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Kasuistika | Case report

An unexpected complication. First descriptions of intercostal artery rupture in Ehlers-Danlos syndrome

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SOUHRN

Pacient s cévním typem Ehlersova-Danlosova syndromu a s implantovanou mechanickou chlopní byl hospitalizován pro dyspnoe a bolest na hrudi. Po vyloučení infarktu myokardu následovalo vyšetření CT angiografií, které prokázalo spontánní rupturu jedné z mezižeberních tepen s následným vznikem hemothoraxu a rozvojem kompresivní atelektázy. Celý případ komplikovala současná antikoagulační léčba warfarinem po implantaci mechanické chlopně. Po neúspěšné konzervativní léčbě byl případ řešen chirurgicky. Jak ukazuje naše kasuistika, může implantace mechanické chlopně s nezbytnou odpovídající a dlouhodobou antikoagulací warfarinem u těchto pacientů představovat nežádoucí přídatné riziko.

ABSTRACT

A case of Ehlers-Danlos syndrome of vascular type with a history of mechanical valve prosthesis was admitted with dyspnea and chest pain. After exclusion a myocardial infarction, an angio-CT was performed and revealed a spontaneous rupture of an intercostal artery with subsequent hemothorax and compression atelectasis. The case was complicated by the ongoing anticoagulant therapy for the mechanical valve prosthesis with warfarin. After failure of conservative therapy, the case underwent surgical treatment. As shown in this case, mechanical valve prosthesis with necessary adequate anticoagulation, using long lasting warfarin derivates, could be an unneeded added risk for these patients.

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Introduction

Ehlers-Danlos syndrome is a clinically and genetically heterogenous connective tissue disorder, caused by a defect in the synthesis of collagen leading to increased connective tissue elasticity [1]. There are seven types of the syndrome, based on the clinical manifestation and the type of gene defect [2]. Vascular Ehlers-Danlos syndrome, also known as Ehlers-Danlos syndrome type IV, is a life-threatening autosomal dominant type, resulting from mutations in the *COL3A1* gene coding for type III procollagen, leading to severe fragility of connective tissues with possible arterial dissections or rupture [3].

Being prone to spontaneous rupture, the risk of spontaneous bleeding may be complicated if these are subjected to permanent anticoagulation. Here we discuss a case of Ehlers-Danlos syndrome type IV with spontaneous vessel rupture in a patient under anticoagulation due to a history of mechanical valve prosthesis.

The case

A 56-year old patient was presented in the chest pain unit suffering from dyspnea and left sided chest pain. After exclusion of an acute coronary syndrome, meticulous history taking revealed a positive history of Ehlers-Danlos syndrome and a history of Bentall-procedure with mechanical valve prosthesis based on a severe aortic valve insufficiency and an aneurysm of the ascending aorta in 2003. The chest x-ray revealed a new pleural effusion of unknown etiology (Fig. 1). To further evaluate the pleural effusion and to exclude a dissection, an angio CT was performed. The CT (Fig. 2) excluded an aortic dissection, but revealed an extravasation of the contrast agent from the 3rd intercostal artery with the hemothorax compressing the left lung. The case was stabilized by partial release of hemothorax using a chest tube and correction of the coagulation factors using fresh frozen plasma. However, an anticoagulant therapy with heparin had to be resumed to prevent possible prosthetic valve thrombosis. After initial improvement under conservative therapy, the follow-up angio CT showed persistent compression atelectasis of the left lung despite relative improvement of the hemothorax. The lung function tests showed a marked loss of the vital capacity, so that the hemothorax was to be removed surgically. The patient underwent a video-assisted thoracoscopic removal of the pleural hematoma. The patient did well and went back home with stable hemodynamics and no residual loss of vital capacity. Due to the low incidence of intercostals artery rupture and lack of alternative anticoagulation, the patient again was put on warfarin, however with lower target INR of 2–2.5.

Comment

The Ehlers-Danlos syndrome is an inherited disease with no cure but only supportive treatment [2]. The main cause of death in patients with this type is spontaneous rupture of blood vessels due to fragility of the vessel wall [3]. Based on this fact, permanent anticoagulant therapy would complicate any spontaneous bleeding and could lead to a life-threatening condition.

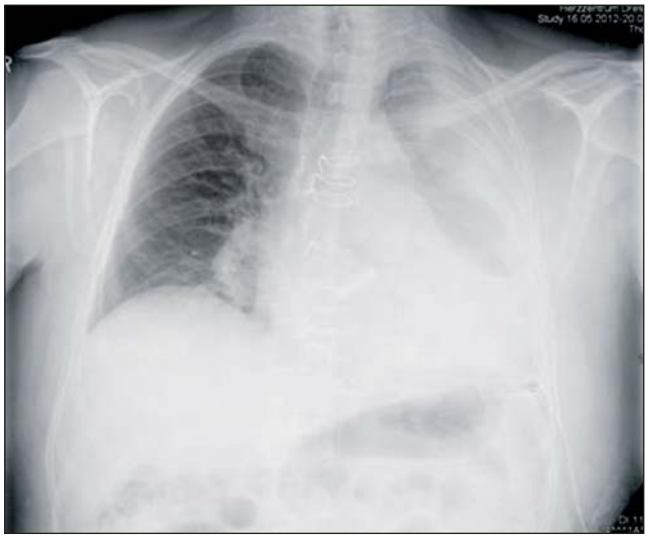

Fig. 1 – X-ray showing a new pleural effusion.

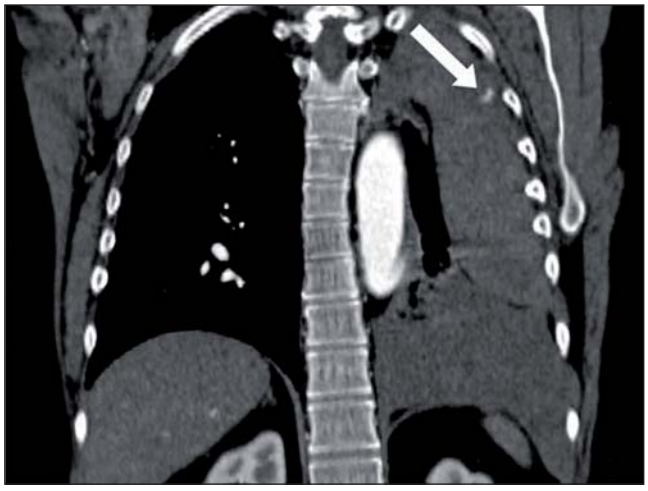

Fig. 2 – An angio CT-image showing extravasation of contrast agent from spontaneous rupture of the intercostal artery (arrow), left sided hemothorax, left-sided atelectasis, and no sign of aortic dissection.

The Ehlers-Danlos syndrome could be associated with valvular diseases. The weakness of connective tissue in the vascular bed in Ehlers-Danlos syndrome is not confined to small and medium-sized vessels, but extends to large arteries, leading it to be one of the causes of aneurysmal formation in the ascending aorta with possible aortic valve insufficiency [4]. The weakness of the connective tissue extends beyond the vascular bed and could even lead to mitral valve prolapse [5].

When a surgical correction of the valvular disease is considered, a valve-sparing procedure should be tried if the valve does not suffer structural damage. If a valve replacement is planned, we think that the bleeding tendency of the Ehlers--Danlos syndrome patients should be considered while choosing the valve prosthesis. As we saw in this case, mechanical valve prosthesis with necessary adequate anticoagulation, using long lasting warfarin derivates, could be an unneeded added risk for these patients.

Conflict of interest
None declared.

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None.

Ethical statement
Authors state that the research was conducted according to ethical standards.

Informed consent
Informed consent was obtained from the patient participating in this study.

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