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ANNULAR SUBVALVULAR LEFT VENTRICULAR ANEURYSM ASSOCIATED WITH CARDIAC LYMPHOSTASIS

R.W. Lupinski, S. Shankar, K.Y. Wong, Y.H. Chan

Departments of Cardiothoracic Surgery (RL), Groote Schuur Hospital, Capetown, South Africa, and KK Hospital (SS,KYW, YHC), Singapore

Annular Subvalvular Left Ventricular Aneurysm (ASLVA) is a rare cardiac lesion of unknown etiology predominantly seen in young patients of African origin. A patient with ASLVA is presented in whom histopathological features of the aneurysm wall revealed cardiac lymphostasis. This finding may bring new light on understanding this unusual heart condition.

CASE REPORT

A 19 year old Bantu woman was admitted to Groote Schuur Hospital (Cape Town) for evaluation of a suspected cardiac aneurysm. One year earlier she developed a nocturnal productive cough associated with palpitations. One month before admission, she developed pulmonary edema and was found to have severe mitral valve incompetence with an unusual left heart border on plain chest radiograms. On admission, she had an upper respiratory tract infection, widespread skin rash and low-grade fever. Cardiac auscultation revealed a loud 1st sound, a normal 2nd sound and a grade 3/6 pansystolic murmur at the apex with an associated 3rd sound. A mid-diastolic murmur was also audible. Bilateral crepitations were present in the lower zones.

Laboratory Findings

White blood cell was 16,000/mm^3, sedimentation rate was 132/hr, C-reactive protein 6.6, antistreptolysin titer and antinuclear factor were negative. Blood tests for syphilis and AIDS were also negative. Blood culture were negative x3. Chest radiogram revealed an unusual left heart contour *(Fig 1)*. There was mediastinal lymphadenopathy in the right paratracheal region with compression of the right tracheal border *(Fig 1)*. Electrocardiogram showed sinus tachycardia. There were pathologic Q waves in leads I and AVL with associated T wave inversion in AVL. On angiography, the coronary arteries were patent. An echocardiogram revealed attenuation of the posterolateral portion of the mitral valve annulus, which was bridged by a large thin membrane, i.e., a roof of submitral aneurysm. She underwent operative repair of the aneurysm and remains well four years later.

Histopathology of the aneurysm wall revealed fibrous tissue lined by endothelium. There also was dilatation, deformation and ruptured vessels with extravasated lymph *(Fig. 2)*. "Tooth-like" processes also were seen.

COMMENT

This is the first known report of ASLVA associated with cardiac lymphostasis. Mediastinal lymphadenopathy perhaps restricted lymph outflow from the heart (1), a phenomenon previously observed in patients

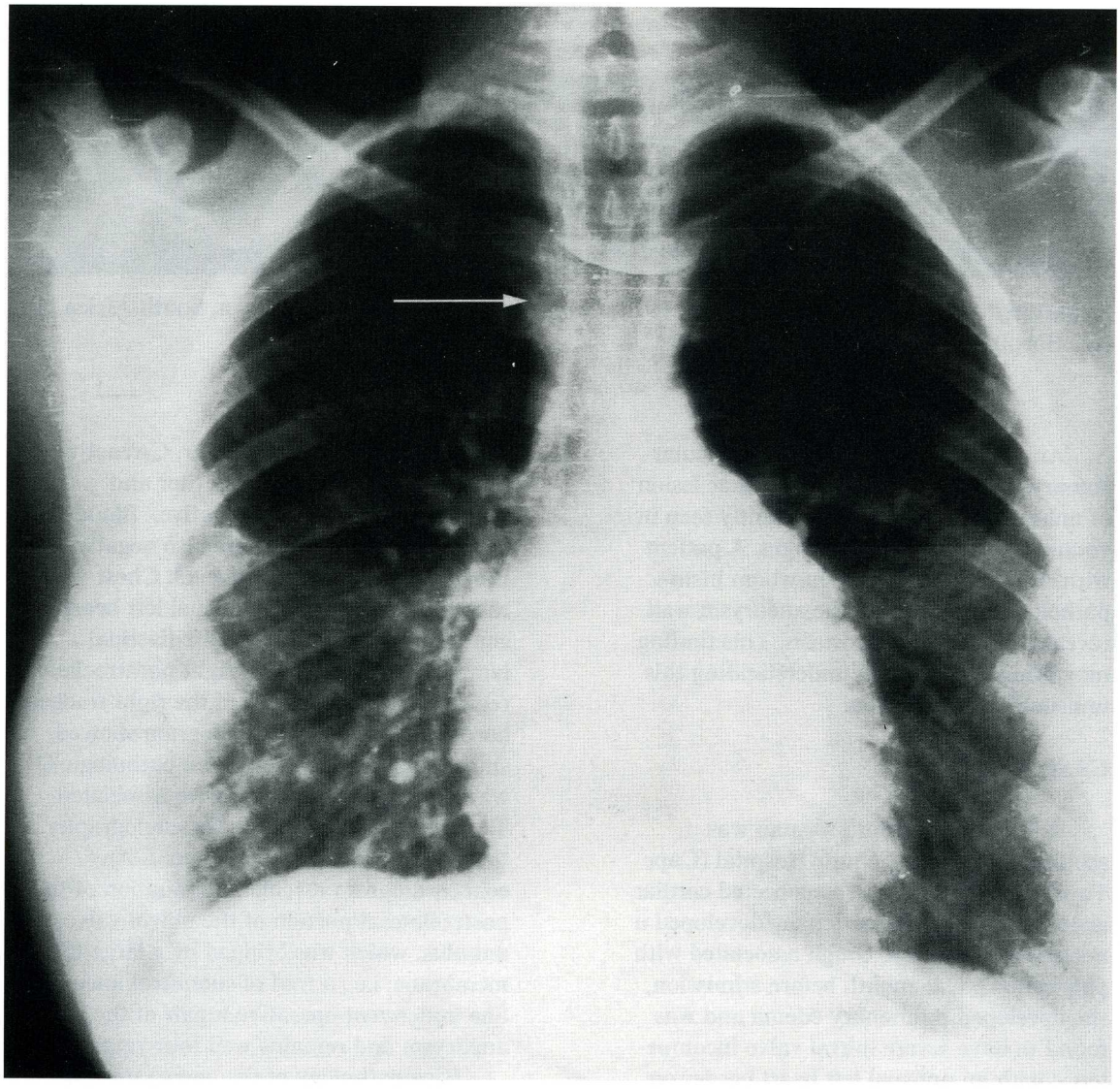

Fig. 1 Chest x-ray demonstrating unusual left cardiac border with irregular mass in right paratracheal region with compression of the trachea (arrow)

with ASLVA (2,3). "Cardiac lymph nodes," which collect lymph from the heart, are located in the right paratracheal and/or subcarinal group of lymph nodes (4). Similar histopathological features of cardiac lymphostasis as described in our patient have been described in experimentally induced cardiac lymphostasis (5). Extensive overgrowth of collagen may relate to intravascular shear stress in the presence of cardiac lymphostasis with activation of endothelial pathways leading to tissue fibrosis.

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Fig. 2. Cardiac lymphostasis associated with ASLVA. Between arrows dilatation and deformation of a vascular channel containing lymph (L) is seen.

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Richard W. Lupinski, MD, PhD
Department of Cardiothoracic Surgery
KK Hospital
100 Bukit Timah Road
Singapore 229899