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Primary Right Atrial Cardiac Angiosarcoma in Patient With Poland Syndrome: Case Report and Review of the Literature

Poland Sendromlu Hastada Primer Sağ Atriyal Kardiyak Anjiorsarkom: Olgu Sunumu ve Literatür Taraması

CASE REPORT
OLGU SUNUMU

ABSTRACT

This article presents the case of a 24-year-old woman with Poland syndrome who developed primary right atrial cardiac angiosarcoma. The patient presented to the hospital with dyspnea and chest pain, and imaging studies revealed a large mass attached to the right atrium. Urgent surgery was performed to remove the tumor, and the patient underwent adjuvant chemotherapy afterward. Follow-up exams showed no signs of the tumor or any complications from treatment. Poland syndrome is a rare congenital disorder characterized by the absence of unilateral large pectoral muscle, ipsilateral symbrachydactyly, and other malformations of the anterior chest wall and breast. Although the condition does not predispose patients to malignancy, different pathologies can be seen in these patients due to the unknown etiology of the syndrome. Primary right atrial cardiac angiosarcoma is a rare malignancy, and its coexistence with Poland syndrome has not been well established in the literature. This case report highlights the need to consider cardiac angiosarcoma as a possible diagnosis in patients with Poland syndrome who present with cardiac symptoms.

Keywords: Cardiac angiosarcoma, Poland syndrome, pericardial effusion

ÖZET

Bu yazıda, sağ atriyal kardiyak anjiyosarkom gelişen Poland sendromlu 24 yaşındaki bir kadın olgu sunulmaktadır. Nefes darlığı ve göğüs ağrısı şikayetleri ile hastaneye başvuran hastanın görüntüleme tetkiklerinde sağ atriyuma yapışık büyük bir kitle saptandı. Tümörü çıkarmak için acil ameliyat yapıldı ve daha sonrasında hastaya adjuvan kemoterapi uygulandı. Hastanın takiplerinde tümör belirtisi veya tedaviden kaynaklanan herhangi bir komplikasyon görülmedi. Poland sendromu başlıca pektoralis majör kasının hipoplazisi, ipsilateral değişken derecede üst ekstremite deformiteleri ve göğüs malformasyonları ile karakterize, konjenital bir sendromdur. Sendromun nedeni bilinmediği için hastaların maligniteye yatkınlığı olmasa da, bu hastalarda farklı patolojiler görülebilir. Sağ atriyal kardiyak anjiyosarkom nadir bir malignitedir ve literatürde Poland sendromu ilişkisi iyi bir şekilde belirlenememiştir. Bu olgu sunumu, kardiyak semptomlarla başvuran Poland sendromlu hastalarda kardiyak anjiyosarkomun olası bir tanısı göz önünde bulundurulması gerektiğini vurgulamaktadır..

Anahtar sözcükler: Kardiyak anjiyosarkom, Poland sendromu, perikardiyal effüzyon

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Poland syndrome is a rare congenital disorder presenting with an absence of unilateral large pectoral muscle, ipsilateral symbrachydactyly, and occasionally other malformations of the anterior chest wall and breast.[1] Poland syndrome usually remains undiagnosed until puberty, its etiology is unknown, and its diagnosis is usually based on its symptoms.[2] There is no clear relationship in the literature about the coexistence of Poland syndrome and malignancy. It does not predispose to malignancy.

Case Report

A 24-year-old woman with Poland syndrome was referred to our hospital with dyspnea and chest pain for 2 days. The patient had a history of an artificial left breast prosthesis operation 3 years ago. On admission, her blood pressure was

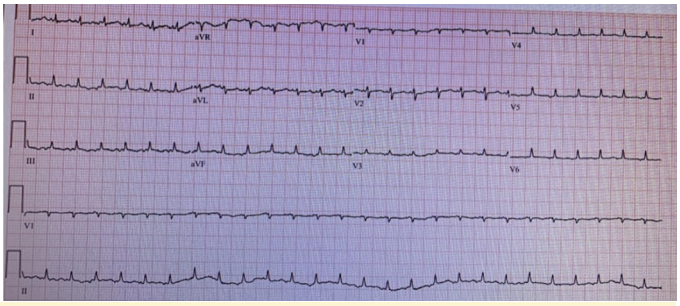

Figure 1. An initial electrocardiography at first ER admission in a 24-year-old woman with sinus tachycardia and low voltage was seen.

85/45 mmHg, her pulse was regular at 140 beats per minute; oxygen saturation was 82% with ambient air. The initial electrocardiogram showed sinus tachycardia (Figure 1).

Laboratory findings revealed leukocytosis and mildly elevated C-reactive protein. Chest radiography and thorax computed tomography (CT) showed cardiomegaly and bilateral pleural effusion (Figures 2 and 3).

Echocardiographic findings showed cardiac tamponade with large amounts of pericardial effusion. Urgent pericardiocentesis was performed through apical approach and about 730 cm^3 of hemorrhagic fluid was drained. Transthoracic echocardiography (TTE) showed an ejection fraction of 60% with

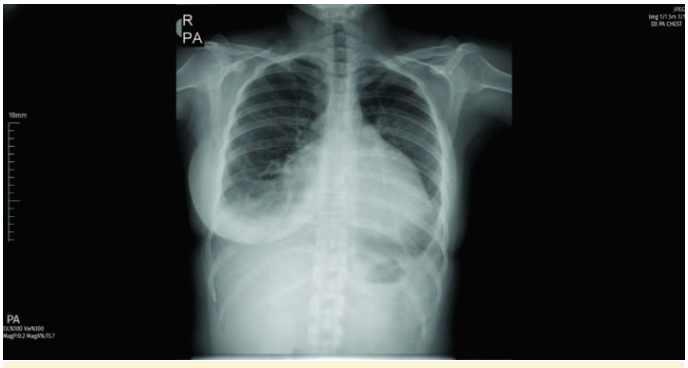

Figure 2. Chest radiography of the patient showed enlargement of cardiac shadow and bilateral pleural effusion.

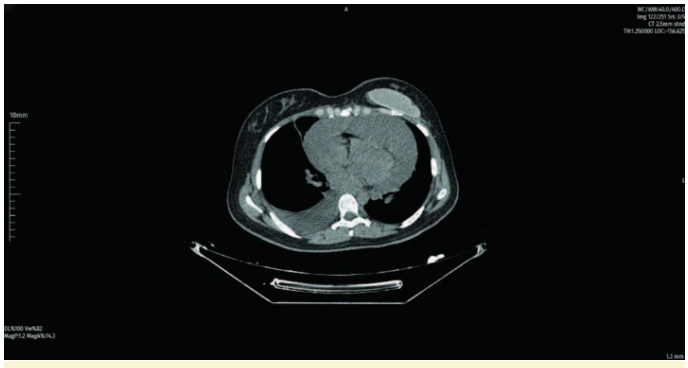

Figure 3. Thorax computed tomography scan of the patient showed pericardial and bilateral pleural effusion.

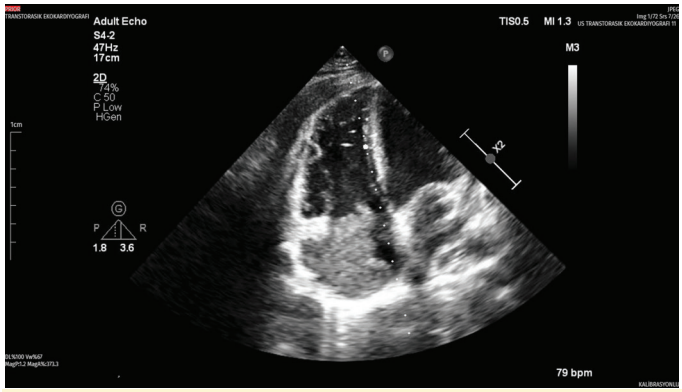

Figure 4. On a pre-operative 2D transthoracic echocardiogram of the patient, about 39 × 42 mm sized lobulated echogenic mass is identified in the right atrium

right atrial mass with an irregular border (Figure 4).

Computed tomography demonstrated a mobile mass (39 × 42 mm) attached to the right atrium extending toward the superior vena cava. Mass in the right atrium was seen in FDG-PET/CT, and in addition, metastases were not seen. In addition to PET/CT, cardiac magnetic resonance imaging (MRI) was performed (Figures 5 and 6).

She was followed by echocardiography regularly in Coronary Intensive Care Unit. Regular echocardiography follow-up showed that tumor size doubled in 2 days. She consulted with surgeons again and then her operation was planned urgently. In the operation, median sternotomy and right atriotomy were performed at first, then the right atrium mass was resected. Next, a bovine pericardium was sutured to reconstruct the right atrium. Postoperative TTE showed no sign of tumor mass and mild-moderate tricuspid regurgitation (Figures 7 and 8).

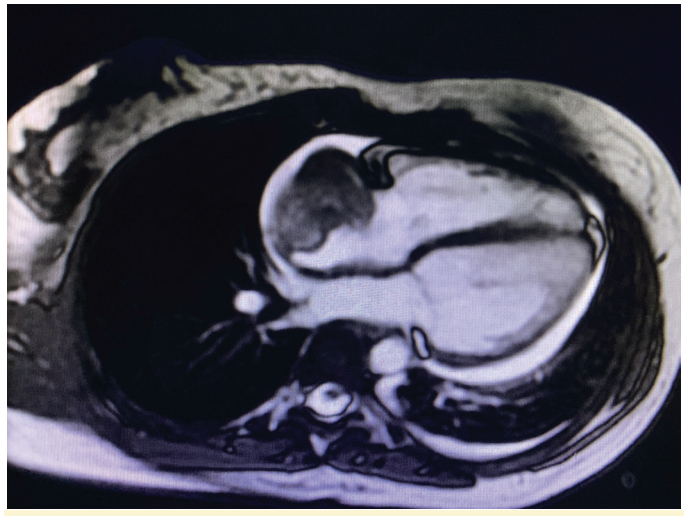

Figure 5. Horizontal and sagittal sections of gadolinium-enhanced cardiac magnetic resonance images demonstrated heterogeneous enhancement within the mass typical of a neoplastic lesion.

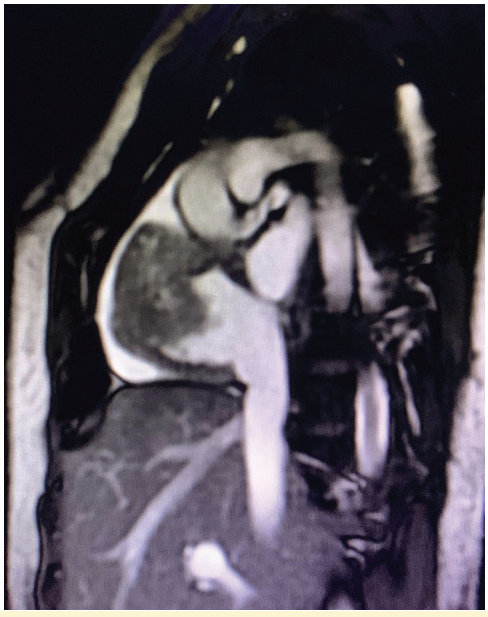

Figure 6. Horizontal and sagittal sections of gadolinium-enhanced cardiac magnetic resonance images demonstrated heterogenous enhancement within the mass typical of a neoplastic lesion.

After 2 weeks, she was discharged. Her effort capacity improved significantly; complaints were completely resolved. After surgery, she underwent adjuvant chemotherapy. She started treatment with concurrent adriamycin (25 mg/m^2), ifosfamide (2500 mg/m^2), and mesna (1500 mg/m^2). At 3-month follow-up, no side effects or complaints were observed, and the patient's complaints were completely resolved. Postoperative third-month transthoracic echocardiography showed no signs of the tumor mass and mild-moderate tricuspid regurgitation.

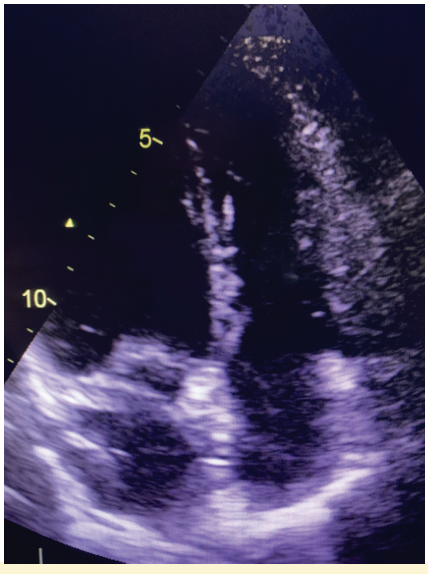

Figure 7. Postoperative third-month transthoracic echocardiography of the patient showed no signs of tumor mass.

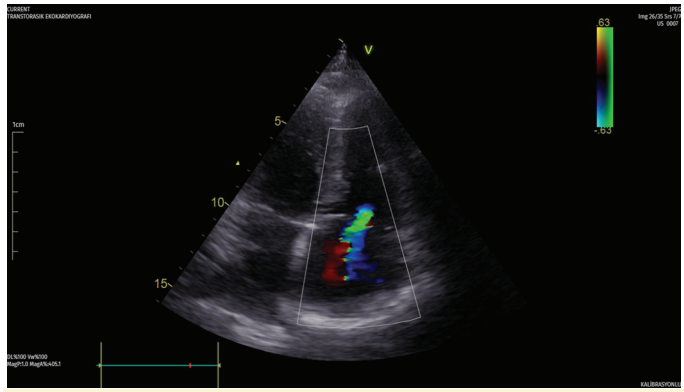

Figure 8. Postoperative third-month transthoracic echocardiography of the patient revealed mild-moderate tricuspid regurgitation.

Discussion

Poland syndrome affects about 1 in 20 000 newborns, and males are affected twice as often as females. The anomalies are usually unilateral and involve the right hemithorax.[2] Etiology of Poland syndrome is unknown, and the pathophysiology is not absolutely understood. There are some theories of its etiology like teratogenic factors, drugs, smoking, and genetic factors. The most believed theory is a transient decrease or interruption in the flow of the subclavian and vertebral arteries during the first 6 weeks of gestation.[3] In Poland syndrome, cardiac pathologies are rarely seen. In the literature, there were case series showing that cardiac dextroposition has been reported with Poland syndrome. The relationship between left-sided Poland syndrome and dextrocardia was shown in the Italian case series. In this Italian case series, 122 patients have registered whose ages varied from 6 months to 53 years, with a median age of 8.9 years. The male-to-female ratio was 2 : 1; 39 patients were female (32%), and 83 patients were male (68%). In 14 patients of this case series, dextrocardia was seen. In these patients with dextrocardia were seen with left-sided Poland syndrome, and there were no environmental factors during pregnancy. Parents of 14 patients with dextrocardia had scoliosis and mild scapular asymmetry. In this case series, mechanical factors during gestation are the most mentioned hypothesis which reveals the relationship between left-sided Poland syndrome and dextrocardia.[3]

Malignant tumors in Poland syndrome are very rare, and the review results of the patients are shown in Table 1. There are case series of breast cancer in patients with Poland syndrome. In these case series, Poland syndrome is usually seen in ipsilateral breast cancer.[4]

Even though Poland syndrome is mostly benign, there are reports of associated malignancies, including leukemia, non-Hodgkin lymphoma, and breast and lung carcinomas. To the best of our knowledge, this is the first case report of

Table 1. Reported Cases of Malignancy in Patients with Poland's Syndrome

	Sex	Age	PS side	Cardiac Defect	Malignancy
Zhang et al.[8]	female	43	left	-	breast cancer
Kurt et al.[9]	male	21	left	-	gastric cancer
Loharkar et al.[10]	male	44		-	gastric cancer
Parikh et al.[11]	male	28	left	-	acute lymphoblastic leukemia
Athale et al.[12]	male	17-month	right	-	Wilm's tumor
Ahn et al.[1]	male	69	right	-	Lung ca
Caksen et al.[13]	male	15	right	-	neuroblastoma
Mojallal et al.[14]	female	28	right	-	contralateral breast ca
Katz et al.[15]	female	42	left	-	ipsilateral breast cancer
Shaham et al.[16]	female	56	right	-	leiomyosarcoma
Wong et al.[17]	female	51	left	-	ipsilateral breast cancer
Okamo et al.[18]	female	59	right	-	contralateral breast cancer
DeFazio et al.[4]	female	62	right	-	contralateral breast cancer
Nakagawa et al.[19]	female	70	left	-	breast cancer
Fukushima et al.[20]	female	57	right	-	ipsilateral breast cancer
Fukushima et al.[20]	female	53	left	-	ipsilateral breast cancer
Havlik et al.[21]	female	33	right	-	ipsilateral breast cancer
Khandelwal et al.[22]	female	71	right	-	ipsilateral breast cancer
Tamiolakis et al.[23]	female	53	left	-	ipsilateral breast cancer
Salhab et al.[24]	female	52	left	-	ipsilateral breast cancer
Yesilkaya et al.[25]	female	39	left	-	2ipsilateral breast cancer
Ji et al.[26]	female	58	left	-	ipsilateral breast cancer
Una et al.[27]	female	39	right	-	ipsilateral breast cancer
Wang et al.[28]	female	46	right	-	ipsilateral breast cancer
Caussa et al.[29]	female	43	left	-	ipsilateral breast cancer
Gerlinger et al.[30]	male	57	right	-	tonsillo-lingual carcinoma
Our case	female	24	left	-	cardiac angiosarcoma

cardiac angiosarcoma in a patient with Poland syndrome. Although cardiac sarcomas are very rare among cardiac tumors, sarcomas consist of different histopathological types, which are angiosarcomas, leiomyosarcomas, and rhabdomyosarcomas, and angiosarcomas is the most common histopathologic type. Right atrium sarcomas are usually angiosarcomas. A cardiac angiosarcoma usually originates from the right atrium close to the atrioventricular sulcus and shows locoregional infiltration which may invade the atrium, ventricles, valves, and pulmonary arteries.[2] Symptoms depend on the degree of tumor infiltration and metastasis. Patients with angiosarcoma may present with arrhythmias, pericardial effusion, emboli, and constitutional symptoms like fever and weakness.

The diagnosis of angiosarcoma is based on multimodality imaging findings. Cardiac MRI is an imaging modality that is more useful than TTE in order to obtain more detailed information about soft tissue characterization and invasion of adjacent tissues of cardiac angiosarcoma.[5] Cardiac MRI can also show the tissue composition of cardiac angiosarcoma which are hypervascularization, necrosis, hemorrhage, and calcification. Definitive diagnosis of a cardiac angiosarcoma is based on cytology and immunohistochemistry of pericardial fluid. In 75%-87% of cases, pericardial fluid cyto-pathology is positive.[6] In our patient, cytologic evaluation of the pericardial fluid showed no evidence of malignancy. Even though malignant cells are not seen in pericardial fluid cytology, cardiac angiosarcoma should be considered in the differential diagnosis of a patient presented with a pericardial effusion.[7] Because cardiac angiosarcoma has poor survival, chemotherapy and radiation therapy are often used to treat after surgery.[7] Postoperative adjuvant chemotherapy regimens include doxorubicin, vincristine, taxanes (paclitaxel and docetaxel), and cyclophosphamide. Since cardiac angiosarcomas have aggressive course and poor prognosis, patients ought to be followed regularly.

Conclusion

Poland syndrome is a rare congenital disorder that may accompany dextrocardia as well as malignancies. Even though solid tumors are rarely seen in a patient with Poland syndrome, it may accompany a cardiac angiosarcoma. Due to its unknown etiology, different pathologies can be seen in these patients.

Informed Consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

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