CASE IMAGE

An unusual cause of pulmonary arterial hypertension: An acquired extracardiac arteriovenous fistula

Pulmoner arteriyel hipertansiyonun alışılmadık bir nedeni: Edinilmiş ekstrakardiyak arteriyovenöz fistül

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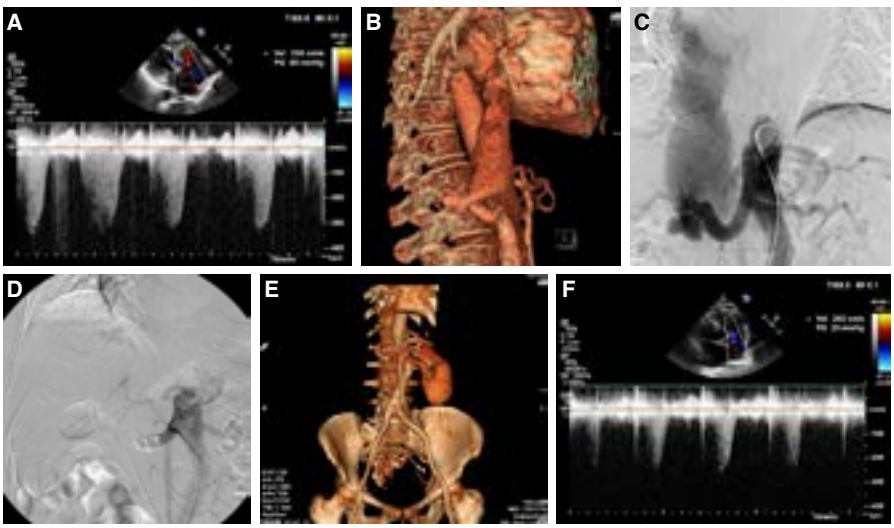
A 68-year-old female patient was referred due to clinical and echocardiographic findings compatible with pulmonary hypertension. Her medical history revealed a unilateral nephrectomy operation 19 years earlier due to a benign mass in the right kidney. The lung fields were clear. A 3/6 loud systolic ejection murmur originating from the upper abdomen, which radiated to the left parasternal border was discerned. Two-dimensional echocardiography revealed normal-sized left heart chambers with an ejection fraction of 60% and no significant left-sided valvular disease. The right ventricle and right atrium were slightly dilated with mild-to-moderate tricuspid valve regurgitation. A dilated (24 mm) non-collapsing inferior vena cava and severe pulmonary hypertension (60 mm Hg) were also noted. (Fig. A, Video 1*) An intracardiac shunt was not detected via transesophageal echocardiography. An abdominal computed tomography (CT) showed a tortious aortocaval fistula originating from the remnant of the right renal artery (Fig. B). It was thought that this arteriovenous fistula (AVF) was probably linked to vascular injury dating to the nephrectomy performed 19 years prior. On cardiac catheterization, the pulmonary capillary wedge pressure and mean pulmonary artery pressure (PAP) were determined to be 5 mmHg and 33 mmHg, respectively. Using the indirect Fick method, pulmonary flow was calculated at 12.7 L/minute, which yielded a pulmonary vascular resistance of 176.8 dynes/second/cm^{-5} (2.2 Wood units). The tract of the aortocaval fistula was visualized with aortography (Fig. C, Video 2*) and subsequently closed using one 16 mm Amplatzer vascular plug I and one 16 mm Amplatzer Vascular Plus II (St Jude Medical, Inc., St. Paul, MN, USA) (Fig. D, Video 3*). A repeat abdominal CT image obtained before discharge did not demonstrate any residual flow in the AVF (Fig. E). At 3 months, the patient was free of symptoms and a follow-up transthoracic echocardiography at 3 months showed normalization of estimated systolic PAP (28 mm Hg) (Fig. F, Video 4*).



Figures– Echocardiographic and angiographic findings of the patient before closure of the defect. **(A)** High tricuspid regurgitation velocity (3.4 m/v) with estimated systolic pulmonary artery pressure of 60 mm Hg (45+15 mm Hg) obtained at presentation of the patient. **(B, C)** Show computed tomography and invasive angiographic views of the arteriovenous fistula originating from the stump of the renal artery and draining to the inferior vena cava after a tortious course. Echocardiographic and angiographic findings of the patient following closure of the defect. **(D)** After insertion of 2 coils on the remnant of the right renal artery, no residual flow was observed in the fistula. **(E)** An abdominal computed tomography image obtained before hospital discharge indicated a total disappearance of the fistula. **(F)** Three months after the procedure, tricuspid regurgitation velocity dropped below 2.8 m/s, with disappearance of the symptoms.

*Supplementary video files associated with this presentation can be found in the online version of the journal.