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Blue toe syndrome following modified Bentall procedure: a case report

Modifiye Bentall işlemi sonrası görülen mavi parmak sendromu: Olgu sunumu

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Cholesterol crystal emboli syndrome, also named "blue toe syndrome" can be rarely seen following heart surgery and invasive cardiology procedures. A 64-year-old man was operated on for ascending aortic aneurysm and severe aortic regurgitation. On the third postoperative day he complained of his cold, pale toes with severe pain. Lower extremity arterial Doppler ultrasonography examination showed a triphasic flow pattern in all segments. Complaints of the patient increased on the following day and cyanosis began to be evident on the distal end of his toes. Oral anticoagulation therapy was continued because of the new aortic prosthesis, and clinical signs recovered spontaneously. The patient has had no ischemic symptoms in the last six months.

Key words: Bentall procedure; blue toe syndrome; cholesterol crystal embolism.

Blue toe syndrome, also known as purple toe syndrome or cholesterol crystal emboli (CCE) syndrome, is a systemic disorder due to an embolization of atheromatous material from ulcerated atherosclerotic plaques in the aorta and its major branches. The skin and the kidneys are most frequently involved, but any organ can be affected. Livedo reticularis of the lower extremities and acrocyanosis are the most common cutaneous manifestations.^[1-3]

CASE REPORT

A 64-year-old man was admitted to our hospital with chest pain, shortness of breath and weakness. He had hypertension and dyslipidemia. At physical examination, a holodiastolic murmur was heard along the left sternal border. Pulse pressure was widened. Electrocardiography (ECG) and chest X-ray show left ventricular enlargement. Transthoracic echocardiography showed an ascending aortic aneurysm, severe aortic regurgitation, and a mildly Mavi parmak sendromu olarak da adlandırılan kolesterol kristal emboli sendromu kalp cerrahisi ya da invaziv kardiyolojik girişimlerden sonra nadiren görülür. Altmış dört yaşındaki erkek hasta çıkan aort anevrizması ve ciddi aort yetmezliği nedeniyle ameliyat edildi. Ameliyat sonrası üçüncü gün şiddetli ağrı ile ayak parmaklarında soğuma ve solukluk yakınması oldu. Alt ekstremite arteryel Doppler ultrason incelemesinde bütün segmentlerde üçlü akım paterni saptandı. Sonraki gün hastanın yakınmaları arttı ve ayak parmaklarının uç kısmında belirgin bir şekilde siyanoz başladı. Yeni protez aort kapak nedeniyle oral antikoagülasyon tedavisine devam edilen hastanın klinik bulguları süratle düzeldi. Hastada son altı ay içinde herhangi bir iskemik semptoma rastlanmadı.

Anahtar sözcükler: Bentall işlemi; mavi parmak sendromu; kolesterol kristal embolisi.

dilated left ventricle. Coronary angiography revealed normal coronary arteries.

Operation was performed through median sternotomy. Right femoral artery and two stage venous cannulation was performed under moderate (28 °C) hypothermia. After aortic cross clamping, myocardial protection was performed using combined antegrade/retrograde blood cardioplegia. The ascending aorta was dilated and its inner surface was normal. We replaced the ascending aorta with a 25 mm valved composite graft.

The patient was discharged from the intensive care unit on his first day after surgery. On the third day we noticed cold, pale toes with severe pain. Lower extremity arterial Doppler ultrasonography examination showed a triphasic flow pattern in all segments.

We supposed that it could be cholesterol crystal emboli. His complaints increased on the following

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Fig. 1. Blue toe is seen in the picture which was taken on the third postoperative day.

day and cyanosis began to be evident on his toes (Figure 1). Blood eosinophil and plasma creatinine levels increased, renal replacement therapy was not required. We carried on oral anticoagulation for his new aortic prosthesis. Clinical signs recovered spontaneously. The patient was discharged on the 13th day postoperatively. The patient has had no ischemic symptoms in the last six months.

DISCUSSION

Cholesterol crystal embolization may occur spontaneously, and can also be seen after cardiovascular surgery, percutanous coronary interventions, and in some cases following anticoagulant or thrombolytic treatments, or both.^[1-2] The term "atherombotic microembolism" is also used to refer to the dislodgement of vascular plaque material that contains cholesterol crystals plus red blood cells and fibrin. These can occlude major systemic vessels and result in organ infarction.^[3] Cholesterol crystal emboli have a wide prognostic spectrum. The prognosis depends on the extent of the systemic disease and a high rate of mortality can be observed. Risk factors for CCE are old age, peripheral vascular disease and severe atherosclerosis of the ascending aorta.^[3]

The onset of clinical symptoms and signs of CCE syndrome are variable, and in part depends on the mechanism for cholesterol release. Early symptoms are usually relatively rapid after physical dislodgement: days to weeks. The triad of pain, blue toe and intact peripheral pulses is pathognomic for CCE. Diagnosis can be made with skin biopsies, but these may also be normal.^[4]

The kidneys are frequently affected by cholesterol emboli because of the proximity of the renal arteries to the abdominal aorta and also because of the enormous amount of blood flow they have.^[5]

The management of blue toe syndrome is initially supportive. Anticoagulation is controversial, because thrombolytic therapy and anticoagulants appear to precipitate cholesterol emboli by dissolving protective thrombi and fibrin deposits coating an atheromatous plaque, permitting the release of cholesterol.^[5] Carrying on anticoagulation orally for his new aortic prosthesis was essential in our patient. If anticoagulation is necessary, warfarin can be used safely. Even though cyclophosphamide and corticosteroids are recommended in the case of renal insufficiency, there is still no specific treatment for CCE.^[6]

In patients with diffuse atherosclerosis, surgical strategies like off-pump surgery, total circulatory arrest, and clamping techniques are important during the operation to prevent this undesired event.

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