Folia Cardiologica 2021
vol. 16, no. 2, pages 130–133
DOI: 10.5603/FC.2021.0015

ISSN 2353–7752
e-ISSN 2353–7760

Severe chronic thromboembolic pulmonary hypertension in a patient after splenectomy for hereditary spherocytosis: the need for long-term surveillance and closer cooperation between haematologists and cardiologists

Ciężkie przewlekłe zakrzepowo-zatorowe nadciśnienie płucne po splenektomii z powodu sferocytozy wrodzonej — potrzeba długoterminowego nadzoru i ścisłej współpracy hematologów i kardiologów

Maja Klimkiewicz[1], Grzegorz Sławiński[1], Anna Faran[1], Alicja Dąbrowska-Kugacka[1], Grzegorz Romanowicz[2], Marcin Kurzyna[3], Ewa Lewicka[1]

[1]Department of Cardiology and Electrotherapy, Medical University of Gdansk, Poland
[2]Department of Nuclear Medicine, Medical University of Gdansk, Poland
[3]Department of Pulmonary Circulation, Thromboembolic Diseases and Cardiology CMKP, Otwock, Poland

Abstract

We present the case of a 34-year-old female patient with severe pulmonary hypertension diagnosed > 15 years after splenectomy to emphasize the need to monitor patients after splenectomy for the development of chronic thromboembolic pulmonary hypertension (CTEPH). The lack of screening tests for this group of patients, as in the described case, resulted in a serious clinical condition of the patient at the time of diagnosis. Without specialized treatment, the probability of her survival at 5 years was only 10%. Close collaboration between haematologists and cardiologists is mandatory in the management of post-splenectomy patients Life-long surveillance is needed in these patients due to the risk of CTEPH. The development of a screening program for post-splenectomy patients is required.

Key words: chronic thromboembolic pulmonary hypertension, CTEPH, pulmonary hypertension, PH, splenectomy

Folia Cardiologica 2021; 16, 2: 130–133

Introduction

We present a case of a young patient with severe pulmonary hypertension (PH) diagnosed > 15 years after splenectomy to emphasize the need for monitoring of patients after splenectomy for the development of chronic thromboembolic pulmonary hypertension (CTEPH).

Case report

A 34-year-old woman was admitted to the hospital due to suspected PH. In childhood, she underwent splenectomy due to congenital spherocytosis at the age of 16 (2002). In December 2018, there was a pulmonary embolism (PE) treated with thrombolysis, and a month later she underwent

Address for correspondence: Prof. Ewa Lewicka MD, PhD, Klinika Kardiologii i Elektroterapii Serca, Gdański Uniwersytet Medyczny, ul. Debinki 7, 80–211 Gdańsk, Poland, phone: +48 58 349 39 10, e-mail: elew@gumed.edu.pl

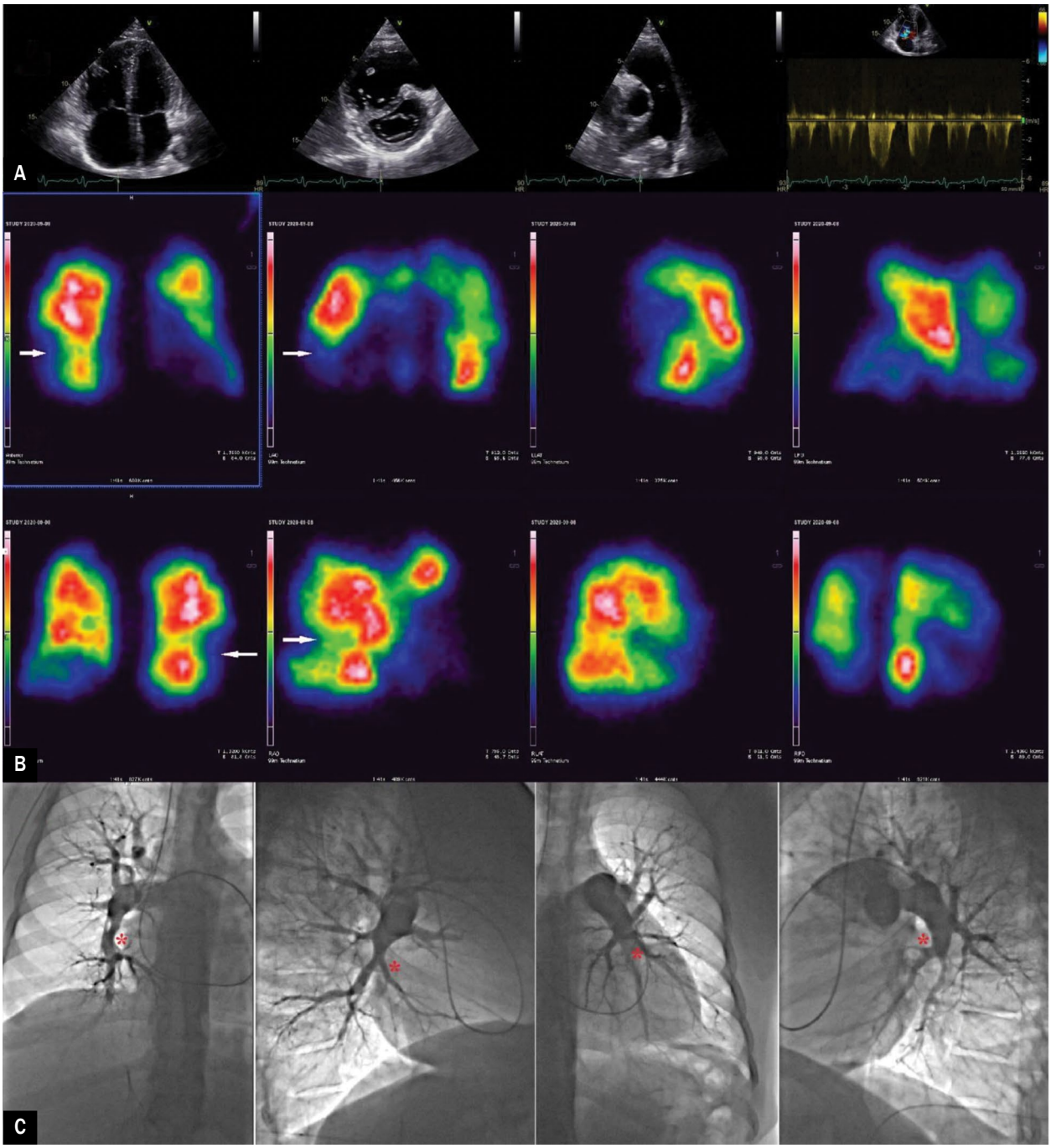

Figure 1A. Echocardiography shows the enlargement of the right heart chambers and pulmonary trunk: right ventricle (RVID) 53 mm, right atrial area 28 cm^2, RV:LV ratio 1.4. D-shaped left ventricle in systole, secondary to interventricular septum flattening, indicates the right ventricular pressure overload. Severe tricuspid regurgitation is shown with the peak velocity of 4.8 m/s; **B.** Lung scintigraphy shows numerous segmental and subsegmental perfusion deficits in both lungs (white arrows); **C.** Pulmonary angiography reveals the widening of main branches of the pulmonary artery, organized clots located in the distal parts of both lower lobes branches (red asterisks) and multiple webs lesions at segmental levels. From the left: right pulmonary artery (posteroanterior and right anterior-posterior oblique 90 views) and left pulmonary artery (posteroanterior and left anterior-posterior 90 oblique views)

surgical recanalization of deep veins in the right lower limb and filter implantation into the inferior vena cava. Three months later, deep vein thrombosis was diagnosed, while taking enoxaparin. Despite anticoagulation, PE recurred in August 2019 and left iliac vein thrombosis was diagnosed. Thrombophilia was excluded.

On admission, the patient complained of progressive dyspnoea [New York Heart Association (NYHA) III] and recurrent syncopes.

In laboratory tests: haemoglobin 17 g/dL, haematocrit 53%, platelets 560 × 10^9/L and BNP 169 pg/mL. Electrocardiography (ECG) showed sinus tachycardia, right axis deviation, P-pulmonale, right ventricular (RV) hypertrophy.

Echocardiography revealed (see Supplementary videos):

- normal left ventricular (LV) systolic function: LV ejection fraction 60%;
- right heart and pulmonary trunk enlargement;
- impaired RV systolic function: three–dimensional RV ejection fraction 31%, RV free wall longitudinal strain –10%, RV S' 10 cm/s;
- severe tricuspid regurgitation, and RV systolic pressure of 100 mmHg (Figure 1A).

Angio-CT showed no evidence of acute PE. Perfusion scintigraphy revealed numerous perfusion deficits in both lungs at the segmental and subsegmental levels (Figure 1B). Right heart catheterization indicated precapillary PH: mean pulmonary artery pressure (mPAP) was 57 mm Hg, pulmonary vascular resistance 8.93 Wood units, pulmonary capillary wedge pressure 12 mm Hg, and the cardiac index was 2.56 L/min/m^2. Pulmonary artery angiography showed numerous contrast deficits (Figure 1C), and finally, CTEPH was diagnosed. The patient was qualified for urgent pulmonary endarterectomy (PEA) and removal of the inferior vena cava filter.

Discussion

Splenectomy is considered a risk factor for CTEPH development and a history of splenectomy is found in 3.4–9.1% of CTEPH patients [1, 2]. CTEPH may occur many years after splenectomy [3]. The diagnosis of CTEPH may be challenging, but this does not fully explain the late diagnosis in the presented case. This could be due to low awareness of the risk of CTEPH after splenectomy and the lack of recommendations for monitoring these patients. CTEPH is a rare disease that, if untreated, leads to RV failure and death. In the case of untreated CTEPH and mPAP exceeding 50 mm Hg (57 mm Hg in the presented patient), 5-year survival was only 10% [4]. However, successful treatment with PEA reduces the risk of death by 67% compared to non-operated patients [5]. In patients not eligible for surgery, percutaneous balloon pulmonary angioplasty is an option. Also targeted pharmacotherapy for PH is administered, mainly using riociguat.

Conclusion

Close collaboration between haematologists and cardiologists is mandatory in the management of post-splenectomy patients. They require life-long surveillance due to the risk of CTEPH. The development of a screening program for post-splenectomy patients is required.

Conflict of interest

The authors declare no conflict of interest.

Streszczenie

Przedstawiono przypadek 34-letniej pacjentki z ciężkim nadciśnieniem płucnym (PH) zdiagnozowanym ponad 15 lat po splenektomii, aby podkreślić potrzebę monitorowania pacjentów po tym zabiegu pod kątem rozwoju przewlekłego zakrzepowo-zatorowego nadciśnienia płucnego (CTEPH). Jak dotąd, brakuje zaleceń odnośnie do badań przesiewowych u chorych po splenektomii, co w opisanym przypadku skutkowało ciężkim PH u chorej w momencie podstawienia diagnozy. Bez podjęcia specjalistycznego leczenia prawdopodobieństwo jej 5-letniego przeżycia wynosiło zaledwie 10%. Pacjenci po splenektomii wymagają ścisłej współpracy między hematologami i kardiologami oraz stałej obserwacji przez całe życie ze względu na ryzyko wystąpienia CTEPH. Konieczne jest opracowanie programu badań przesiewowych dla pacjentów po splenektomii.

Słowa kluczowe: zakrzepowo-zatorowe nadciśnienie płucne, CTEPH, nadciśnienie płucne, PH, splenektomia

Folia Cardiologica 2021; 16, 2: 130–133

References

1. Jaïs X, Ioos V, Jardim C, et al. Splenectomy and chronic thromboembolic pulmonary hypertension. Thorax. 2005; 60(12): 1031–1034, doi: 10.1136/thx.2004.038083, indexed in Pubmed: 16085731.
2. Pepke-Zaba J, Delcroix M, Lang I, et al. Chronic thromboembolic pulmonary hypertension (CTEPH): results from an international prospective registry. Circulation. 2011; 124(18): 1973–1981, doi: 10.1161/CIRCULATIONAHA.110.015008, indexed in Pubmed: 21969018.
3. Galiè N, Humbert M, Vachiery JL, et al. ESC Scientific Document Group. 2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension: The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS): Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC), International Society for Heart and Lung Transplantation (ISHLT). Eur Heart J. 2016; 37(1): 67–119, doi: 10.1093/eurheartj/ehv317, indexed in Pubmed: 26320113.
4. Riedel M, Stanek V, Widimsky J, et al. Longterm follow-up of patients with pulmonary thromboembolism. Late prognosis and evolution of hemodynamic and respiratory data. Chest. 1982; 81(2): 151–158, doi: 10.1378/chest.81.2.151, indexed in Pubmed: 7056079.
5. Delcroix M, Lang I, Pepke-Zaba J, et al. Long-term outcome of patients with chronic thromboembolic pulmonary hypertension: results from an International Prospective Registry. Circulation. 2016; 133(9): 859–871, doi: 10.1161/CIRCULATIONAHA.115.016522, indexed in Pubmed: 26826181.