We describe a previously healthy 48-year-old man who presented with clinical characteristics suggestive of internal carotid artery dissection, confirmed by magnetic resonance imaging. He developed a massive infarction of the left cerebral hemisphere and died after 3 days of transtentorial herniation. Post-mortem examination identified a dissection of the thoracic aorta caused by Erdheim-Gsell cystic medionecrosis, with the characteristic degeneration of the elastic fibers of the lamina media. The dissection showed an unusually large extension not only distally into both iliac arteries, but also proximally into both carotid arteries. To our knowledge, such an extensive dissection has not been described previously. Underlying vessel wall disorders of the aorta, such as Erdheim-Gsell cystic medionecrosis, should be considered in young patients with spontaneous arterial dissection.

Although ischemic stroke is more common with increasing age, it is certainly not rare in young adults (ie, <50 years). Estimates of its incidence in patients aged 15 to 39 years, for example, range from 6 per 100,000 in whites to 15 per 100,000 in blacks.1 In this case report, we describe a 48-year-old male patient with a massive infarction of the left cerebral hemisphere caused by a rather uncommon nonatherosclerotic vasculopathy that has received little attention in the neurological literature.

REPORT OF A CASE

On January 18, 1997, a previously healthy, nonsmoking 48-year-old male patient was admitted to our hospital. A few hours before admission, he experienced acute pain on the left side of the neck while he was exercising on a running belt. Thirty minutes later, he experienced failing vision in his left eye, followed shortly thereafter by the loss of muscle strength in his right arm and expressive dysphasia. There was complete restoration of neurological function after approximately 30 minutes. The patient's family history was negative for vascular events. General examination on admission showed blood pressure of 140/80 mm Hg; auscultation revealed normal heart and lung sounds. Results of routine hematologic and biochemical blood investigation were unremarkable. Computed tomography of the brain showed no abnormalities. One hour after admission, the patient suddenly developed mixed aphasia with flaccid hemiparalysis of the right side and an ipsilateral Babinski sign. Subsequent magnetic resonance imaging revealed a thrombus in the left internal carotid artery with a large arteria cerebri media infarction. No anticoagulant treatment was started because of the extent of the infarction. Three days later, he died with the clinical symptoms of a transtentorial herniation.

Autopsy showed a dissection of the thoracic aorta, with a tear of about 1.5 cm just below the opening of the left subclavian artery that continued distally into the abdominal aorta and both iliac arteries and proximally into both internal carotid arteries. At the level of the left carotid bifurcation, there was almost complete obstruction of the internal carotid artery caused by a thrombus in the dissected vessel wall.
More distally into the left internal carotid artery, continuing into the left middle cerebral artery, a thrombus completely occluded the vessel lumen. The left cerebral hemisphere was edematous, with signs of transtentorial herniation. Microscopic examination of the aortic tissue and other larger arteries showed elaborate degeneration of the elastic fibers in the media that was compatible with the histopathological diagnosis of Erdheim-Gsell cystic medionecrosis (Figure).

In other parts of the arteries that were investigated, mild atherosclerotic changes were observed that were compatible with the age of the patient. Besides minor steatosis hepatis, no other abnormalities were detected.

**COMMENT**

Our patient presented with several clinical characteristics suggestive of internal carotid artery dissection (eg, left-sided neck pain, followed after several hours by transient ischemia in the ipsilateral territory of the ophthalmic artery and eventually an ischemic stroke in the left medial cerebral artery). Magnetic resonance imaging confirmed the presence of left internal carotid artery dissection. In our patient, a dissection in the thoracic aorta caused by Erdheim-Gsell cystic medionecrosis was found with an unusually large extension proximally into both carotid arteries and distally into both iliac arteries. To our knowledge, such an extensive dissection has not been described previously.

Erdheim-Gsell segmental cystic medionecrosis is presumably a rare cause of dissection and cerebral infarction in young adults. This histopathological entity was introduced by Gsell² in 1928 and Erdheim³ in 1929; they described necrosis of the smooth musculature of the lamina media (Gsell’s principle) and the mucoid degeneration of the lamina elastica and propria (Erdheim’s principle) in pathological aortas. Although cystic medionecrosis is widely recognized, the term is now considered to describe only the mucoid (ie, cystic) degeneration. In these cystic areas, there is extensive pooling of acid mucopolysaccharide material, leading to a severe diminution of the vessel wall strength. In the noncystic areas, there is only minimal degeneration of the elastic fibers. The reduced vessel wall strength may subsequently allow irritation of blood into the vascular wall, with advancement along specific planes of cleavage, causing constriction of the arterial lumen and thereby hampering the blood flow.

The aorta is the usual site of the dissecting aneurysms, and these lesions may extend into the major branches of this vessel⁴,⁵ (eg, into the splenic⁶,⁷ or femoral⁸ artery). In addition, next to these distally extending dissections, as shown by our patient, dissections originating primarily within the thoracic aorta may also spread proximally into the cervical arteries. Such proximal extensions are certainly a rare cause of stroke.⁹ Schievink et al¹⁰ recently described a comparable patient with Marfan syndrome who presented with bilateral atherosclerotic fuga after surgical repair of an aortic dissection. Angiography revealed a dissection of the ascending aorta that extended bilaterally into both carotid arteries. Although rarely described, dissecting aneurysms may also originate primarily in the cervical internal carotid arteries; this is caused by cystic degeneration in the lamina media.⁴,⁷,⁹ In such patients, additional (minor) trauma may contribute to the dissection.¹⁰ In some of these patients, similar changes in the thoracic aorta can be found.⁴ As there are no large series of patients with a histopathologically confirmed dissection, the relative contribution of aortic dissections caused by Erdheim-Gsell cystic medionecrosis as a cause of stroke in young adults cannot be estimated.

The etiology of Erdheim-Gsell cystic medionecrosis is still unknown. Some authors consider it an exaggerated age-related change of the vessel wall¹¹ because the histological changes of aortic tissue from these patients resemble normal postmortem aortic tissue; these changes in the aortic wall apparently arise at a much younger age in patients with cystic medionecrosis. Others suggest that Erdheim-Gsell cystic medionecrosis is a final common morphological pathway following exposure to a great number of different risk factors (eg, hypertension, estrogens, toxins, copper deficiency, and being male⁶,¹²). Finally, genetic predisposition may play a role in some patients. Nicod et al¹³ described a large family with aortic dissecting aneurysms and histological characteristics similar to those of Erdheim-Gsell cystic medionecrosis. The pattern of inheritance suggested autosomal dominant inheritance. However, whether family members of patients with Erdheim-Gsell cystic medionecrosis are at risk for developing a dissection of the
carotid artery is not known. With the advent of improvements in magnetic resonance imaging techniques, dissections may be diagnosed more often during life, but, unfortunately, the diagnosis of Erdheim-Gsell cystic medionecrosis rests on pathological examination of the diseased vessel. Therefore, given the current state of knowledge about this disease, we think it is not useful to employ magnetic resonance imaging as a screening tool in family members with no symptoms.

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