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Eisenmenger syndrome presenting with chronic thromboembolic disease

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ABSTRACT

Objectives: Eisenmenger syndrome is characterized by the reversal of blood flow due to increased pulmonary vascular resistance. It can be prevented with early diagnosis and surgical treatment. Thromboembolism is a leading cause of death in patients with Eisenmenger syndrome. Pulmonary endarterectomy is the primary treatment, but medical treatments may be considered in inoperable cases. Regular follow-up and a multidisciplinary approach are important for diagnosis and treatment. Lifestyle changes and medical therapy can improve patient's quality of life and prevent complications. Our case is presented because of the chronic thromboembolic disease in addition to the pulmonary hypertension due to the partial atrioventricular septal defect and the management of the treatment.

Keywords: Eisenmenger syndrome, pulmonary hypertension, chronic thromboembolic disease

Eisenmenger syndrome (ES) is defined as the reversal of blood flow (pulmonary-systemic shunt) or bidirectionality of blood flow due to a severe increase in pulmonary vascular resistance (PVR) in the largest systemic-pulmonary shunts. ES often occurs in people with heart disease. The most frequent cardiac diseases include congenital heart diseases (CHD) such as ventricular septal defect (VSD), atrial septal defect (ASD), and patent ductus arteriosus (PDA) [1]. Erythrocytosis, hyperviscosity, and multiple organ system involvement develop as a result of chronic hypoxia. Treatment of ES may vary according to the severity of symptoms and the patient's age. The aim is to improve heart and lung function and reduce symptoms. Treatment options include drug therapy, oxygen therapy, surgical interventions, and other procedures [2]. Our case is presented because of the chronic thromboembolic disease (CTEH) in addition to the pulmonary hypertension due to the partial atrioventricular septal defect (AVSD) and the management of the treatment.

CASE PRESENTATION

A 66-year-old woman presented to our emergency department with pre-syncope. The patient's oxygen saturation was 60%, respiratory rate was 18/min and arterial blood pressure was 180/110. In history, it was learned that the patient had limited exertion and cyanosis for a long time. Antero-posterior chest radiography showed enlargement of the right hilum and increased density in the left middle and lower zones (Fig. 1). On pulmonary computed tomographic an-

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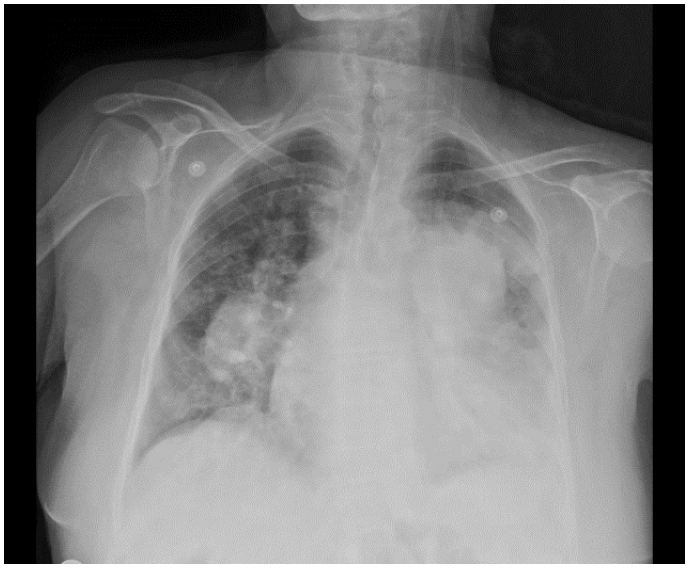

Fig. 1. Antero-posterior chest radiography showed enlargement of the right hilum and increased density in the left middle and lower zones.

giography (CTA), the diameter of the pulmonary artery was approximately 74 mm at its widest point, the diameter of the right main pulmonary artery was 52 mm, the diameter of the left main pulmonary artery was 41 mm at its widest point, and chronic thrombus formation was present in the right main pulmonary artery, middle lobe and lower lobe branches (Figs. 2a and 2b). Echocardiographic examination revealed a single AV valve, partial atrioventricular septal defect (AVSD), pulmonary artery diameter 8 cm, dilated atria, left-sided ventricle hypertrophic, mean pulmonary artery pressure (mPAP): 55 mmHg (Fig. 3). Right heart catheterization was performed (mPAP: 77 mmHg, pulmonary capillary wedge pressure (PCWP): 13 mmHg, PVR: 7.68 WU, vasoreactivity test was negative). The patient's functional classification (FS) was IV according to the New York Heart Association and anticoagulant, diuretic, endothelin receptor antagonist (ERA), and long-term oxygen therapy (LTOT) treatment was started. At the 3-month treatment control, the patient regressed to FS-III, mean PAP: 45 mmHg, and thrombus burden on CTA continued to decrease with a decrease in thrombus burden and was considered inoperable for endarterectomy, so Riociguat was added to the treatment. At the 6th-month treatment check, the patient's resting oxygen saturation was 92% and FS was III, so it was decided to continue clinical follow-up.

DISCUSSION

Eisenmenger syndrome and CTEH are two different entities causing pulmonary arterial hypertension (PAH) with different pathophysiological mechanisms. In cases of Eisenmenger's syndrome due to congenital heart disease, pulmonary hypertension can be prevented with early diagnosis and surgical treatment, and PAH-specific treatments can provide hemodynamic improvement in delayed cases reaching adulthood [1]. The risk of thromboembolism has been studied in several cohorts of congenital heart disease. The results have been variable, depending on the study, the size of the population, and the study methods. Importantly, the risk varies according to the type and complexity

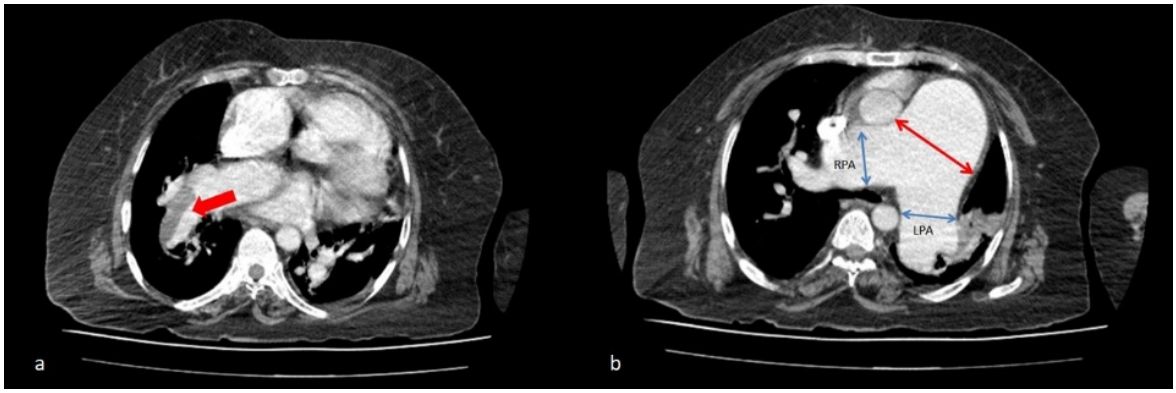

Fig. 2. (a) Chronic thrombus formation in the right main pulmonary artery (red arrow), (b) Enlarged Pulmonary truncus (red line), enlarged right and left main pulmonary artery (blue line).

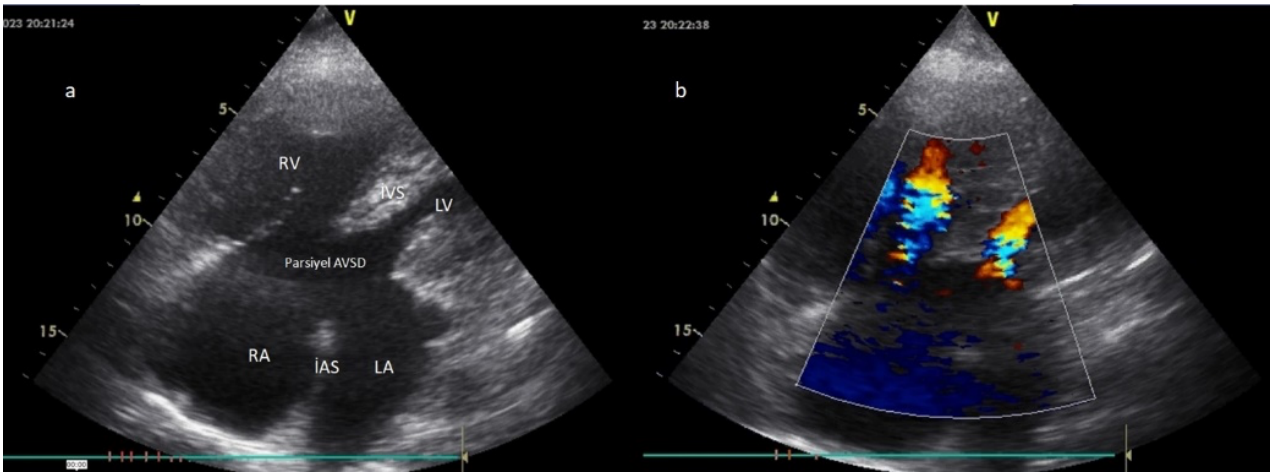

Fig. 3. Partial atrio-ventricular septal defect (AVSD) appearance on echocardiogram.

of the CHD; patients with transposition of the great arteries, univentricular hearts, and cyanotic CHD are at increased risk [4, 5]. There is a significant risk of pulmonary artery thrombus formation in 21-29% of patients with Eisenmenger syndrome [6]. In the study of 34 patients with ES by Silversides K *et al*. the prevalence of proximal pulmonary artery thrombi was 21% (7/34). Thrombosis of more distal vessels was detected in 43% (3/7%) of patients with thrombus in the proximal pulmonary arteries, and patients with thrombus were more likely to be female (86% vs. 37%, P=0.04) and to have lower oxygen saturation (72±9% vs. 85±6%, P=0.01) [7]. Hjortshøj *et al*. [8] in a retrospective study of 1546 patients between 1977 and 2015 to determine cause-specific mortality in ES, thromboembolism was found to be 8% of the leading causes of death. CTEH is also a condition that should be evaluated among the etiological causes of PAH. Although the primary treatment is pulmonary endarterectomy in appropriate cases, medical treatments should be considered in inoperable cases [3].

CONCLUSION

Pulmonary hypertension is a serious health problem and early diagnosis and treatment are very important. The association of Eisenmenger's syndrome and CTEH is a rare condition that can have serious consequences. The diagnosis and treatment of these conditions requires a multidisciplinary approach and regular follow-up of patients is important. Medical therapy, surgical intervention, and lifestyle changes play an important role in the management of these diseases. This can improve patients' quality of life and prevent serious complications.

Patient' Consent

Patients was informed about the purpose of the case report, and informed consent was obtained from the patient for this publication.

Authors' Contribution

Study Conception: ACP, KK, TY; Study Design: ACP, KK, TY; Supervision: ACP, KK, TY; Funding: ACP, KK; Materials: ACP, KK; Data Collection and/or Processing: ACP, KK; Statistical Analysis and/or Data Interpretation: ACP; Literature Review: ACP; Manuscript Preparation: ACP and Critical Review: ACP.

Conflict of interest

The authors disclosed no conflict of interest during the preparation or publication of this manuscript.

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REFERENCES

1. Basit H, Wallen TJ, Sergent BN. Eisenmenger Syndrome. [Updated 2023 Feb 13]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023. Available from: https://www.ncbi.nlm.nih.gov/books/NBK507800/
2. Diller GP, Lammers AE, Oechslin E. Treatment of adults with Eisenmenger syndrome-state of the art in the 21st century: a short overview. Cardiovasc Diagn Ther. 2021;11(4):1190-1199. doi: 10.21037/cdt-21-135.
3. Otani N, Watanabe R, Tomoe T, Toyoda S, Yasu T, Nakamoto T. Pathophysiology and Treatment of Chronic Thromboembolic Pulmonary Hypertension. Int J Mol Sci. 2023;24(4):3979. doi: 10.3390/ijms24043979.
4. Diller GP, Kempny A, Alonso-Gonzalez R, et al. Survival Prospects and Circumstances of Death in Contemporary Adult Congenital Heart Disease Patients Under Follow-Up at a Large Tertiary Centre. Circulation. 2015;132(22):2118-2125. doi: 10.1161/CIRCULATIONAHA.115.017202.
5. Engelfriet P, Boersma E, Oechslin E, et al.The spectrum of adult congenital heart disease in Europe: morbidity and mortality in a 5 year follow-up period. The Euro Heart Survey on adult congenital heart disease. Eur Heart J. 2005;26(21):2325-2333. doi: 10.1093/eurheartj/ehi396.
6. Broberg CS, Ujita M, Prasad S, et al. Pulmonary arterial thrombosis in Eisenmenger syndrome is associated with biventricular dysfunction and decreased pulmonary flow velocity. J Am Coll Cardiol. 2007;50(7):634-642. doi: 10.1016/j.jacc.2007.04.056.
7. Silversides CK, Granton JT, Konen E, Hart MA, Webb GD, Therrien J. Pulmonary thrombosis in adults with Eisenmenger syndrome. J Am Coll Cardiol. 2003;42(11):1982-1987. doi: 10.1016/j.jacc.2003.07.022.
8. Hjortshøj CMS, Kempny A, Jensen AS, et al. Past and current cause-specific mortality in Eisenmenger syndrome. Eur Heart J. 2017;38(26):2060-2067. doi: 10.1093/eurheartj/ehx201.