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CASE REPORT

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Sudden death due to giant cell coronary arteritis

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Abstract An 89-year-old woman was found dead lying in her bed. Autopsy demonstrated a pronounced thickening of all coronary arteries except for the first 2–4 cm. Death was due to a recent myocardial infarction. Microscopically, the coronary arteries showed a substantial concentric thickening of all three layers with 90% narrowing. There was a dense transmural inflammatory infiltration with lymphocytes, macrophages, and numerous multinucleated giant cells. The CD68 positive giant cells were mostly located at the media-intima border in the vicinity of fragmented fibers of the lamina elastica interna. The aorta and its major branches including the carotid arteries, however, were free of inflammation and thickening. The findings were characteristic for giant cell arteritis, the equivalent of temporal Horton arteritis, but isolated involvement of the coronary arteries is exceptional.

Introduction

Sudden or unexpected death from natural causes may be due to a wide range of sometimes extraordinary disorders (e.g., [5, 11]). Atherosclerotic coronary artery disease is by far the most frequent cause of sudden death, but there are a number of differential diagnoses. Inflammatory diseases of the coronary vessels are one [1, 2, 10, 12]. A case of giant cell arteritis leading to fatal myocardial infarction is presented where the vasculitic changes were confined to the coronary arteries.

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Case report

An 89-year-old woman was found dead lying in her bed in the morning. She was known to have mild coronary heart disease but a cardiologic examination 1 year before death, including echocardiography and long-term electrocardiography, yielded no substantial pathologic findings. During the last year, erythrocyte sedimentation rate varied between 9/25 and 79/93 mm and C-reactive protein (CRP) varied between <0.3 and 12.7 mg/dl. Leukocytes were never elevated.

The main findings demonstrated by autopsy were moderate to pronounced atherosclerosis of the aorta and its major branches, hypertrophy of the left ventricle (heart weight 385 g and body weight 51 kg), and congestion of the liver. The coronary arteries showed important thickening and glassy bloating except for the first 2–4 cm from the branching of the aorta. This had caused long-distance narrowing and obstruction of the lumen of the vessels. In the apex of the heart, including the septum and posterior wall, there was a pallor of the myocardium, which was 3 cm in diameter. The heart valves and the right ventricle did not show pathological changes.

Histology including immunohistochemical staining (C_{5b-9}) evidenced an acute myocardial infarction not older than 24 h. Old myocardial scars were absent. All coronary branches, except for the very proximal sections, showed important thickening of the walls, which had produced luminal narrowing and occlusion through intimal hyperplasia (Fig. 1). The arteries showed a dense transmural inflammatory infiltrate, which was primarily made up of lymphocytes but also included neutrophil granulocytes and macrophages (Fig. 2). In addition, numerous multinucleated giant cells were located at the media-intima border in close association to fragmented fibers of the lamina elastica interna (Figs. 2 and 3). The giant cells and surrounding macrophages reacted positively to the macrophage-specific antibody CD68 (Fig. 4), but were negative for the antibodies MRP 8 and 14 [6]. Histologically, the aorta and its major branches, including the carotid arteries, showed moderate to intense atherosclerosis, but were free of inflammation.

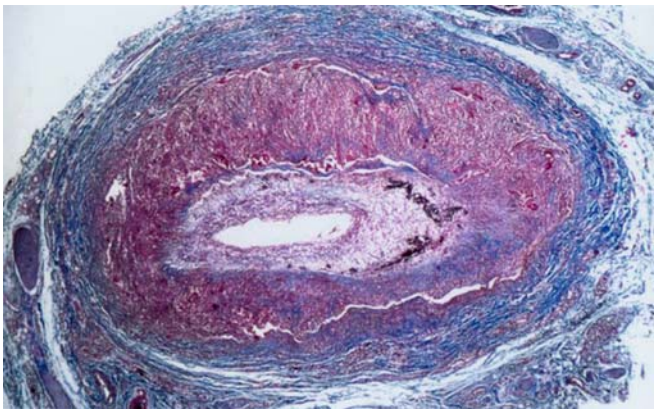

Fig. 1 Left anterior descending coronary artery, Azan, original magnification ×20. Thickening of arterial wall and concentric narrowing of the lumen by intimal hyperplasia

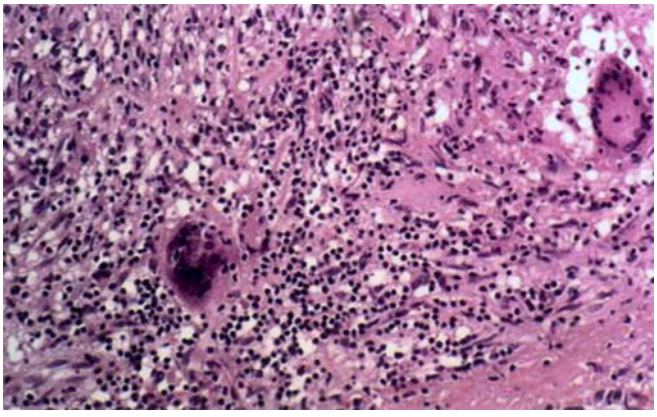

Fig. 2 Left anterior descending coronary artery, hematoxylin-eosin, original magnification ×164. Dense lymphocyte-rich inflammatory infiltrate of all three wall layers and multinucleated giant cells in the vicinity of the media-intima border

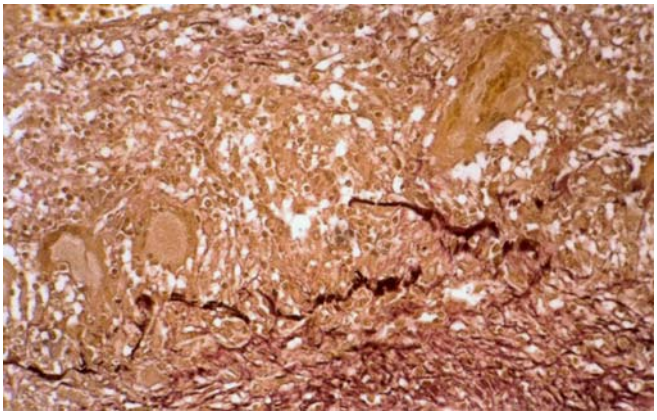

Fig. 3 Right coronary artery, elastica von Giesson, original magnification ×128. Giant cells in close association to the fragmented fibers of the lamina elastica interna

Discussion

The classification scheme for different forms of arteritis includes two entities characterized by giant cells [4, 15]: giant cell arteritis, also called arteritis temporalis Horton,

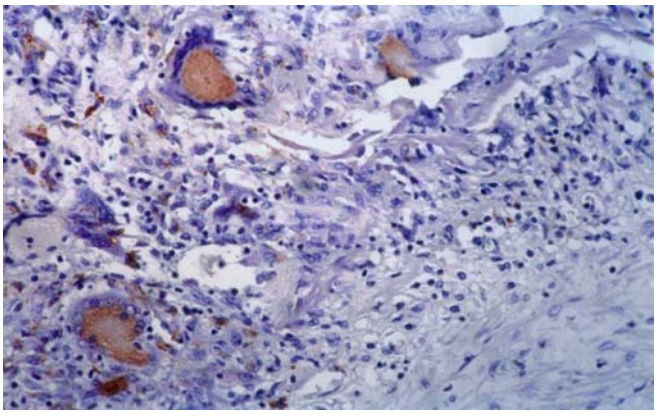

Fig. 4 Right coronary artery, CD68, original magnification ×128. The giant cells reacted positively to this macrophage-specific antibody. The arrangement of the cores of the giant cells was frequently *horseshoe-shaped* or *circular* (compare with Figs. 2 and 3). CD68 positive macrophages in the vicinity

and Takayasu arteritis. The morphological findings in our case, such as lymphocyte-dominated panarteritis, multinucleated giant cells associated with fragmented fibers of the lamina elastica interna, and numerous CD68 positive cells, are characteristic for giant cell arteritis [13–15]. The risk of giant cell arteritis is highest in women older than 75 years [3, 15], whereas women in their second and third decades of life are at highest risk for Takayasu arteritis [8]. Therefore, the diagnosis clearly was "giant cell arteritis."

However, giant cell arteritis commonly affects medium- and large-sized vessels, including the aorta in a systemic or segmental way [7, 15] while, to our knowledge, only one case of isolated coronary giant cell arteritis was published [9]. Although vasculitic lesions can be missed in a focal segmental distribution, this report consequently describes an extremely rare form of giant cell arteritis, probably confined to the midsections of the coronary arteries. This atypical distribution had caused a sudden and unexpected death without typical prodromi of the disease, such as headache, scalp tenderness, or yaw claudication, although, in retrospect, the raised erythrocyte sedimentation rate and CRP can be considered indicative. The heart had not shown major pathologic findings 1 year before death, which may be due to rapid progression of the vasculitic process in the meantime.

In conclusion, this case demonstrates the importance of forensic autopsies in unclear death cases and of a thorough examination of the heart to clarify the exact cause of death.

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