

Cardiac Involvement in Granulomatosis with Polyangiitis (GPA): Pathologic Study of 11 Fatal Cases

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Abstract: Background: Clinically significant heart involvement is a rare manifestation of GPA. However, the prevalence of cardiac involvement in GPA is probably underestimated due to their frequent asymptomatic nature.

Objectives: To investigate the variation of cardiac pathologic findings in autopsy cases of GPA.

Methods: Microscopic examinations of the heart were performed in 11 fatal cases of GPA. There were 8 males and 3 females, ranging from 16 to 74 years of age. The median duration of disease was 2,2 years with a range of 1 month to 4 years. The causes of death in patients with GPA were combined pulmonary and renal failure; profuse hemorrhage from the eroded vessels of the upper respiratory tract, lungs, intestine.

Results: The pathological changes in the heart and the coronary vessels have been found in all cases of GPA. The destructive-productive panvasculitis affecting small sized vessels was the most common finding. The vessels most affected were predominantly intramyocardial small arteries, arterioles, and capillaries, or more rarely epicardial. Histologically, the remodelling of coronary artery tree in GPA had a wide range of acute and chronic changes which likely were depended on the duration of the disease. The acute stage has been characterized by abnormalities of vessel wall from mucoid swelling to fibrinoid necrosis. The destructive changes were accompanied by the inflammatory responses with intramural and perivascular infiltrates, mainly composed of lymphocytes, and neutrophils. The chronic lesions were characterized by intimal hyperplasia, fibrosis, hyalinosis of the vessels with their luminal narrowing. The intima has become thickened, and the vessels converted into a fibrosis cord. In the affected vessels develops thrombosis with completely or particular obliterated lumen. Complete obliteration of the arterial lumen with recanalization of organized thrombi has been presented in cases with a longer duration of disease. The intimal proliferation, fibrosis, and thrombus formation leading to stenosis or occlusion of the vascular lumens, and finally to myocardial ischemia, necrosis of cardiomyocytes, and myocardial fibrosis. The destruction of the collagen fibers and extracellular matrix has been detected not only in vessels, but also in the myocardial interstitial space. The most common lesion of myocardial interstitial matrix was extravascular granulomatous inflammation with a variegated cellular infiltrate. Microscopically, the cellular infiltrate made up of lymphocytes, plasma cells, neutrophils, histiocytes, and epithelioid cells. It should be noted that in our series of autopsy cases of GPA the giant cell reaction occurred rare. Macroscopically, the chambers of the hearts were dilated. In addition, other non-specific findings of GPA included pericarditis that was observed in two cases.

Conclusions: Our data suggest that cardiac involvement is common in GPA. Analysis of cardiac pathological changes in 11 fatal cases of GPA showed that the manifestations were multiple. It can be assumed that the most prominent histologic feature is microcirculatory vasculitis with coronary flow reduction in the microcirculatory bed of myocardium. The second typical morphological finding is extravascular granulomatous inflammation of myocardial interstitial matrix.

Disclosure of Interest: None declared

Key words: Granulomatosis with polyangiitis, cardiac involvement, pathologic study