MALIGNANT FIBROUS HISTIOCYTOMA DURING PREGNANCY: A CASE REPORT

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SUMMARY

Objective: We present a case of a 38-year-old postpartum woman who had antepartal undiagnosed sarcoma with multiple metastasis. Although the patient underwent aggressive treatment with surgery and chemotherapy, she died 3 months after the vaginal delivery of a healthy female infant weighing 2,090 g at 35 weeks of gestation.

Case Report: The patient had right shoulder pain and mild chest discomfort during the last trimester of the pregnancy. Six days after delivery, she came to our emergency room because her pain had become more severe. A humeral neck tumor with bone destruction was found in the right shoulder on X-ray. After detailed evaluation, right humeral surgery, cardiac surgery, and liver biopsy were performed. All the removed specimens were sent for pathologic examination, and the results showed a sarcoma favoring malignant fibrous histiocytoma with its primary origin in the left atrium.

Conclusion: Obstetricians should be aware that any non-specific complaint may be due to severe disease. It is better to evaluate all symptoms and signs that persist. In this case, early intervention such as radiologic imaging of the bone or echocardiography could have been performed during pregnancy to prevent tumor spread, maternal morbidity, and even death. [Taiwanese J Obstet Gynecol 2006;45(1):86–88]

Key Words: heart, histiocytoma, pregnancy, sarcoma, shoulder pain

Introduction

Malignant fibrous histiocytoma (MFH) is a malignant tumor of fibroblasts and pleomorphic histiocytoid cells with a large number of bizarre, multinucleated giant cells [1]. MFH accounts for 20–24% of soft tissue sarcomas, making it the most common soft tissue sarcoma occurring in late adult life. MFH occurs most commonly in the extremities (70–75%, with lower extremities accounting for 59% of cases), followed by the retroperitoneum. Tumors typically arise in the deep fascia or skeletal muscle. MFH has rarely been reported in the lung, kidney, bladder, heart, stomach, small intestine, and other soft tissues or organs. It is the second most common primary cardiac sarcoma with a frequency of 11.7% [1]. Only 46 cases of primary cardiac MFH have been reported until now [2]. MFH is not often associated with pregnancy [3].

We present the case of a postpartum woman who had antepartal undiagnosed sarcoma with multiple metastasis. Although she underwent aggressive treatment with surgery and chemotherapy, she died 3 months after the vaginal delivery of a healthy baby.

Case Report

A 38-year-old mother, gravida 2, para 1, visited our hospital for the first time at 35 weeks of gestation due to chest tightness. She had normal prenatal care from a local doctor and did not have any systemic disease or history of surgery. She complained of intermittent chest pain and shortness of breath for 1 month. No other symptoms or signs were noted. The physical examination...
revealed stable vital signs and symmetrical expansion with clear breathing sounds. Local tenderness on the left sternal border was noted. We ordered complete electrocardiography (ECG) and transferred the patient to the cardiovascular department under the suspicion of ischemic heart disease. The ECG revealed non-specific ST-segment and T-wave changes, and the patient was recommended to come for outpatient follow-up and echocardiography was scheduled.

After this examination, the patient returned home and preterm premature rupture of the membrane (PPROM) was noted early next morning. A female infant (Apgar score, 9–10; weight, 2,090 g) was born vaginally 6 hours after PPROM. During the 3 days of hospitalization, the patient’s vital signs were stable and intermittent chest pains were noted, but they did not become severe. In addition, she complained of right arm and shoulder pain during hospitalization. We traced her history and she had felt the pain for 3 months but because she thought that her shoulder pain was related to her pregnancy, she did not seek treatment, and only had massage to relieve her pain. She could not describe the pattern of the pain but it was vague and irritating. Physical examination revealed no palpable mass or any visible lesion on the right arm or shoulder. No limitation of the range of motion in the right arm was noted. Therefore, musculofascial pain was diagnosed and a nonsteroidal anti-inflammatory drug was prescribed. Six days after delivery, she came to our hospital emergency room because her right arm pain had become more severe. A humeral neck tumor with bone destruction was found in the right shoulder during X-ray examination (Figure 1). She was transferred to our orthopedic department for further evaluation. Frozen section of the incision biopsy showed pleomorphic sarcoma. After biopsy, scheduled echocardiography revealed a left atrial mass, which suggested myxoma. Transesophageal echocardiography revealed a huge mass (about 4 × 4 cm) over the left atrium. Chest computed tomography showed a mass of about 4 × 4 × 3 cm in the left atrium of the heart (Figure 2) and a heterogeneous mass of about 10.8 × 10 cm in the right lobe of the liver. Cardiac surgeons removed the mass in the left atrium and the patient recovered well. Ten days after cardiac surgery, the orthopedic surgeon excised the right shoulder tumor and reconstructed the shoulder. Liver biopsy was also performed during this admission.

All the removed specimens were sent for pathologic examination, and the results showed a sarcoma favoring MFH with its primary origin in the left atrium (Figure 3).

The bone scan showed multiple bony metastasis. The patient was transferred to the oncology department.

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**Figure 1.** X-ray of right shoulder 6 days after vaginal delivery shows a humeral neck tumor with bone destruction (white arrow).

**Figure 2.** Computed tomography with contrast enhancement of the heart shows a hypodensity mass about 4 × 4 × 3 cm in the left atrium of the heart (black arrow).

**Figure 3.** Pathologic specimen (hematoxylin and eosin, × 200) of a heart sarcoma, composed of markedly pleomorphic, large, and ovoid to irregularly shaped tumor cells, as well as spindle tumor cells, favoring malignant fibrous histiocytoma.
for two courses of adjuvant chemotherapy with liposomal doxorubicin (Lipo-Dox).

Unfortunately, the patient died 3 months after delivery from MFH with multiple metastasis complicated by pneumonia and respiratory failure.

Discussion

MFH sometimes originates from the heart, frequently the left atrium. Because of its location, the symptoms of cardiac MFH are related to pulmonary congestion due to pulmonary vein obstruction, mitral stenosis, mitral regurgitation, and right ventricular failure [1]. Echocardiography is the primary tool for diagnosis of intracardiac mass lesions [4], but a histologic study of the operative specimen is necessary.

Patients with MFH have had poor outcomes because of the high propensity for local recurrence and hematogeneous spread [5]. The most common sites of metastasis are the lung and bone [5]. In our case, we first discovered this tumor during an incisional biopsy of the right humerus.

When this pregnant woman complained about her right shoulder pain and mild chest discomfort during a routine prenatal care examination in the last trimester, we thought the discomfort was possibly related to the last trimester of pregnancy. That is why it is so difficult to diagnose this malignancy during prenatal care.

Early radical surgery, even during pregnancy, can improve survival [6]. The survival of patients with MFH is determined by the metastatic outcomes. Zagars et al reported a 5-year survival rate of 68% with this tumor [5]. In our case, the low immune response during pregnancy and the delayed diagnosis were the most important factors accounting for the metastasis of the tumor.

After discussion with the physicians in the department of hematology and oncology, we decided to prescribe Lipo-Dox as the chemotherapy regimen. For more than 20 years, the Soft Tissue and Bone Sarcoma Group of the European Organization for Research and Treatment of Cancer has been investigating different chemotherapy regimens for advanced and metastatic soft tissue sarcomas. Only a few drugs are active in this disease. Doxorubicin remains the most active single agent, with response rates of 20–25%. In an effort to improve results, many combination regimens have been investigated as first-line treatment in advanced soft tissue sarcoma. Despite higher response rates in some studies, no multidrug regimen has yet been proven superior to single-agent doxorubicin in terms of overall survival [7].

Based on the findings of this case report and other reports described in the literature, we suggest that obstetricians should be aware of the possible existence of MFH, and that careful measures should be taken to avoid tumor spread, maternal morbidity, and even death.

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References