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A case of cardiac undifferentiated pleomorphic sarcoma in late pregnancy: A case report

Junlei Zhang[1], Xiaoyan Lin[2] and Yuanhui Yang[1]

Abstract

Cardiac tumors are uncommon and most of them are benign. Although cases of malignant cardiac tumors are rare, it is still necessary to improve awareness in both clinical and pathological diagnosis. Since cardiac tumors often have a high degree of malignancy, it is vital to determine what form of intervention can increase recurrence-free survival and overall survival. In this paper, we report on a 42-year-old woman in the third trimester of pregnancy who had a cardiac undifferentiated pleomorphic sarcoma. According to her medical history, the patient had never had a cardiac tumor or any other disease. She was treated surgically and a left atrial mass was removed immediately after cesarean section. No other treatments were applied after the surgery, and, unfortunately, the tumor reoccurred 6 months later. We reviewed some literature and found one case in which the patient was treated using radiotherapy and survived for another 2 years after the third tumor recurrence. This suggests that neoadjuvant therapy effectively improves the survival rates of such patients.

Keywords

Cardiac malignant, cardiac undifferentiated pleomorphic sarcoma, cardiac tumor

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Introduction

Cardiac undifferentiated pleomorphic sarcoma (UPS) is a high-grade sarcoma that most commonly occurs in the left atrium. The reported cases have not revealed any sex predilection. Both its lobulated appearance and some histopathological structures sometimes mimic myxoma. As high-grade sarcomas, many of them reoccur after surgical excision, and the prognosis of the patients is poor, with survival typically being weeks to months.[1]

At our clinic, we received a 42-year-old female patient who had been pregnant for 36 weeks. Her chief complaint was coughing and difficulty breathing for 4 days. Ultrasound revealed a mass in her left atrium. She was diagnosed with mass in left atrium (cardiac myxoma?) and accepted surgery. Pathological examination suggested that the mass was a cardiac UPS. We are reporting on this case because cardiac UPS is extremely rare, and the patient was a woman in the third trimester of pregnancy. To improve awareness of diagnosing and treating cardiac UPS, we performed a literature review and discussed the treatment and prognosis of the condition. The patient provided written informed consent for the publication of the case report.

Case report

A 42-year-old female in her 36th week of pregnancy was referred to our clinic with the chief complaints of coughing and difficulty breathing for 4 days after having a cold. Before visiting our clinic, she took antibiotics by herself and also received additional antibiotics from her local hospital, both of which were ineffective.

The patient had no history of cardiac diagnosis or surgery and no related signs were detected in routine prenatal examinations during her pregnancy. Due to the blockage caused by the mass, a physical examination disclosed that she had orthopnea, and her cardiac function was evaluated as Grade IV. The electrocardiogram revealed a regular rhythm with a

[1]Medical Integration and Practice Center, Shandong University, Jinan, Shandong, China
[2]Department of Pathology, Shandong Provincial Hospital Affiliated to Shandong First Medical University, Jinan, Shandong, China

Corresponding Author:
Junlei Zhang, Medical Integration and Practice Center, Shandong University, Jinan, Shandong 250100, China.
Email: zhangjl247@163.com

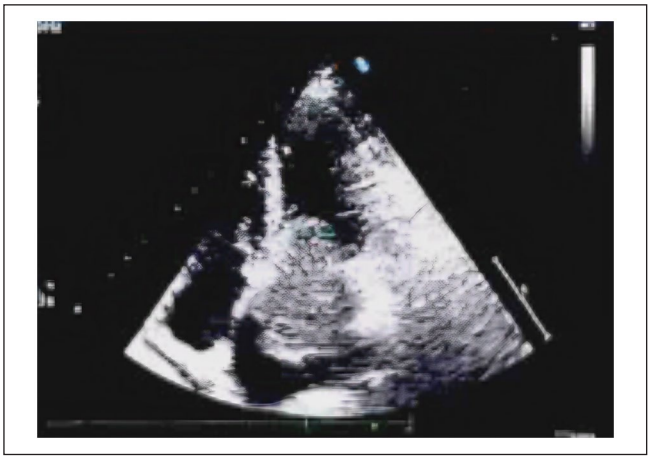

Figure 1. Ultrasound shows space occupying in left atrium.

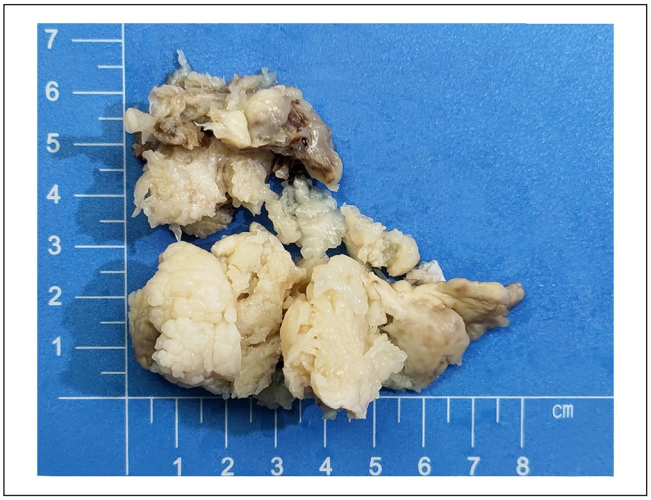

Figure 2. Macroscopically, most parts of the tumor are white and solid lobulated, while some parts are dark red.

heart rate of 92 beats/min. Ultrasound examination suggested that there was a mass in her left atrium (Figure 1), and the patient also had pulmonary hypertension, tricuspid regurgitation, pericardial effusion, and pleural effusion. Cesarean section and heart surgery were performed simultaneously because the presence of a tumor was dangerous to both the patient and her fetus. Ultimately, the fetal delivery was successful, and the atrial tumor was successfully removed.

After surgery, the mass was sent to the pathology department for further examination. Macroscopic examination of the cardiac specimen revealed a solid, white, lobular, villiform mass with no obvious envelope. It was 8 cm long, 4.5 cm wide, and 2.8 cm thick (Figure 2). Histological examination with hematoxylin-eosin (H&E) staining revealed a solid growth with pleomorphic, fusiform-shaped cells. Additionally, the tumor cells were atypical and crowded. Frequent mitosis and necrosis were also observed (Figure 3(a)). In some areas, the tumor cells were surrounded by myxoid stroma (Figure 3(b)).

Based on the information and results mentioned above, we performed immunohistochemistry (IHC) tests. The tumor cells were immunoreactive for vimentin (Figure 4(a)), CDK4 (Figure 4(b)), MDM2 (Figure 4(c)), CD99, SMA, calretinin, WT-1, and caldesmon, and partially positive for keratin, CD34, desmin, and calponin. However, the cells were negative for S-100 (Figure 4(d)), CD31, EMA, and D2-40, while the Ki-67 proliferation index was 70%.

Based on these findings, the tumor was eventually diagnosed as Cardiac UPS. Postoperatively, the patient did not receive any further treatment, and the tumor recurred 6 months later.

Discussion

Cardiac UPS normally occurs in the atrium of the heart. Symptoms of circulation obstruction often appear as the initial sign. In our case, the patient was in her third trimester of pregnancy, so the burden on the heart was more than when she was not pregnant. As a result, her symptoms of cardiac insufficiency were more acute, thereby increasing the chances of tumor detection. According to its position and the probability of all cardiac tumors, we first assumed it was a myxoma.

The gross appearance of cardiac myxoma is variable, but it is generally solid and villiform. Besides, the cut surface is usually variegated due to myxoid tissue and areas of intratumoral hemorrhage.

Microscopically, cardiac myxoma exhibits considerable histological variability. One defining characteristic is the presence of a cytologically bland cell with eosinophilic cytoplasm and an oval or round nucleus, which is called the myxoma cell.[1] The tumor cells are scattered and oriented around vascular channels, while the stroma of the tumor is myxoid and rich. Additionally, degeneration, such as hemorrhage or calcification, is common.

IHC showed that the tumor cells were variably positive for S100, smooth muscle, and endothelial markers CD34, CD31, and factor VIII.[2] In the histological structure of our case, some areas of the tumor were slightly atypical, expressing CD34 and desmin, and surrounded by mucinous matrix. Therefore, we initially believed that this part of the tumor was a myxoma. However, according to her medical history, the patient had never had a myxoma. Although cardiac myxoma often involves a rearrangement of 12p1 and 17p1, there was no genetic evidence to support our inference of cardiac myxoma since the patient had never undergone any genetic testing.

In this case, the macroscopic shape of the tumor was similar to that of a benign myxoma. However, under the microscope, we observed that most areas of the tumor were malignant. In these areas, the tumor cells were densely arranged and poorly differentiated, while the nuclei were

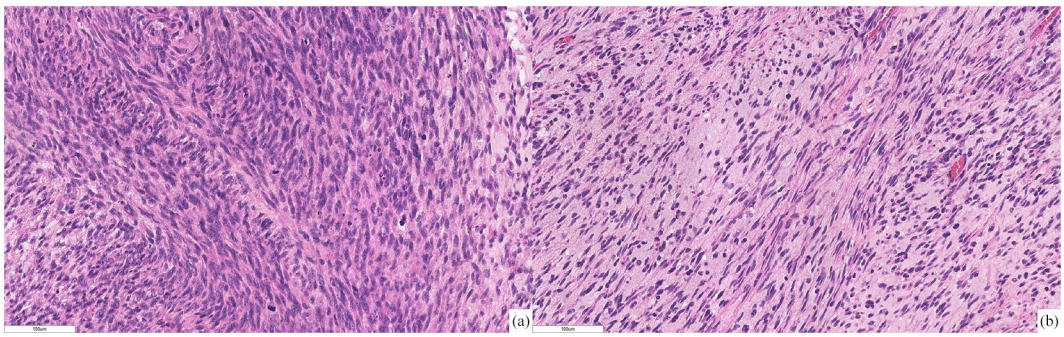

Figure 3. Tumor under microscope, H&E staining, whole slide imaging, 20×; (a) The tumor cells were atypical and crowded; (b) In some areas, the tumor cells were surrounded by myxoid stroma.

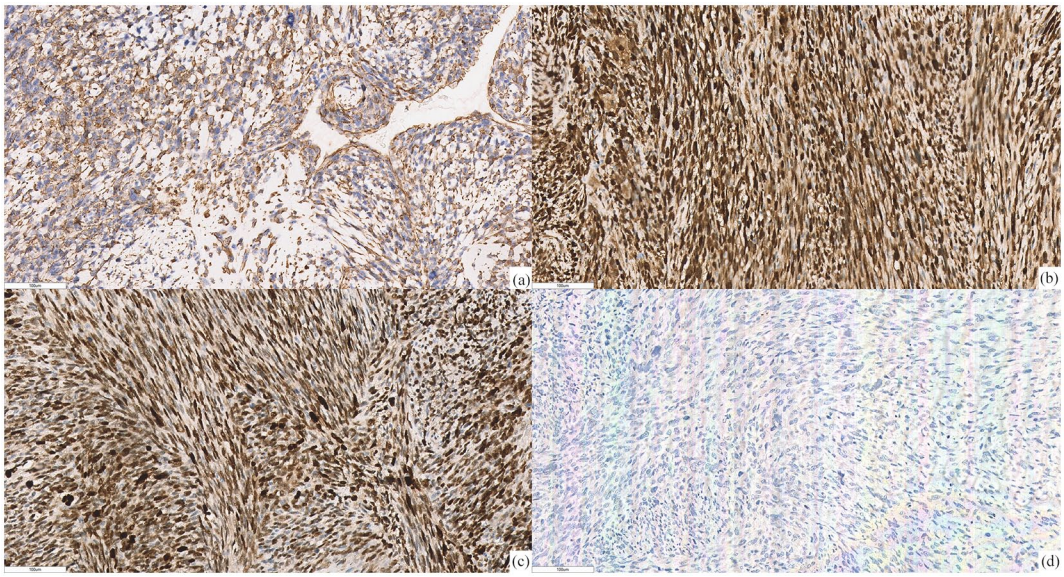

Figure 4. Tumor under microscope, immunohistochemistry staining, whole slide imaging, 20×. (a) tumor cells are positive for vimentin; (b) tumor cells are positive for CDK 4; (c) tumor cells are positive for MDM2; (d) tumor cells are negative for s-100.

large and heteromorphic. Moreover, the Ki-67 proliferation index also reached 70%. Therefore, we made the diagnosis of cardiac undifferentiated pleomorphic sarcoma.

The gross and the microscopic structures of cardiac UPS are often similar to cardiac myxoma. UPS is composed of multiform spindle cells with a large number of mitotic images and common necrosis. Intimal sarcoma is immunoreactive for vimentin and desmin, while MDM2 amplification is common.[3] A retrospective analysis conducted by Neuville et al.[4] assessed 100 cases of cardiac sarcomas. They found that most endometriomas expressed CDK4 and MDM2; some cases partially expressed CD34, but none expressed CD31. Now, cardiac undifferentiated pleomorphic sarcoma and intimal sarcoma are being merged into a single spectrum,[2] so the positive results of vimentin, desmin, MDM2, CDK4, and CD34 and the negative CD31 result in our case were consistent with our diagnosis of UPS. Moreover, another case reported by Higashi et al.[5] confirmed that UPS was positive for desmin and negative for factor VIII, CD31, CD34, and S100. In another case study conducted by Kim et al.,[6] an UPS recurred from a benign myxomatoid hemorrhagic cyst, expressing vimentin but not S100. Based on the similarities among these cases, we inferred that the mass should be diagnosed as cardiac UPS.

Cardiac sarcoma generally has a very poor prognosis. According to the reported cases, it is very common that patients with UPS experience a recurrence only a few months after surgical resection.[7,8] Specifically, the recurrence period in our case was 6 months after surgery. However, some studies have reported that cases have not recurred 9 months after surgery.[9] Furthermore, it is worth noting that neoadjuvant therapy may be a feasible option for prolonging survival.

Hirooka reported on an individual case that involved three instances of surgical resection and recurrence, and the patient received only radiotherapy after surgery. This confirms that radiotherapy may be extremely effective since the patient survives for another 2 years after the third recurrence.[10] Unfortunately, the patient in our case did not receive further treatment other than surgery because of her financial situation and our lack of understanding of the tumor. Thus, it is necessary to improve awareness of pathological diagnosis and provide neoadjuvant therapy or other treatments to enhance recurrence-free survival and overall survival.

Conclusion

Due to the relative rarity of Cardiac UPS, it is crucial to report cases and conduct a literature review to improve awareness of diagnosis and treatment. In our case, the tumor was found during the late pregnancy of the patient, and it was surgically removed immediately after cesarean section. Both the patient and her baby survived the surgery, but the tumor was diagnosed as a cardiac UPS after pathological examination. The patient did not undergo any additional treatment, and the tumor recurred 6 months after surgery. Cardiac UPS is highly malignant, and the prognosis is generally poor. Therefore, more awareness is critical as greater familiarity with the diagnosis and treatment of this disease leads to better outcomes for patients.

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The patient provided written informed consent for the publication of the case.

Author contributions

The original manuscript was written by J.Z., reviewed and edited by X.L. and Y.Y. All authors read and approved the final manuscript.

Declaration of conflicting interests

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Ethics approval

Our institution does not require ethical approval for reporting individual cases or case series.

Informed consent

Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

ORCID iD

Junlei Zhang https://orcid.org/0009-0000-6799-0342

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