# Rupture of Isolated Abdominal Aortic Aneurysm and Dissection in a Patient With Marfan Syndrome

## *MARFAN SENDROMLU BİR OLGUDA İZOLE DİSSEKAN ABDOMİNAL AORT ANEVRİZMA RÜPTÜRÜ*

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**Abstract**

Marfan syndrome is a dominantly inherited connective tissue disorder characterized by cardiovascular, skeletal and ocular manifestations. Aortic aneurysm and dissection in Marfan syndrome are commonly found in the thoracic part of the aorta. Whereas, abdominal aortic involvement is very rare. We, hereby, present a Marfan's patient who presented with rupture of aneurysm and dissection involving the abdominal aorta from the level of the renal arteries to the right iliac artery. The patient was successfully operated and a week later was discharged. Careful examination and serial imaging studies are essential in detecting new aneurysms or dissections at different sites of the aorta

**Key Words:** Marfan Syndrome; aorta, abdominal; aortic rupture

**Özet**

Marfan sendromu kardiyovasküler sistem, iskelet ve göz tutulumu ile karakterize dominant geçişli bir konnektif doku hastalığıdır. Marfan sendromlu hastalarda, aort anevrizması ve disseksiyonu en sıklıkla torasik aortada bulunur. Abdominal aort tutulumu ise oldukça seyrektir. Biz burada, Marfan sendromlu bir hastada abdominal aortada renal arterlerin distalinden başlayıp sağ iliyak artere uzanan rüptüre anevrizma ve diseksiyon olgusunu sunuyoruz. Hasta başarılı bir şekilde ameliyat edilip 1 hafta sonra taburcu edildi. Dikkatli fizik muayene ve seri radyolojik tetkikler farklı lokalizasyonlardaki yeni başlangıçlı anevrizmaların ve disseksiyonların tespiti için gereklidir.

**Anahtar Kelimeler:** Marfan sendromu; abdominal aorta; aortik rüptür

**Turkiye Klinikleri J Cardiovasc Sci 2007, 19:95-97**

Marfan syndrome (MFS) is an inherited connective tissue disorder transmitted as an autosomal dominant trait with a prevalence of 1 in 3000 to 5000 individuals.[1] The most common cardiovascular complications seen in patients with MFS are thoracic aortic aneurysm, dissection, and aortic rupture.[2-4] Isolated abdominal aortic aneurysm and dissection are very rare in patients with MFS. We report a case of a 30 year old man with MFS who was admitted with sudden aortic rupture secondary to isolated abdominal aortic aneurysm and dissection for which he was operated successfully.

**Geliş Tarihi/*Received:*** 26.10.2006 **Kabul Tarihi/*Accepted:*** 25.01.2007

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## Case Report

A thirty year old male patient was referred to the emergency unit of our hospital with complaints of sudden onset abdominal pain emerged during an effort to lift a heavy tree-stump. The patient has been followed up with the diagnoses of MFS and mild mental retardation since he was 5 years old. Physical examination findings revealed poor general status, open conscious, cooperating, blood pressure: 60 /40 mmHg, heart rate: 120 / min, body temperature: 36.7°C. A dissecting aortic aneurysm, reaching 5.5 cm at its widest site, beginning from the level of renal artery and extending to the right iliac artery was detected in thoracoabdominal CT examination (Figure 1, 2a). A periaortic hemorrhage secondary to rupture was also detected (Figure 2b). Heart valves and the ascending aorta were determined to be normal with echocardiography. Physical examination revealed high palate and extremity abnormalities. The patient was 198 cm in

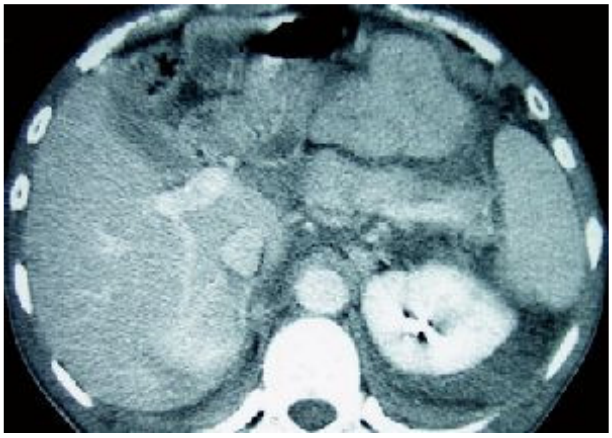


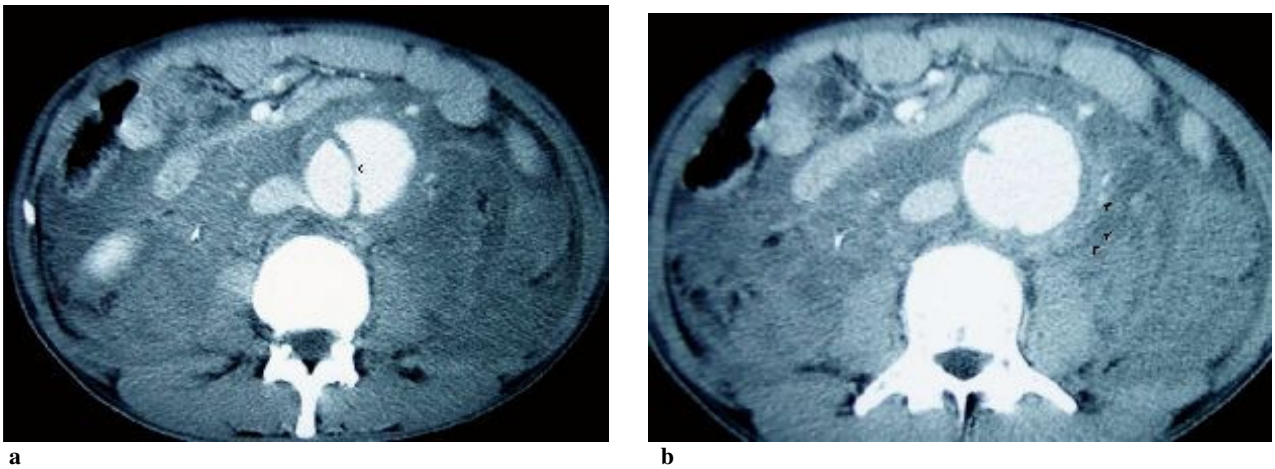
**Figure 1.** Axial CT image shows normal suprarenal abdominal aorta.

height and had no characteristic findings related to MFS in the family history. Patient was urgently operated with the diagnosis of "rupture of dissecting aneurysm in the abdominal aorta/right iliac artery". The patient was operated by transmedian laparatomy approach where the aneurysmatic abdominal aorta was reached. A wide retroperitoneal hematoma was detected during operation. A cross clamp was applied below the renal arteries. There was a rupture in the posterolateral portion of the aorta of about 2 cm below the renal arteries. In addition to aneurysm a dissection extending into the iliac artery was present. A bifurcated (16 x 8 mm) dacron graft was used. The proximal anastomosis was carried out using running 4/0 polypropylene suture. Teflon felts were used to reinforce the anastomosis. After the proximal anastomosis was constructed, the distal anastomosis was performed to the iliac artery bifurcations with end to end fashion. Finally, the inferior mesenteric artery was reanastomosed to the dacron graft. No additional problems were seen in the intensive care unit and the service during the postoperative period. Patient was discharged on 7$^{th}$ day. In the examination of pathological specimen, hyaline degeneration was found in the wall of the aorta. In the following 1 year the patient had no complaint.

## Discussion

Dilatation, dissection or rupture of the proximal aorta are a typical characteristic feature of MFS. Untreated cardiovascular complications are the major cause of death in the first 4 decades of life in MFS. Elongation of the lifetime is depended on the control or prevention of the cardiovascular complications.[1]

MFS is an autosomal dominant disease resulting from a mutation in the fibrillin gene located on chromosome 15 and leads to a connective tissue disorder characterized by cardiovascular, skeletal, and ocular abnormalities.[5-7] The major cause of morbidity and mortality in these patients is the dilatation of the aortic root. Aortic failure is a complication that develops secondary to the dilated



**a** **b**

**Figure 2a, 2b.** Axial CT images show dissection (arrow) and ruptured abdominal aortic aneurysm. Notice periaortic hemorrhage (arrowheads).

aortic root. In addition to the pathologies of the aorta, the most frequently observed cardiac involvement, is that of the mitral valve. Rarely, the aneurysms of the coronary, axillary, and subclavian arteries have been reported.[8]

The typical histological finding observed in the aorta in MFS is cystic medial necrosis which may also be seen in other connective tissue disorders. In this condition, there is degeneration in the elastic fibers of the media. Advanced subintimal thickening, hyaline degeneration and fibromuscular proliferation with occasional mixoid degeneration, in addition to degeneration of the muscular fibers due to significant fibrosis was observed in the aortic wall in the preparations of all the pathological material we obtained.

MFS involves a wider portion of the aorta to include both the thoracic and abdominal parts, as compared to the degenerative thoracoabdominal aneurysms (27-50%) (Crawford type 2). However, isolated aortic involvement (Crawford type 4), is observed in 18% of patients with MFS. Aortic dissection is observed in 21-82% of these patients.[4,5] Aoimi S et al, identified isolated aortic aneurism in only 2 of 141 Marfan's patients with aortic involvement.[9] We did not find any case of rupture secondary to the aneurysm and dissection of the abdominal aorta and right iliac artery in the literature searching of patients with MFS.

Surgical therapy is generally accepted to be performed in aneurysms exceeding 5-5.5 cm in diameter. However, since there is a risk of rupture in aneurysms of smaller diameter in MFS, surgical treatment is recommended in aneurysms larger than or equal to 4.5 cm in diameter. Surgical treatment is satisfactory in these patients and mortality rate is lower. However, recurrence rate after the surgery is high, therefore regular control with tomography or magnetic resonance at 6-month intervals is mandatory.

## Conclusion

Presentation with sudden rupture secondary to dissection is common in MFS patients with cardiovascular involvement. Therefore, these patients should be closely followed up and surgical treatment should be performed before the development of any complication. In addition to close follow up, counseling the family and recruitment of the proper medical treatment modality shall reduce the rate of complications due to cardiovascular involvement.

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