal surgery with clear margins.[10] This treatment approach has led to surgery in initially inoperable

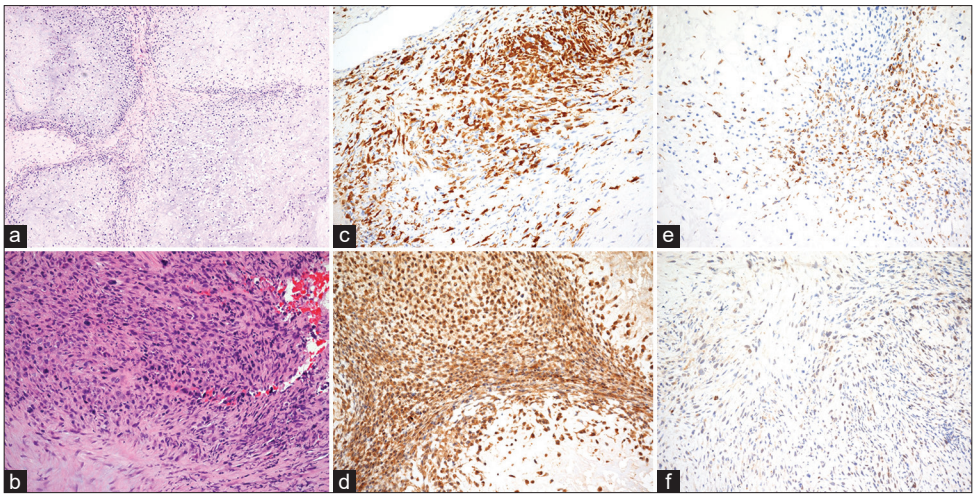

Figure 2: Marked cartilaginous differentiation of the cancer cells admixed with pleomorphic, spindle, and round cells (a and b). They were immunoreactive for S-100 protein (c) and isocitrate dehydrogenase 1 (d), focal and weak for desmin (e) and murine double minute-2 (f)

patients with a positive impact on survival. On the other hand, the available data are not clear concerning the benefits of adjuvant chemotherapy for IS of the pulmonary artery, even though many case reports have demonstrated successful results in favor of chemotherapy in the postoperative setting. Adjuvant therapy in the study by Wong *et al*. was administered to 25% of their patients, with a trend toward improved survival compared to surgery alone (24 vs. 8 months, $P = 0.3417$).[11] Further studies are warranted.

Conclusion

We present a very rare case of IS with chondrosarcoma differentiation arising in the pulmonary artery. Although a diagnosis is often challenging, early detection and histological confirmation of the tumor followed by complete surgery and multidisciplinary collaboration between clinicians, radiologists, and pathologists are fundamental to achieve the best long-term outcomes.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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