Rare Manifestation of Abdominal Aortic Aneurysm and Popliteal Aneurysm in a Patient with Marfan's Syndrome

Wolfgarten, Bernd
Kruger, I.
Gawenda, M.

Vascular Surgery; Jan/Feb2001, Vol. 35 Issue 1, p81, 4p, 4 bw

Aneurysmatic dilatation of the left popliteal artery; Primary location of aneurysmatic dilatations; Diagnosis of Marfan's syndrome.

1535

00422835

4063549


Biomedical Reference Collection: Comprehensive
RARE MANIFESTATION OF ABDOMINAL AORTIC ANEURYSM AND POPLITEAL ANEURYSM IN A PATIENT WITH MARFAN'S SYNDROME

Introduction
Marfan's syndrome is an autosomal dominant disease with high penetration and variable expression. Prevalence of the disorder lies between 40 and 60 per one million population. The diagnosis is based on results of family history and clinical examination. The skeletal system, eye, and cardiovascular systems are commonly affected. In the extreme range of clinical findings, a typical case involves somatomegaly, arachnodactyly, aortic dissection and aneurysm, and lens dislocation. The genetic origin of Marfan's syndrome appears to be located on chromosome 15. Histologic findings show a mucoid degeneration of the media accompanied by the destruction of elastic and collagenous fibers and possible necrosis of the smooth musculature. Aneurysmatic dilatations are primarily located in sections I and II of the thoracic aorta, whereas aneurysms in sections IV and V are much rarer. To our knowledge, only 30 cases of Marfan's syndrome involving abdominal aortic aneurysms have been published. The rare combination of an abdominal aortic aneurysm and left-sided popliteal aneurysm accompanying Marfan's syndrome is presented.

Case Report
A 37-year-old patient with back pain and somatomegaly was found to have a penetrating aneurysm of sections IV and V of the abdominal aorta. Results of a family history and clinical examination confirmed suspicions of Marfan's syndrome. Further angiologic studies depicted an aneurysmatic dilatation of the left popliteal artery. Aneurysmatic dilatations are primarily located in sections I and II of the thoracic aorta, whereas aneurysms in sections IV and V are much rarer. The rare combination of an abdominal aortic aneurysm and left-sided popliteal aneurysm accompanying Marfan's syndrome is presented.

Discussion
The diagnosis of Marfan's syndrome is primarily based on clinical manifestations, which have an extraordinary degree of variation. Chromosome analyses of gene defects on chromosome 15 and histologic investigations of cystic media necrosis (Erdeghm Gsell) continue to be less reliable. Considering the life expectancy of these patients, an early diagnosis is extremely important to prevent typical complications. Cardiovascular complications, which in 30% to 60% of the cases lead to a reduction in life expectancy, are of special relevance.

In larger studies, a mean life expectancy was determined to be 32 years. Less than 10% of the patients with...
cardiovascular manifestations aged more than 40 years.[8] The cause of the life-threatening condition is a cystic media necrosis, a non-arteriosclerotic vascular disease that can lead to dissecting aortic aneurysms.[4, 9-11]

Figure 3 shows a complete occlusion of the distal superficial femoral artery and indicates a combination of occlusive and aneurysmal disease, which can occur in Marfan’s syndrome. The aneurysms can be found in segments I and II and are often accompanied by a simultaneous aortic insufficiency. There are two reasons for this location. First, in the ascending aorta there are twice as many elastic fibers as in the abdominal aorta. Second, the thoracic aorta is subject to a much larger degree of pressure fluctuation and, in accordance with Laplace’s law, the larger diameter of the thoracic portion leads to a greater pull on the wall of this segment.[12] In contrast, abdominal aneurysms are rare,[2, 9, 13] especially those in the peripheral arteries among patients with Marfan’s syndrome.[14-16]

Despite the family history and typical Marfan’s syndrome habitus of our patient, this condition had not been diagnosed. Considering the vulnerability of the vascular wall damaged by media necrosis, the indication is given for a rapid bypass of the aneurysmatic vessel. In our case, the increased fragility of the aorta necessitated a two-cavity procedure. To prevent relapse, tri-annual examinations are justified. Ultimately, surgery is the only possibility for the increase in life expectancy of patients with Marfan’s syndrome.

Conclusion
Aortic dissection and aneurysms of the thoracic aorta are typical vascular manifestations of Marfan’s syndrome. Abdominal and peripheral vascular aneurysms are rare but possible manifestations. Angiologic diagnostics encompassing the peripheral arteries are indicated to detect multifocal manifestations of the disease in the vascular system.

Vascular Surgery 35:81-84, 2001

From the Department of Visceral and Vascular Surgery, University of Cologne, Germany

Correspondence: Bernd Wolfgarten, MD, c/o St. Vinzenz-Hospital Köln, Merheimer Str. 221-223, D-50733 Köln, Germany

(C) 2001 Westminster Publications, Inc., 708 Glen Cove Avenue, Glen Head, NY 11545, USA

PHOTO (BLACK & WHITE): Figure 1. Marfan’s syndrome with arachnodactyly and pronounced stem and lateral branch varicosis.

PHOTO (BLACK & WHITE): Figure 2. Intraarterial angiography of the aorta thoracoabdominalis.

PHOTO (BLACK & WHITE): Figure 3. Intraarterial angiography of the lower appendage shows an aneurysm in the left popliteal artery and an occlusion of the distal superficial femoral artery.

PHOTO (BLACK & WHITE): Figure 4. Procedure according to Crawford. Intraoperative situs with implantation of the 22-mm coated Dacron tubular prosthesis.

REFERENCES


~~~

By Bernd Wolfgarten, MD; I. Kruger, MD and M. Gawenda, MD, Cologne, Germany

Adapted by MD, MD and MD, Cologne, Germany

Copyright of Vascular Surgery is the property of Sage Publications, Inc. and its content may not be copied or emailed to multiple sites or posted to a listserv without the copyright holder's express written permission. However, users may print, download, or email articles for individual use.