Discussion

As a pathologic entity this particular subtype may not constitute a distinct tumor from the other types of clear cell tumors, as suggested in the early 1980s.^{2,4} However, further clinicopathologic, immunohistochemical, and ultrastructural studies of subsequent case reports support CCC as a distinct subtype of large cell carcinoma with purely clear cell features or classified into squamous or adenomatous variants, depending on the type of cellular differentiation.³ In our case, no squamous or adenomatous differentiation was seen, and the tumor grew into and completely occluded the right main bronchus. This biologic behavior appears to be more benign than that seen in other types of non-small cell lung cancer.

Conclusion

CCC has rarely been reported in the literature, and cases of endobronchial compromise are even rarer. Histopathologically, this tumor

must be differentiated from metastatic renal cell carcinoma, benign sugar tumor of the lung, predominant squamous mucoepidermoid carcinoma, and myoepithelial tumor. Immunohistochemical staining of tumor cells facilitates the differential diagnosis of these entities. Because of the low metastatic potential of these lesions and their rarity of recurrence, the role of chemotherapy for these lesions is dubious.

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Successful enucleation of a giant cardiac hemangioendothelioma showing an unusual proliferation pattern

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Primary cardiac hemangioendothelioma is extremely uncommon, and just a few cases treated with surgical intervention are reported in the English literature.¹⁻³ We report a case involving the peculiar appearance of a huge hemangioendothelioma that arose from the right ventricle and was successfully enucleated.

Clinical Summary

A previously healthy 36-year-old woman presented at our cardiac clinic with a 4-month history of cough and increasing edema in both legs. Cardiac examination revealed a Levine grade 2/6 systolic murmur and a sinus rhythm of 76 beats/min. Blood examination was unremarkable, including tumor markers, except for slight anemia (hemoglobin of 9.2 g/dL).

Chest radiography revealed cardiomegaly. On transthoracic echocardiography, it appeared as if the right ventricle was filled by a huge mass encircled with calcification and moderate tricuspid regurgitation. Computed tomography and magnetic resonance imaging of her chest revealed pericardial effusion and a mass lesion along the inferior border of the right ventricle, with calcification of its verge (Figure 1, A). The inner space of the mass matched to a hematoma on both images. Coronary angiography from the right coronary artery (RCA) revealed that the distal RCA was extended and occluded (Figure 1, B). Coronary angiography from the left coronary artery demonstrated late filling of the occluded distal RCA by reversed flow from the left coronary artery.

We diagnosed a cardiac tumor and operated on April 30, 2002. Through a median sternotomy, the pericardium was opened. The heart was surrounded by fibrin tissue, and there was a giant cardiac tumor that pushed up the whole heart in a cephalad manner.

Cardiopulmonary bypass was established with ascending aortic, superior vena caval, and right femoral venous cannulation. After cardiac standstill, the right atrium was opened. The right atrial intima and the tricuspid valve appeared intact. The right ventricular wall, including the RCA, was thin and extended by the cardiac tumor (Figure 1, C). We then cut the right ventricular wall and removed the tumor, which could be enucleated from the right ventricular mural wall without connection to the intraventricular cavity, except for small pin holes. We repaired these small pin holes and the right ventricular wall after trimming.

Histopathologic examination showed the excised tumor was an oval monolocular cyst of 12 × 11 cm in diameter and clearly encapsulated by fibrous connective tissue. There was no extracapsular

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Figure 1. A, Preoperative computed tomography reveals pericardial effusion and a mass lesion along the inferior border of the right ventricle, with calcification of its verge. B, Coronary angiography from the right coronary artery reveals that the distal right coronary artery is extended and occluded. C, The right ventricular wall, including the right coronary artery, is thin and extended by cardiac tumor. D, The inner space of the tumor is filled by hematoma and fibrin.

Figure 2. A, Atypical cells line the inner layer of the tumor wall. B, Formation of delicate vascular channels. C, Mitosis and oval vesicular nucleus or distorted nucleus, large clear nucleolus, and acidophil vacuole. D, Positive staining of CD34 and factor VIII-related antigen.

invasion pathologically. The inner space of the tumor was filled by hematoma and fibrin (Figure 1, D). The wall of the tumor was about 4 mm thick and consisted of fibrous connective tissue with hyaline degeneration and calcification. Atypical cells lined the inner layer of the tumor wall (Figure 2, A). Within the fibrous stroma, there were spindle and polygonal cells that formed a mesh or cordlike structure, and some cells appeared canalized in a formation of delicate vascular channels (Figure 2, B). Furthermore, some of these cells showed mitosis and possessed oval vesicular or distorted nuclei, large clear nucleoli, and acidophil vacuoles (Figure 2, C).

Immunohistochemical analysis showed negative staining for keratin and calretinin; however, positive staining of CD34 and factor VIII-related antigen (Figure 2, D) were clearly revealed. Some showed as heteromorphous, and the Ki-67 labeling index was approximately 25%. Histologic diagnosis was hemangioendothelioma with an unusual proliferation pattern.

The postoperative course was uneventful, and the patient was discharged on the 18th postoperative day. Thereafter, the patient was followed up for about 3 years, with no sign of recurrence.

Discussion

Hemangioendothelioma is a rare cardiac tumor, and just a few cases treated with surgical intervention are reported in the English literature.¹⁻³ The tumor in the case we report here was huge and clearly encapsulated by calcification and fibrous connective tissue, which is why we could enucleate the tumor without difficulty.

Hemangioendothelioma is classified as an intermediate malignancy that sometimes metastasizes and recurs.^{4,5} This might indicate that we should have undertaken additional therapy, but Allaire and colleagues⁴ have reported that irradiation therapy or chemotherapy resulted in only palliation, whereas Hongquan and associates¹ revealed the prognosis of the hemangioendothelioma was relatively good once it was completely resected. In the present case, the tumor was successfully enucleated, and therefore we decided not to undertake additional therapy. The patient was then followed for about 3 years, with no sign of recurrence or metastasis.

Although it appears to have been clinically benign, this is an unprecedented giant tumor that showed an unusually proliferative pattern, and therefore further close follow-up is inevitable.

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Lymphangiomatous hamartoma: Cause or bystander of the isolated chylopericardium?

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Isolated chylopericardium is an extremely rare pathologic entity, and about its cause there is a series of reasons sporadically reported in the literature. Lymphangiomatous hamartoma, a benign tumor of lymphangiomatous origin sparsely located exclusively to the mediastinum, has been considered one of the causes of the chylopericardium.

Clinical Summary

A 25-year-old male smoker of 7 pack-years was referred to our department from a district hospital for rapidly recurrent pericardial effusion (Figure 1) diagnosed 15 days before and showing no response to medication. During the 2 previous days, this progressively increasing effusion had caused clinical and echocardiographic features of pericardial tamponade and had been submitted twice to pericardiocentesis, with removal of 2.5 L and 800 mL of milky fluid, respectively. The laboratory results

Figure 1. Preoperative chest x-ray film demonstrating enlargement of the cardiac silhouette contour caused by tamponade.

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