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*Giant aortic aneurysm and rhabdomyomas in infant with tuberous sclerosis (Case report)**

Emilia Patiño Bahena,* Juan Calderón-Colmenero,* Alfonso Buendía,* Antonio Juanico*

Summary

We present the case of a giant aneurysm and dissection of the abdominal aorta in an eight month old infant. Imaging studies especially a helicoid computerized axial tomography with 3D reconstruction, showed a lesion which extended from the diaphragm to the iliac arteries. The clinical findings and the above mentioned studies revealed non-obstructive heart rhabdomyomas, tubers in the brain and the abdominal aortic aneurysm, all of which were consistent with the diagnosis of tuberous sclerosis.

Resumen

ANEURISMA GIGANTE DE LA AORTA ABDOMINAL Y
RABDOMIOMAS EN UN LACTANTE CON ESCLEROSIS
TUBEROSA

Presentamos el caso, de un niño de 8 meses con un aneurisma gigante de la aorta abdominal. La imagen de la tomografía axial computarizada helicoidal con reconstrucción tridimensional, mostró que dicha lesión iniciaba a nivel del diafragma y llegaba hasta las arterias ilíacas con disección de su pared. El cuadro clínico y los estudios apoyaron el diagnóstico de esclerosis tuberosa con rabdomiomas cardíacos, no obstructivos. Por la gravedad de dicha lesión, el desenlace fue fatal.
(Arch Cardiol Mex 2005; 75: 448-450)

Key words: Aortic aneurysm. Rhabdomyomas. Tuberous sclerosis.

Palabras clave: Aneurisma aórtico. Rabdomiomas. Esclerosis tuberosa.

Introduction

Aortic aneurysms in pediatric patients are rare lesions related to illnesses of the aorta, as in Marfan and Ehlers-Danlos syndromes, or of inflammatory conditions, such as secondary mycotic aneurysms. They usually involve the thoracic aorta which in some cases are the result of interventionist catheterization procedures.¹

There are abdominal aortic aneurysms with a history of catheterization of the umbilical artery

in neonates.² Aneurysms of the abdominal aorta in tuberous sclerosis have been reported since 1971, usually in school-age children.³ We present a case of early onset of various manifestations of this condition.

Case summary

An 8-month-old male was seen in our institution with a history of generalized tonic seizures at age 4 months, which alternated with myoclonic spasms. He also exhibited retarded psychomotor de-

* Pediatric Cardiology Department, Instituto Nacional de Cardiología "Ignacio Chávez".

Address correspondence to: Emilia Patiño Bahena. (INCICH, Juan Badiano Núm. 1 Tlalpan Sección XVI, 14080 México D.F.) Tel: México (55) 55-73-29-11 1336 Fax (55) 54-85-15-68. E-mail: emjopaba@cardiologia.org.mx

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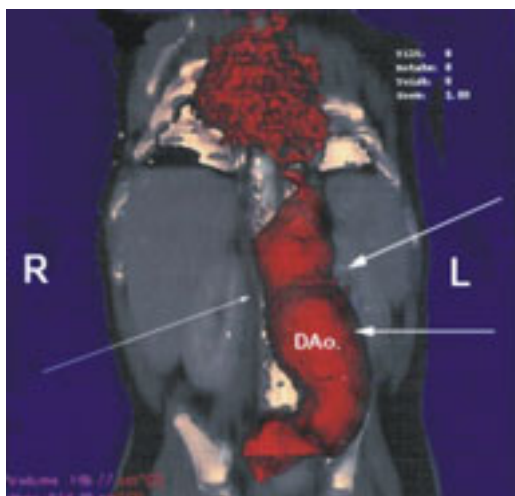


Fig. 1. Anteroposterior view of contrast-enhanced, 3D abdominal computerized axial angio-tomography which shows a long fusiform aneurysm of the aorta (arrows).

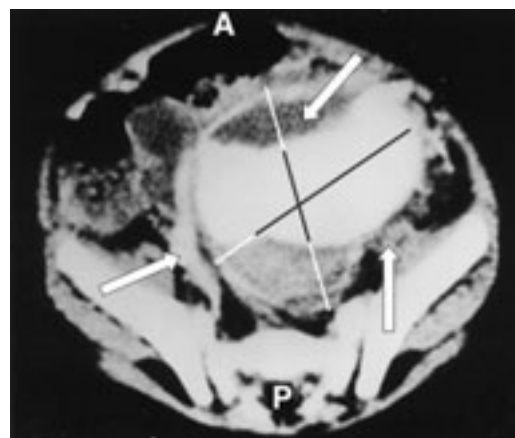


Fig. 2. Contrast-enhanced computerized axial tomography which shows the abdominal aorta and an aneurysm measuring of 6 x 7 cm. A thrombus is seen in the anterior and posterior walls (arrows).

velopment. At age 7 months, an abdominal mass was detected clinically. Physical examination. The child appeared pale; the heart sounds were normal. A firm, irregular, pulsating mass extending from the hypochondria to the iliac fossa was palpated on the left side of the abdomen.

An echocardiogram revealed the presence of three tumors; one apical mass 4 mm in diameter in the left ventricle and two in the right ventricle. One of the right ventricular tumors was located in the anterior wall close to the tricuspid valve, and the other one was on the right septum; none were obstructive. A computerized axial tomography scan of the skull showed tubers in the wall of the right lateral ventricle.

Contrast-enhanced, 3D abdominal computerized axial angiotomography corroborated the presence of a fusiform aneurysm of the aorta, extending 13.5 cm from the diaphragm to the iliac arteries with a thrombus in its distal portion, near the iliac vessels. It involved the renal and mesenteric arteries, and dissected the aortic wall, clearly apparent in a computerized axial tomography at the abdominal level; there an aortic dissection a thrombus in the abdominal aortic wall was also seen (*Figs. 1 and 2*). The patient died suddenly as a result of the dissection of the aorta.

Discussion

This case shows an early onset of tuberous sclerosis, with lesions in different areas: the heart

exhibited tumors in both ventricles, the brain had tubers; the abdominal aorta showed a giant aneurysm and aortic dissection which caused the death of the patient. The possibility of surgical treatment of the aortic lesion was suggested; however, dissection of the aorta precluded this intervention. Bavdekar et al reported a similar case in a 6-year-old child with a toracoabdominal aneurysm without rhabdomyomas, who was successfully operated. They identified increased collagen content, especially in the elastic fibers, in the media of the aorta. They suggested observation of these patients and tracking this complication with serial abdominal ultrasounds.^{4,5}

Tuberous sclerosis can affect the skin, brain, kidney, heart, retina, and pancreas; it is a heritable condition in an autosomal dominant manner with variable entrance. The loci that underlie it have been identified on two chromosomes, one on chromosom 9 (9q34) that codes for hamartin and the other at 16 p13.3, which codes for tuberlin.⁶

Cardiac involvement in this syndrome varies; 40 percent of the tumors are benign;^{7,9} when they obstruct cardiac cavities or valves, they are amenable to surgical treatment.

Figures 1 and 2 show computerized axial tomography images with 3D reconstruction of our patient with tuberous sclerosis exhibiting an infrequent complication, i.e., an aneurysm and dissection of the aorta.

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