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A **Lethal Case of Thoracic Aortic Dissection with Bilateral Patellar Subluxation Likely Due To Ehlers-Danlos Syndrome**

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Abstract: A 42-year-old male spontaneously fell down in front of his wife. His past medical history included gastroesophageal reflux disease (GERD) and hypertension without receiving medication. Upon arrival, he was in cardiopulmonary arrest and his initial rhythm was asystole. His chest roentgen revealed a dilated heart and an echocardiogram demonstrated cardiac tamponade. He underwent pericardiocentesis and infusion of 1 mg of adrenalin for three times at 4-minute intervals. Unfortunately, he could not obtain a return of spontaneous circulation. Postmortem computed tomography findings revealed cardiac tamponade, deformity of the contour of the ascending aorta, suggesting Stanford type A aortic dissection, and bilateral patellar subluxation. Bilateral patellar subluxation tends to occur in young females who have an elastic softness. This case was a middle-aged male, thus ligamentous laxity may have existed. Ehlers-Danlos syndrome is a heterogeneous group of heritable connective tissue disorders mainly characterized by joint hypermobility, skin hyperextensibility and tissue fragility. Among congenital or secondary conditions associated with medial degeneration of the aorta, only Ehlers-Danlos syndrome can result in bilateral patellar dislocation. In addition, GERD is common in Ehlers-Danlos syndrome. Accordingly, we strongly suspected that this case was the vascular form Ehlers-Danlos syndrome. A physician should evaluate joint hypermobility or skin hyperextensibility to find causative factor for aortic dissection in young patients.
Keywords: aortic dissection; patellar subluxation; Ehlers-Danlos syndrome

INTRODUCTION

Clinical aortic dissection risk factors fall into two broad categories: conditions that contribute to medial degeneration and those that increase aortic wall stress, such as hypertension [1]. Congenital or secondary conditions associated with medial degeneration include Marfan syndrome, Loeys-Dietz syndrome, the vascular form of Ehlers-Danlos syndrome, inflammatory diseases of the aorta, Turner syndrome, bicuspid aortic valve, familial thoracic aortic aneurysm and dissection syndrome [1]. We herein experienced a case of thoracic aortic dissection with bilateral patellar subluxation likely due to Ehlers-Danlos syndrome and discuss the mechanism of these two combinations.

CASE PRESENTATION

A 42-year-old male spontaneously fell down in front of his wife. When an emergency medical technician checked him, he was in cardiopulmonary arrest (CPA). His initial rhythm had pulseless electrical activity. After receiving instructions over the telephone, he received cardiopulmonary resuscitation by his wife and was subsequently transferred to Numazu City Hospital with 1 mg of adrenalin. His past medical history included gastroesophageal reflux disease (GERD) and hypertension without receiving medication. Upon arrival, he remained in CPA with dilated nonreactive pupils and his initial rhythm was asystole. Immediate tracheal intubation and mechanical ventilation were performed. The results of a venous gas analysis were pH 6.780, PCO_2 112.0 mmHg, PO_2 25.0 mmHg, HCO_3^- 16.3 mmol/l and base excess -18.0 mmol/l. His chest roentgen revealed a dilated heart and an echocardiogram demonstrated cardiac tamponade. He underwent pericardiocentesis and infusion of 1 mg of adrenalin for three times at 4-minute intervals. Unfortunately, he could not obtain a return of spontaneous circulation. Postmortem computed tomography (CT) findings revealed cardiac tamponade, deformity of the contour of the ascending aorta, suggesting Stanford type a aortic dissection (Figure 1), and bilateral patellar subluxation (Figure 2).

The CT reveals cardiac tamponade, deformity of the contour of the ascending aorta, suggesting Stanford type A aortic dissection.

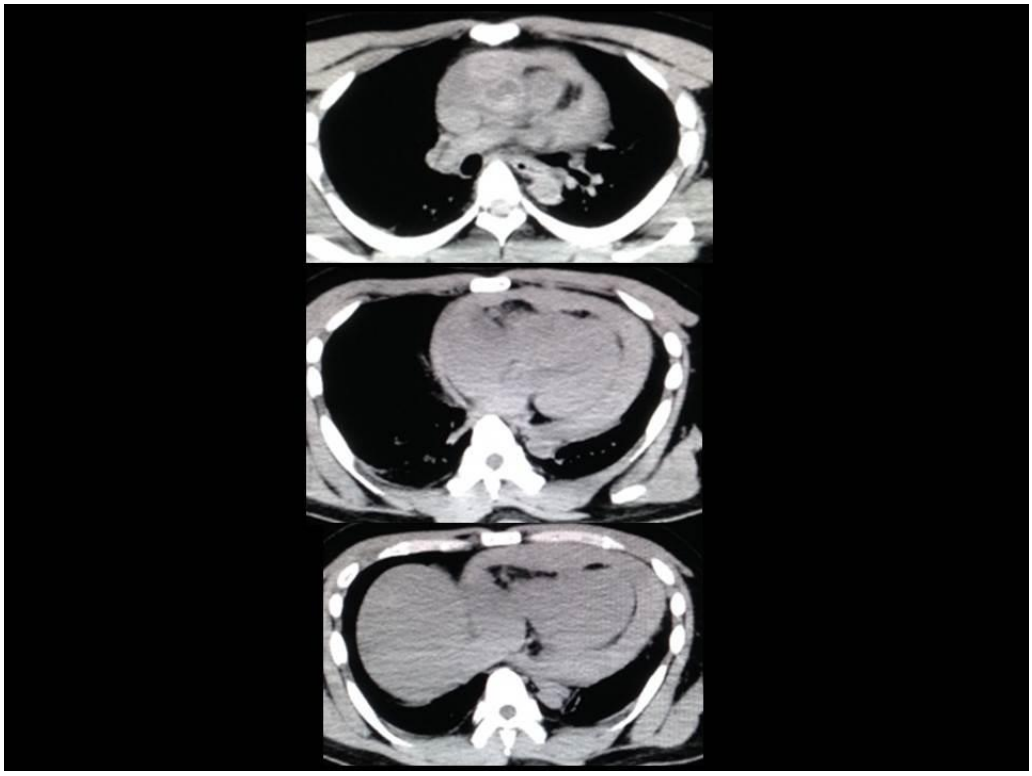

Fig-1: **Postmortem computed tomography (CT) images**

The CT reveals bilateral patellar subluxation.

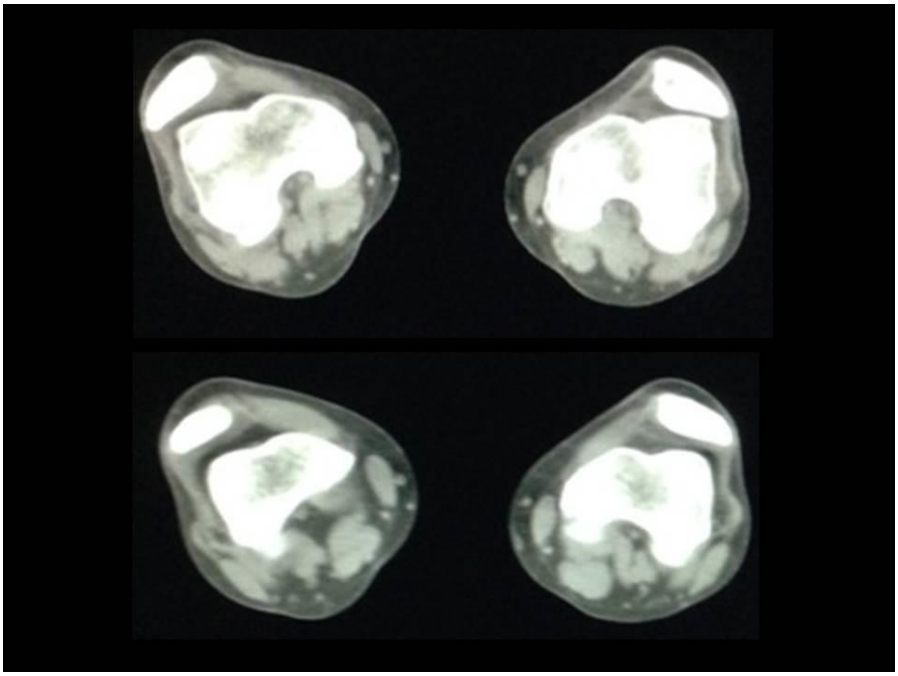

Fig-2: **Postmortem computed tomography (CT) images**

DISCUSSION

Our case was a middle-aged person. Previous data have shown that the average age for the occurrence of dissection is in the 60s [2]. The predominant etiologies for type A dissection in young patients are connective tissue diseases, bicuspid aortic valve, severe hypertension, vascular diseases and cocaine abuse [3]. This patient did not have severe hypertension, vascular diseases or cocaine abuse; therefore, he may have had genetic connective tissue disease.

In general, bilateral patellar subluxation tends to occur in young females who have an elastic softness [4,5]. This case was a middle-aged male, thus

ligamentous laxity may have existed. Ehlers-Danlos syndrome is a heterogeneous group of heritable connective tissue disorders mainly characterized by joint hypermobility, skin hyperextensibility and tissue fragility [5]. Among Marfan syndrome, Loeys-Dietz syndrome, the vascular form of Ehlers-Danlos syndrome, inflammatory diseases of the aorta, Turner syndrome and bicuspid aortic valve, only Ehlers-Danlos syndrome can result in bilateral patellar dislocation [4-7]. In addition, GERD is common in Ehlers-Danlos syndrome, but rare in the other aforementioned diseases [8,9].

Marfan syndrome, Loeys-Dietz syndrome, Turner syndrome or the vascular form of Ehlers-Danlos syndrome has characteristic facial features, however, it is possible that the vascular form of Ehlers-Danlos syndrome without the characteristic facial features may occur [10]. Accordingly, we strongly suspected that this case was the vascular form Ehlers-Danlos syndrome. Unfortunately, we were unable to perform genetic testing for *COL3A1*, which is necessary to make a definitive diagnosis of the disease [11,12].

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Conflicts of interest: All authors do not have any conflicts of interest.

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