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CASE REPORT

Sudden cardiac death caused by a primary intimal sarcoma of the left coronary artery

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Abstract Primary sarcoma of a muscular artery is extremely rare. A review of the literature revealed only one report of a sarcoma originating in a coronary artery. We report a case of fatal cardiac infarction caused by an intimal sarcoma originating in the left coronary artery which exhibited immunophenotypic features that were consistent with rhabdomyosarcomatous differentiation.

Keywords Sudden death · Sarcoma · Coronary artery · Myocardial infarction · Immunohistochemistry

Introduction

Primary cardiac sarcomas are the second most common primary cardiac tumor, after myxoma. However, they are quite rare, with an incidence of only 0.16% at autopsies on patients with malignant diseases [1]. The most common primary cardiac sarcoma is the angiosarcoma, followed by malignant fibrous histiocytoma, leiomyosarcoma, rhabdomyosarcoma, and others. Almost half of all primary cardiac sarcomas arise in the left atrium [2]. Sarcomas that originate in a coronary artery, on the other hand, are extremely rare, and a review of the literature yielded only a case of hemangioendotheliosarcoma of the right coronary artery [3]. We report a case of intimal sarcoma with rhabdomyosarcomatous differentiation that arose in the left coronary artery and resulted in sudden death due to acute cardiac ischemia.

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Case history

A 42-year-old Chinese woman became unconscious after a quarrel. She was taken to the hospital, but resuscitation attempts failed. A medicolegal autopsy was conducted 6 h after death.

Autopsy findings

There was no evidence of injury except for a mild subcutaneous hemorrhage of the forehead. Both lungs were edematous and congested. The heart weighed 290 g, and there were no abnormal findings in any of the chambers or valves. However, the lumen of the anterior interventricular branch of the left coronary artery was almost completely obstructed by a small glue-like gray tissue at the point of 2 cm from its origin. Small fibrotic foci were observed in the anterior portion of the left ventricular wall. There was no abnormal finding in the right coronary artery, circumflex branch of the left coronary artery, or other parts of the left ventricular wall. No tumor was found in the intima of the great vessels. No abnormal findings were detected in any other organ.

Histological examination

The lumen of the anterior interventricular branch of the left coronary artery 2 cm from its origin was filled with a small

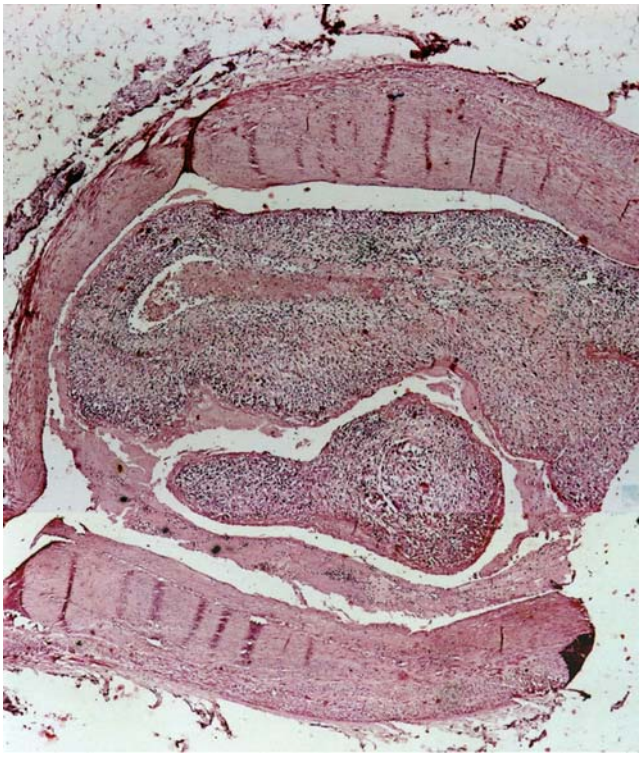

Fig. 1 The lumen of the coronary artery obstructed by a tumor consisting of atypical spindle cells (magnification ×25)

neoplasm. The neoplasm developed within the wall and infiltrated into the muscular layer but not into the adventitia. The neoplasm exhibited a pleomorphic pattern consisting of loose sheaths of intersecting spindle cells and polygonal, tadpole-, or racket-shaped cells (Fig. 1). Multinucleated and binucleated cells were also observed. The nuclei were large with bizarre forms and usually contained prominent nucleoli. Some neoplastic cells contained abundant cytoplasm that stained strongly with eosin (Fig. 2).

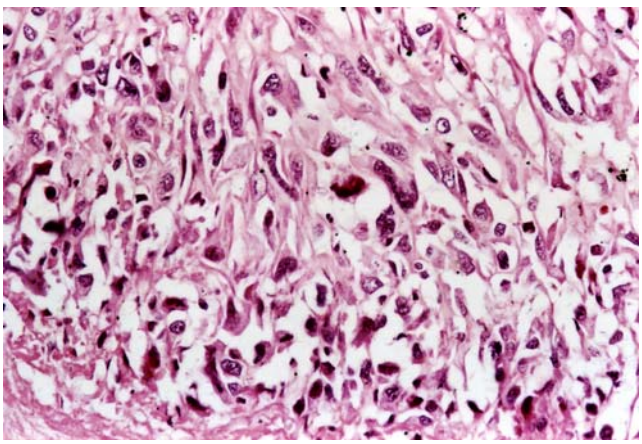

Fig. 2 A higher magnification of the section shown in Fig. 1 showing neoplastic cells containing abundant eosinophilic cytoplasm (magnification ×220)

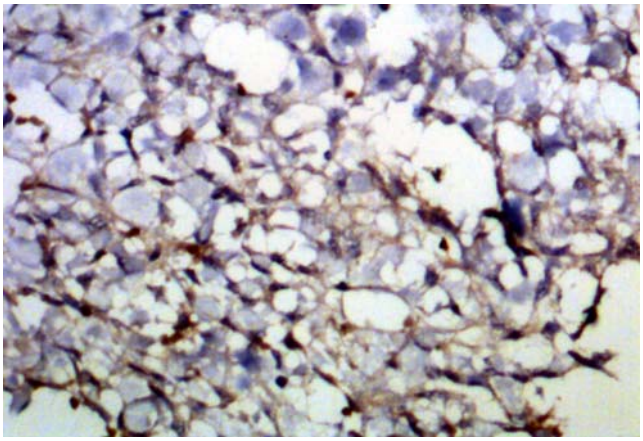

Fig. 3 Cytoplasmic positivity for desmin (magnification ×270)

Histological examinations of the heart exhibited eosinophilia of the muscle fiber, swelling, and depletion of muscle nuclei and interstitial edema in the left anterior wall.

Immunohistochemical staining revealed that the cytoplasm of the neoplastic cells was positive for desmin (Dako; 1:100; Fig. 3) and myoglobin (Dako; 1:1,000; Fig. 4).

Discussion

Arteries are very rare sites of origin for sarcomas, and the most common arteries affected are the pulmonary artery and aorta, especially the abdominal aorta [2, 4, 5]. They can be divided into two types according to their site of origin: an intimal type and a mural type, originating in the media or adventitia [4]. Most primary sarcomas of the aorta and pulmonary artery are intimal, and they are thought to originate from pluripotential mesenchymal cells in the intima. They are consistent with myoblastic tumors with divergent types of cellular differentiation [2]. Approximately 50% of

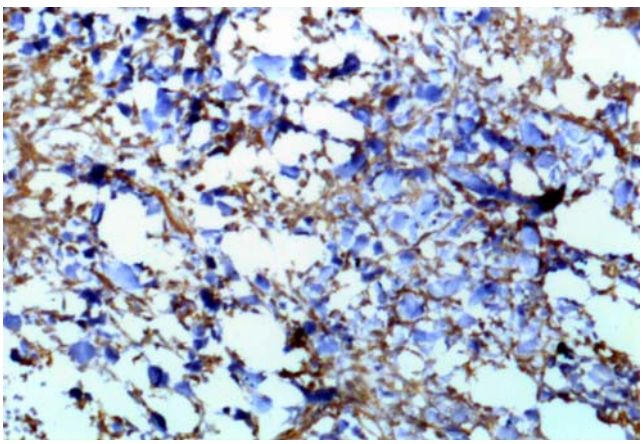

Fig. 4 Cytoplasmic positivity for myoglobin (magnification ×270)

intimal sarcomas of the pulmonary artery are fibroblastic or myofibroblastic sarcomas that are not readily classified and 20% have been classified as leiomyosarcomas. The remaining types of sarcomas have included chondrosarcoma, osteosarcoma, angiosarcoma, malignant mesenchymoma, malignant fibrous histiocytoma, and liposarcoma, and 6% of the sarcomas have been reported to be rhabdomyosarcomas [2]. On the other hand, since many intimal sarcomas of the aorta are not easily classifiable, they have been classified simply as "intimal sarcoma" or "undifferentiated sarcoma" [2, 5]. Application of a wide panel of immunohistochemical markers has recently revealed that undifferentiated intimal sarcomas of the aorta can be further differentiated into two types: sarcomas with myoblastic differentiation and sarcomas with endothelial differentiation [2, 5, 6]. A few cases of aortic intimal sarcomas have been reported as angiosarcoma, malignant fibrous histiocytoma, leiomyosarcoma, and sarcoma with chondrosarcomatous and osteosarcomatous differentiation [2, 5–7]. Only one case of aortic intimal sarcoma has been reported as a sarcoma with rhabdomyosarcomatous differentiation [8]. Since aortic intimal sarcomas cannot be easily classified based on routine histological examinations, immunohistochemical and ultrastructural examinations have been used to make the differential diagnosis of the sarcomas [2, 4– 8]. The histological pattern of large areas in our case resembled that of undifferentiated intimal sarcomas of the aorta with pleomorphic spindle cell growth, although the morphology of some cells suggested rhabdomyosarcomatous differentiation, which was confirmed by the immunohistochemical findings.

Primary sarcomas arising in arteries other than the aorta and pulmonary artery are exceptionally rare, and most are leiomyosarcomas of mural origin [9]. A few cases of sarcomas in muscular arteries have been reported as angiosarcomas [3, 8, 10, 11]. Undifferentiated intimal sarcomas in the muscular arteries that resemble those of the aorta and pulmonary artery are extremely rare. Sebenik et al. [5] reviewed the literature and found only seven cases of undifferentiated intimal sarcoma originating in the carotid, iliac, or femoral artery but found no sarcomas in the coronary arteries. There has been only one report of a case of a sarcoma in a coronary artery. In that case, a hemangioendotheliosarcoma in the right coronary artery was initially manifested by pericardial tamponade secondary to a hemorrhagic pericardial effusion [3]. To our knowledge, ours is the first report of an intimal sarcoma originating in the coronary artery that resembled the intimal sarcomas that occur in the aorta or pulmonary artery.

The prognosis of patients with a sarcoma arising in the aorta or pulmonary artery is generally poor, and metastases by arterial dissemination and tumor embolization to peripheral organs are frequent. On the other hand, some patients live symptom free for years after surgery, especially those with sarcomas of mural origin [2]. Similarly, some patients with leiomyosarcomas in muscular arteries have lived without evidence of metastasis [9]. However, not much is known about the outcome of patients with primary intimal sarcomas of muscular arteries because they are very rare.

Symptoms of primary sarcomas in muscular arteries have included pain and vascular insufficiency. Claudication caused by hypoperfusion due to obstruction of the lumen of the artery by the neoplasm is one of the symptoms of primary sarcomas of the iliac artery and the femoral artery [9]. It is known that myocardial infarction with a non-atherosclerotic coronary artery can occur resulting from numerous conditions in all age group [12] and neoplastic emboli in the coronary arteries are considered a cause of myocardial infarction and death [13]. Moreover, sudden death is one of the topics in forensic pathology [14, 15]. However, primary sarcomas of the coronary artery have been ignored as a cause of myocardial infarction because they are very rare. It is easy to understand that obstruction of a coronary artery by a primary intimal sarcoma results in cardiac ischemia, and our case clearly demonstrates that a primary intimal sarcoma of a coronary artery can cause myocardial infarction and death.

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