CASE REPORT
OPIS PRZYPADKU

PERCUTANEOUS LEFT ATRIAL APPENDAGE CLOSURE IN A PATIENT WITH ATRIAL FIBRILLATION AND OSLER-WEBER-RENDU DISEASE

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ABSTRACT

We present a case report of a 79-year-old woman with permanent atrial fibrillation and Osler-Weber-Rendu disease who underwent percutaneous closure of left atrial appendage. The patient had a history of small bowel resection due to mesenteric embolism and recurrent episodes of gastrointestinal bleeding, epistaxis and hemarthrosis. Bleeding episodes were exacerbated by anticoagulation therapy causing severe anemia that required repeated red blood cells transfusions. Left atrial appendage occlusion with Watchman 30 mm LAA Device was successfully performed. In long-term observation no thromboembolic event, epistaxis or severe bleeding have occurred.

KEY WORDS: Osler-Weber-Rendu disease, Hereditary hemorrhagic telangiectasia, atrial fibrillation, left atrial appendage closure

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INTRODUCTION

Atrial fibrillation (AF) is the most common cardiac arrhythmia. Prevalence of AF is 3% of the general population and still increasing. One of the most important thing about treatment of patients with atrial fibrillation is prevention of thromboembolic complications [1–3]. Anticoagulation therapy by VKA (warfarin, acenocoumarol) or NOAC (apixaban, dabigatran, edoxaban, rivaroxaban) are highly effective however may cause severe bleeding complications especially in patients with coagulation or vascular disorders [4, 5]. One of the vascular disorders is Osler-Weber-Rendu disease (OWRD) known as hereditary hemorrhagic telangiectasia (HHT) [6].

CASE PRESENTATION

We present a case report concerning a 79-year-old woman with atrial fibrillation and Osler-Weber-Rendu disease. The syndrome was diagnosed on the basis of clinical Curacao criteria : recurrent severe epistaxis, presence of skin and mucosa lesions (telangiectasias of face, ears, lips, hands, tongue) and positive family history (first-degree relatives with diagnosed OWR disease) (Fig. 1 A–C) [7]. Genetic testing had not been performed. Comorbidities included atherosclerosis, arterial hypertension, type 2 diabetes , chronic heart failure, chronic kidney disease and hypothyroidism. Additionally, the patient had a history of small bowel resection due to mesenteric embolism.

In March 2015 she was hospitalized in the Department of Internal Medicine because of heart failure symptoms. During hospitalization atrial fibrillation was diagnosed for the first time. Due to high thromboembolic risk (CHA2DS2-VASc score of 9) oral anticoagulant was recommended. After hematologic consultation warfarin was introduced. The patient was also qualified for urgent pacemaker implantation due to sinus sick syndrome. After the procedure oral anticoagulation was sustained.

In July 2015 she was readmitted to the Department of Internal Medicine due to extensive epistaxis. Laboratory tests revealed severe anemia requiring blood transfusions. Because of persistent nosebleeds anterior nasal packing and coagulation was performed. After a few days of observation the patient was discharged in good general condition.

Two weeks later she was hospitalized again for recurrent epistaxis. Laboratory tests showed INR of 7,3 despite proper use of warfarin and moderate anemia. The patient received vitamin K and cyclonamine along with supplementation of iron, cobalamine and folic acid. Oral anticoagulation was continued. She was referred to cardiology department to be evaluated for percutaneous closure of left atrial appendage (LAA).

Due to high thromboembolic and bleeding risk the patient was qualified for transcatheter LAA occlusion. Before the procedure transesophageal echocardiography (TEE) was done. TEE revealed windsock-shaped LAA with no thrombus inside. Based on the result of imaging examination (TEE) LAA morphology was determined to be suitable for occlusion with Watchman device. LAA closure was performed with three dimensional TEE guidance under general anaesthesia and a Watchman 30 mm LAA Closure Device was successfully implanted (Fig. 1D). The procedure and recovery were uneventful and after two days the patient was discharged home in good general condition. Dual antiplatelet therapy was prescribed. After 45 days

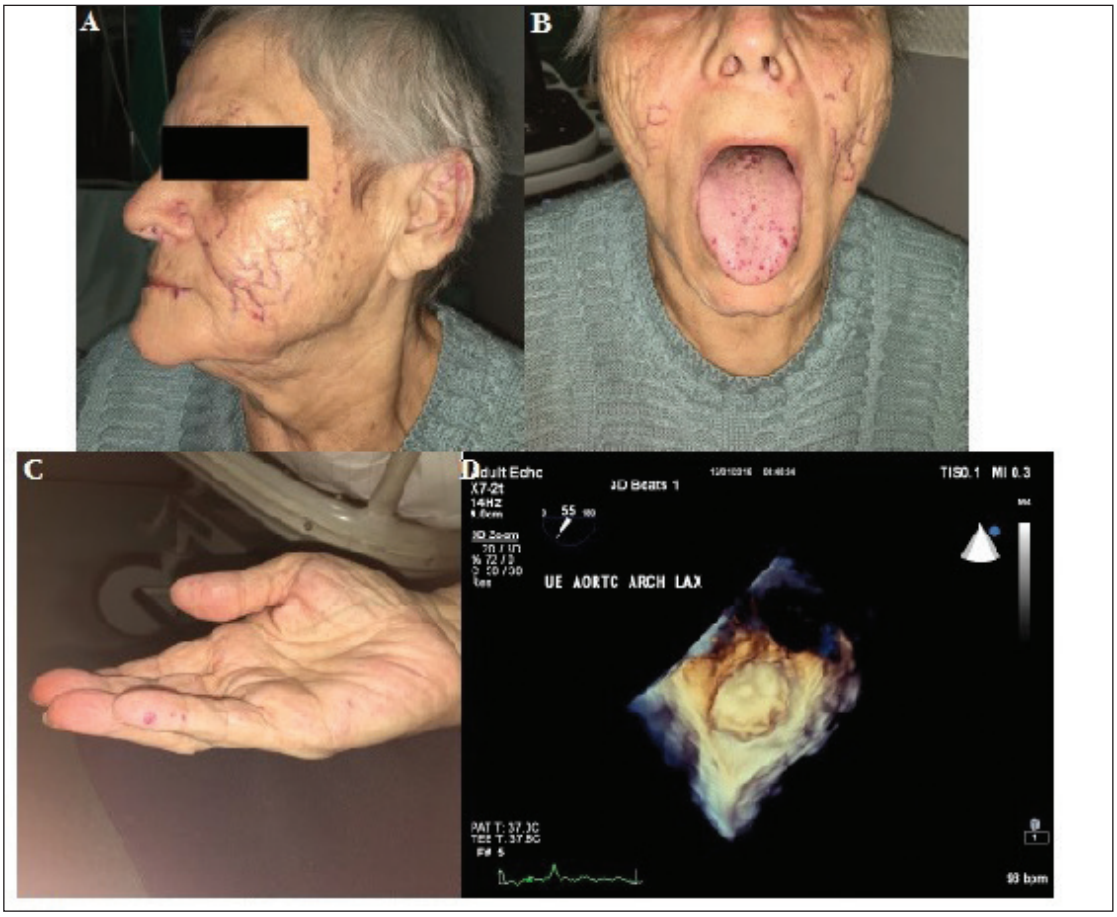

Fig 1: A. Telegiecstasias of face, ears and lips; B. Lesions of face, lips and tongue, C. Telegiecstasias of hand; D. LAA occlusion by Watchman Device 27 mm.

TEE showed proper position of device. In the following 12months antiplatelet therapy was reduced to low-dose aspirin (75 mg), which was well tolerated. In long-term observetion no thromboembolic event, epistaxis or severe bleeding have occurred.

CONCLUSIONS

Osler-Weber-Rendu disease (OWRD) also known as hereditary hemorrhagic telangiectasia (HHT) is a rare autosomal dominant vascular dysplasia which causes telangiectases and arteriovenous malformations of skin, mucosa, and internal organs. The prevalence of OWRD in Europe is 1 case per 5000 to 8000 population and has been estimated to range from 1:200 to 1:100000 worldwide. The clinical diagnosis is based on the Curaçao criteria include epistaxis, telangiectases, visceral lesions and family history (a first-degree relative with HHT). Three or four criteria must be present [6–8]. The most common presentation of OWRD are extensive epistaxis which can lead to severe anemia requiring blood transfusion [9–11].

Atrial fibrillation (AF) is the most common arrhythmia with incidence estimated at 3% of the general population. Thromboembolic events are the most severe complications of this arrhythmia. AF is associated with fivefold higher risk of ischemic stroke [1–3]. Management of AF patients is aimed at prevention of these complications. In order to stratify the individual thromboembolic risk the CHA2DS2-VASc score is used. In patients with high risk of thromboembolism events (a CHA2DS2-VASc score ≥2) the anticoagulation therapy is indicated [12, 13].

Left atrial appendage (LAA) is the main site of thrombi formation. In patients with non-valvular AF over 90% of blood cloths originate from LAA [14, 15]. Non-pharmacological methods of ischemic stroke prevention include surgical exclusion or percutaneous closure of LAA. According to the European Society of Cardiology (ESC) guidelines this procedure may be considered in patients with non-valvular AF, high risk of stroke and contraindications for long-term oral anticoagulation therapy (IIb, level C) [12, 16]. The randomized trials showed that percutaneous LAA closure and VKA treatment (warfarin), were similarly effective in stroke prevention [17–20].

Patients with AF, high risk of stroke and concomitant vascular disorders like OWRD belong to the group at high risk of bleeding complications. This population is extremely problematic with respect to proper anticoagulation - due to complications up to 50% of patients discontinue this therapy [21]. The most commonly used oral anticoagulants are vitamin K antagonist (VKA). Currently there is no data from clinical trials assessing the use of non-vitamin K oral anticoagulants (NOAC) in patients with HTT.

At present there are no recommendations regarding treatment of patients with both HHT and AF. There are no specific guidelines that include the best therapeutic strategy in this population.

The case that we present shows a clinical dilemma. Case reports available via Pubmed report considerable reduction of bleeding complications in patients with AF and HHT who underwent percutaneous LAA occlusion [22, 23]. Further multicenter and randomized clinical trials are required to establish all risks and benefits of LAAC in this group of patients.

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Conflict of interest:

Authors declare no conflict of interest

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