

Atypical presentation of an infant with idiopathic pulmonary arterial hypertension: pericardial tamponade

İdiyopatik pulmoner arter hipertansiyonu olan bir süt çocuğunda atipik sunum: Perikart tamponadı

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Summary– In this report, we present a five-month-old female patient with a diagnosis of idiopathic pulmonary arterial hypertension (IPAH). This patient was accepted by our clinic after a sudden deterioration in her clinical condition. We performed echocardiography and observed a 17 mm pericardial fluid near the left ventricle posterior wall, which was seen to compress the right atrium and right ventricle outflow tract. Cardiac contraction was impaired, the interventricular septum was hypertrophic and the right ventricle systolic pressure measured by tricuspid regurgitation was at systemic level. Pericardiocentesis was performed for pericardial tamponade, and a drainage tube was placed in the pericardium. Cardiac catheterization was performed before and after nitric oxide inhalation, and there were no changes in the parameters of pulmonary vascular resistance, pulmonary vascular resistance/systemic vascular resistance ratio and mean pulmonary arterial pressure. No cardiac lesion that could cause PAH was detected during catheterization. Under mechanical ventilatory support and intensive medical therapy, the patient died on her 16th day of hospitalization. The importance of our case is that it is the first infantile case presenting with pericardial tamponade associated with IPAH.

Pulmonary arterial hypertension (PAH) is a chronic disorder of a pulmonary vascular course that is characterized by progressive elevation of pulmonary arterial resistance leading to right heart failure.^[1] PAH concludes with death when left untreated. PAH is seen more frequently in childhood than adulthood. In this age group, in terms of etiological factors, it has a wide spectrum. PAH can be hereditary (HPAH), idiopathic (IPAH) or secondary to such pathologies

Özet– Bu yazıda, perikart tamponadı kliniği ile karşımıza çıkan ve idiyopatik pulmoner arter hipertansiyonu (İPAH) tanılı beş aylık bir kız çocuğu sunuldu. Klinik durumunda ani bozulma olması üzerine merkezimize kabul edilen hastanın ekokardiyografisinde, sol ventrikül arka duvarında 17 mm ölçülen, sağ ventrikül çıkım yolu ve sağ atriyumda basıya neden olan perikart sıvısı olduğu gözlemlendi. Kalp kasılması bozulmuştu ve ventriküller arası septum hipertrofisi mevcuttu. Triküspit kapak yetersizliği yoluyla ölçülen sağ ventrikül basıncı sistemik düzeyde idi. Bu bulgularla hastaya perikart tamponadı nedeni ile perikardiyosentez uygulandı ve perikart tüpü yerleştirildi. Ayrıca uygulanan kalp kateterizasyonunda nitrik oksit inhalasyonu öncesi ve sonrası, pulmoner damar direnci/sistemik damar direnci oranı, pulmoner damar direnci ve ortalama pulmoner arter basıncı parametrelerinde bir değişiklik olmadığı gözlemlendi. Kalp kateterizasyonunda pulmoner arter hipertansiyonuna neden olabilecek herhangi bir lezyon tespit edilmedi. Hasta, mekanik ventilatör desteği ve yoğun medikal tedavi altında, yatışının 16. gününde yaşamını yitirdi. Olgumuzun önemi, İPAH ile perikart tamponadı birlikteliği olup bu şekilde sunulan ilk süt çocuğu olgusu olmasıdır.

as connective tissue diseases or congenital heart diseases.^[2] IPAH is a progressive disease in which there are irreversible vascular changes, such as Eisenmenger's syndrome, and in this pathology, no cardiac lesions are found. Although it is known that pericardial effusion can be seen during the follow-up period, knowledge in infants is not clear.

Abbreviations:

IPAH Idiopathic pulmonary arterial hypertension
PAH Pulmonary arterial hypertension

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In this report, we present a five-month-old female patient with a diagnosis of IPAH. The importance of our case is that it is the first infantile case presented with pericardial tamponade associated with IPAH.

CASE REPORT

A five-month-old girl was accepted by our clinic after a sudden deterioration in her clinical condition. Her heart rate was 160/per minute, blood pressure was 55/40 mmHg, and saturation was 83%. She was pale, and her capillary filling time was 2.5 seconds. In her last echocardiographic evaluation, supra-systemic PAH and normal ventricular function were found. We performed echocardiography in our clinic, and observed 17 mm of pericardial fluid near the posterior wall of the left ventricle, which was seen to compress the right atrium and right ventricle outflow tract (Figure 1). Cardiac contraction was impaired, the interventricular septum was hypertrophic, and the right ventricle systolic pressure measured by tricuspid regurgitation was at systemic level. The echocardiographic evaluation was normal except for these findings. Under invasive mechanical ventilatory support, dopamine, adrenaline, and milrinone were started as cardiac support and inhaled nitric oxide and intravenous iloprost as pulmonary hypertension therapy. Pericardiocentesis was performed for pericardial tamponade, and a drainage tube was placed in the pericardium. 155 cc pericardial fluid was drained. The fluid was hemorrhagic, as supported by laboratory tests.



Figure 1. Transthoracic echocardiogram showing pericardial tamponade, severe compression and collapse of right heart chambers. LA: Left atrium; LV: Left ventricle; RA: Right atrium; RV: Right ventricle.

No etiological factors for pericardial tamponade, like hypoalbuminemia, hypothyroidism, viral infectious diseases, or other metabolic reasons were determined in our case. On the third day of the follow-up period, heart contractions were better but pulmonary hypertension remained supra-systemic despite medical treatments. Cardiac catheterization was performed before and after nitric oxide inhalation, and there were no changes in the parameters of pulmonary vascular resistance, pulmonary vascular resistance/systemic vascular resistance ratio and mean pulmonary arterial pressure (Table 1). No cardiac lesion that could cause

Table 1. Hemodynamic and oximetric data of the patient

	Before nitric oxide		After nitric oxide	
	Pressure (mmHg)	Saturation (%)	Pressure (mmHg)	Saturation (%)
Superior vena cava		74.2		79.4
Inferior vena cava		70.6		75.8
Pulmonary vein		99		99
Right atrium	14 / 11 (12)		13 / 10 (11)	
Left atrium	12 / 10 (11)		13 / 11 (12)	
Pulmonary artery	119 / 50 (77)	80	125 / 51 (79)	80.6
Aorta	89 / 49 (68)	97.8	90 / 48 (67)	98.2
PVR		6.3		6.2
SVR		7.1		5.7
PVR/SVR		0.8		1.1

PVR: Pulmonary vascular resistance; Qp: Pulmonary flow; Qs: Systemic flow; SVR: Systemic vascular resistance.

PAH was detected during catheterization. Metabolic screening was done in order to explain metabolic acidosis and lactate elevation in the blood gas, but no specific pathology could be found. While under mechanical ventilatory support and intensive medical therapy, our patient died on her 16th day of hospitalization.

DISCUSSION

Pulmonary arterial hypertension is a nomenclature that is used to define some groups of diseases related with different etiologies, but which lead to the same clinical tables and pathological changes in the pulmonary microcirculation. These changes in the pulmonary vascular structures lead to progressive increase in pulmonary vascular resistance and result in right heart failure and death.^[3] The definition of PAH, which was accepted in the Fourth World Pulmonary Hypertension Conference (Dana Point, USA, 2008), is pulmonary artery pressure measured at rest by right heart catheterization of >25 mmHg.^[4]

The insidious and nonspecific symptoms of IPAH at any age are the factors that delay and complicate diagnosis. Patients can be diagnosed with cardiomegaly, which attracts attention on the chest X-ray after an upper respiratory tract infection with long-term improvement. Symptom-onset is more severe in newborns and infants.^[4] Patients can present with feeding difficulties, lethargy, restlessness, retractions, tachypnea, and tachycardia. Peripheral edema and acrocyanosis are the advanced symptoms in this age group, and severe right heart failure symptoms are seen rarely in young children.^[3] Presence of pericardial effusion and cardiac tamponade is not a well-known clinical picture for IPAH. IPAH associated with pericardial effusion carries a poor prognosis. In the study of Honeycutt et al.,^[5] they evaluated 35 patients with PAH and could not demonstrate a correlation between mortality and the size of pericardial effusion. They stated that, in this patient population, the presence of pericardial effusion itself may be more important than the size of the effusion. Further studies with larger patient cohorts will be needed to clarify the proper evaluation and risk stratification of patients with PAH associated with pericardial effusion. This case report is the first IPAH case in the literature showing a worsening clinical picture with cardiac tamponade.

Today, the aim of the therapy for IPAH is to im-

prove the life quality and functional capacity of the patients. The effectiveness of the targeted therapies developed in the last decade was shown in adult patients. Current guidelines suggest that new treatment algorithms that were created according to evidence-based studies for adult patients can be used for pediatric patients (suggestion Class IIa, level of evidence C).^[3,4] However, controlled randomized studies about the effectiveness and safety of these therapies are limited. Prostacyclin and its analogs, endothelin receptor antagonists and phosphodiesterase inhibitors are the main options that are commonly used for targeted therapy. Initial therapies for functional class II patients can be an oral endothelin receptor antagonist bosentan and for functional class II-III patients can be an oral endothelin receptor antagonist or intravenous epoprostenol infusion. Intravenous epoprostenol infusion should be preferred for the treatment of functional class IV patients.^[6] Our patient was in a very poor condition and was referred to our intensive care unit with pericardial tamponade. First, a pericardial tube was placed into the pericardium to drain the tamponade fluid. Then, with the aim of targeted therapy for PAH, a combined therapy with inhaled nitric oxide (15-20 ppm) and intravenous iloprost infusion was started. During the follow-up period, the desired clinical response could not be achieved, so oral bosentan was added to the treatment. However, despite all medications, there was no decline in the level of supra-systemic levels of PAH.

In conclusion, IPAH is an insidious progressive disease with a very poor prognosis. Clinical symptoms are variable and the presenting symptoms are generally nonspecific. Patients can present with unfamiliar clinical symptoms such as pericardial effusion and tamponade. Thus, when deterioration in the general condition, hypotension and shock are observed in patients with IPAH, together with increasing pulmonary pressure, pericardial tamponade must be considered.

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Key words: Cardiac catheterization; cardiac tamponade; infant; pericardial effusion/etiology/therapy; pulmonary arterial hypertension; tamponade, pericardial.

Anahtar sözcükler: Kalp kateterizasyonu; kardiyak tamponad; yenidoğan; perikart efüzyonu/etyoloji/tedavi; pulmoner arter hipertansiyonu; tamponad, perikart.