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A new type of coronary vasculitis (Eosinophilic Coronary Periarteritis, ECPA) showing vasospastic angina and sudden cardiac death

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Eosinophilic coronary periarteritis (ECPA) was recently reported by Kajihara and his co-workers as a new pathologic entity, and clinically showed a vasospastic angina and sudden cardiac death (SCD). The patients were relatively young (mainly 30 to 50 years old) and predominantly male. The characteristic clinical findings of this disease include a) vasospastic angina (Prinzmetal's variant angina) appearing usually from evening to early in the morning, b) all patients experienced SCD early in the morning, and c) allergy or history of allergy was hard to identify in the patients with this disease. Histological findings include a) eosinophilic inflammatory infiltration limited to the adventitia and periadventitial soft tissue is recognized in the epicardial large coronary arteries, b) all 3 main coronary artery branches are affected, with the left anterior descending artery most frequently affected, c) medial smooth muscle cells of the affected coronary artery and both internal and external elastic laminae are well preserved, d) fibrinoid necrosis or granuloma as seen in polyarteritis nodosa (PN) or allergic granulomatous angiitis (AGA) are not found in or around the inflammatory areas, and e) no findings of any type of vasculitis in any other tissue or organs. Spontaneous coronary artery dissection (SCAD) is frequently accompanied by eosinophilic inflammatory infiltration limited to the adventitia and periadventitial soft tissue in the dissected portion of the epicardial coronary arteries, i.e., same as the findings of ECPA, and the patients usually die suddenly. ECPA was recently reported in the patient with cocaine abuse. The diagnosis of ECPA is very difficult to make at the clinical examination stage and is made almost exclusively at autopsy. However, vasospastic angina appearing mainly from the evening to early in the morning (Prinzmetal's variant angina) is the most important symptom of this disease.

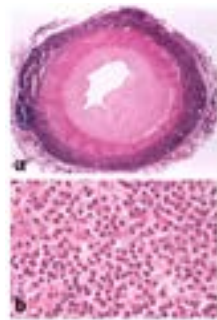


Figure. a, b: right coronary artery of ECPA

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