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Case review

Pseudotumour formation in atheromatous coronary arteries

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ABSTRACT

Three cases with mass like lesions (pseudotumours) surrounding atheromatous coronary arteries were referred to the Royal Brompton Hospital for expert pathology review. All were males with mean age 74 years (range 55–91). In all cases, coronial autopsies were carried out for sudden deaths in the community. Past medical histories of note were hypertension (N = 2) and ischaemic heart disease (N = 1), with one patient having a past history of aortic aneurysm repair.

At autopsy, firm, white and whorled masses surrounded both right and left coronary arteries ranging in size from 9 to 25 mm in diameter. Each coronary artery had intimal atheroma with associated stenosis ranging from moderate to severe. A thrombus was identified in one case.

Histological sections showed a mixed inflammatory infiltrate extending from the media into the adventitia of each coronary artery, composed predominantly of plasma cells and lymphocytes with rare neutrophils and eosinophils. There was accompanying dense fibrosis accounting for approximately 50% of the mass size on microscopic examination of slides. The presence of intimal circumferential atheroma was confirmed in all cases.

Immunohistochemical studies showed staining with IgG4 in two of three cases.

Atheroma may be associated with mild chronic inflammation present in the intima or associated with plaques and adventitia. The differential diagnosis for coronary artery inflammatory masses would include vasculitis, syphilis, inflammatory pseudotumor and IgG4 associated disease.

This is the first report of isolated coronary artery IgG4 related disease in association with atheroma.

1. Introduction

Coronary artery atheroma accounts for the vast majority of sudden cardiac deaths in the community.[1] In most cases autopsy examination yields atheromatous coronary arteries and occasionally a thrombus.[2] The pathogenesis of atheroma is due to endothelial damage and lipid accumulation in foamy macrophages[3] and consequently these lesions are often associated with chronic inflammatory cells.

Coronary artery vasculitis accounts for a rare number of deaths.[4] Similarly mass lesions associated with coronary arteries are infrequent findings at autopsy. Possible aetiologies include aneurysm, pseudo aneurysm or malignancy such as lymphoma.

We report a series of atheromatous coronary arteries with masses seen grossly and histologically confirmed florid lymphoplasmacytic inflammation, the differential diagnosis for which would include vasculitis, syphilis, inflammatory pseudotumor and IgG4 associated disease.

We discuss the histological and immunohistochemical findings and report for the first time isolated coronary IgG4 disease in the setting of atheroma.

2. Case histories

Three cases with mass like lesions surrounding atheromatous coronary arteries were referred to the CRY Cardiac Pathology Unit at Royal Brompton Hospital, a tertiary cardiac pathology referral centre. All cases were referred for an expert opinion, from the State Pathologist's Department at the Institute of Forensic Medicine, Belfast, which carried out 1132 autopsies that year. Routine autopsy

Abbreviations: CRY, cardiac risk in the young; SLE, systemic lupus erythematosus; LAD, left anterior descending; RCA, right coronary artery.

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examination involved review of prior medical history and circumstances of death, detailed external examination, internal examination and dissection of all organs and histological assessment of all major organs (heart, lungs, liver, kidneys) as well as examination of any macroscopic abnormalities seen. Slides and blocks from the coronary arteries and myocardium were referred for review. Immunohistochemical studies were carried out using 4 mm thick formalin fixed, paraffin embedded tissue sections with the antibodies against CD3, CD20, ALK-1, CD68, IgG4 and IgG. The slides were prepared using heat induced epitope retrieval (CD3, CD20, CD68 & ALK-1) and enzyme retrieval (IgG & IgG4) and stained on a Ventana Benchmark platform. Slides for IgG4 and IgG were reviewed and areas of highest concentration of staining cells identified. Three high power fields (40x) were counted and an average calculated along with the IgG4 to IgG ratio.

The three cases were all males with mean age 74 years (range 55–91 years). In all cases coronial autopsies were carried out for sudden deaths in the community. Past medical histories of note were hypertension (N = 2) and ischaemic heart disease (N = 1), with one patient having a past history of aortic aneurysm repair. No history of left heart catheterization, coronary artery angioplasty, and/or stenting was noted.

At autopsy, firm, white and whorled masses were described surrounding atheromatous left and right coronary arteries ranging in size from 9 to 25 mm in diameter (Fig. 1). These extended from the ostia of the vessels all along the route of each to the distal aspect of the vessel. In one case masses were present around both right and left arteries. The other cases involved the main branches of a single coronary artery (right = 1, left = 1). They were surrounding and intimately involved with the wall of the coronary artery. Each coronary artery had intimal atheroma with narrowing of the lumen ranging from moderate (50% stenosis) to severe (>75% stenosis). A thrombus was identified in one case (Fig. 2). No gross infiltration of the myocardium was seen. No vascular abnormalities or lesions were identified elsewhere in two of the cases. No masses were identified elsewhere on gross examination. One of the cases had repair of an abdominal aortic aneurysm five years prior to death, however this was on a background of severe atheromatous aortic disease and hypertension. At autopsy the Dacron graft was patent and there was evidence of extensive atheroma but no inflammatory masses.

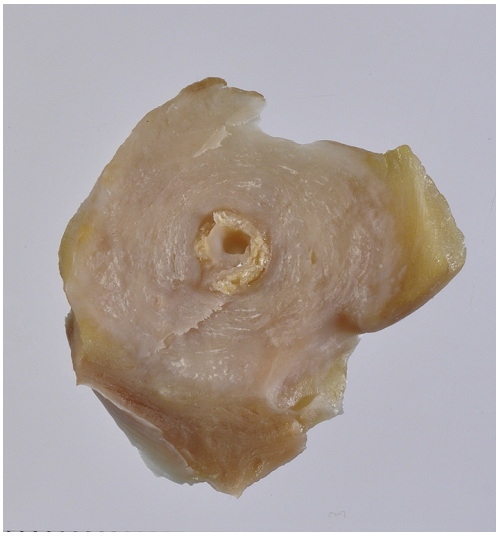

Fig. 1. Macroscopic image of coronary artery showing concentric atheroma with associated fibrotic mass extending into surrounding fat.

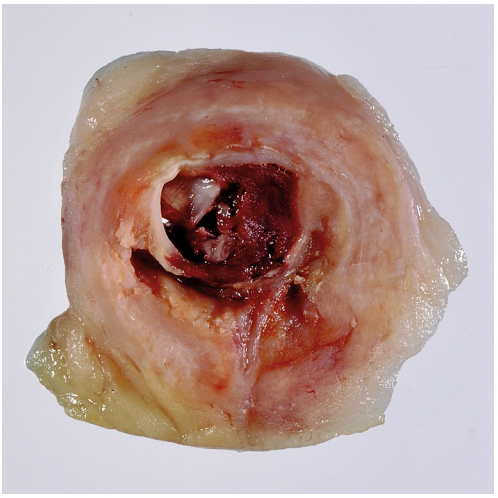

Fig. 2. Macroscopic image of coronary artery with concentric atheroma, plaque rupture with thrombus formation. Surrounding the artery there is fibrosis which shows extension into fat.

Histological sections of coronary arteries showed a mixed inflammatory infiltrate extending from the media into the adventitia, composed predominantly of plasma cells and lymphocytes with rare neutrophils and eosinophils. No giant cells or epithelioid cells were noted. No necrosis was present. Inflammation extended into the surrounding epicardial fat with associated lymphoid aggregate formation at the edge along with focal infiltration of the myocardium by these lymphoid aggregates (Fig. 3). There was accompanying dense fibrosis accounting for 50% of the mass size on microscopic examination of slides (Fig. 4). The presence of intimal circumferential atheroma was confirmed in all cases. Thrombosis associated with rupture was seen in one case. All the intramural vessels were free of atheroma and did not contain this inflammatory infiltrate.

Myocardial fibrosis was identified in one case. Besides the focal lymphoid aggregates described, no inflammatory infiltrate was present in the rest of the myocardium. No vasculitis or lymphoplasmacytic inflammation was seen on histological sections of other organs sampled at autopsy. Histological assessment of the pancreas was carried out in two of three cases and showed no evidence of IgG4 related disease. No histological examination of the aorta was carried out.

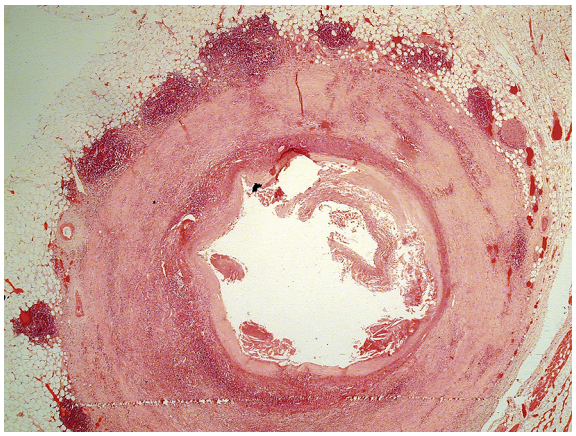

Fig. 3. Low power (4X) microscopic image of coronary artery with mild concentric atheroma and surrounding fibrosis with inflammation including multiple lymphoid aggregates. The inflammation shows infiltration into surrounding fat.