



Pathology Biology Section – 2007

G58 Sudden Death and Keratoderma Associated Cardiomyopathy: An “Affray” of the Heart

Christopher M. Milroy, MD, LLB, and Sacha Kolar, MBChB, Forensic Science Service, The Medico-Legal Center, Watery Street, Sheffield, South Yorkshire S3 7ES, United Kingdom; and Kim Survana, MBChB, Sheffield Teaching Hospitals, Northern General Hospital, Herries Road, Sheffield, South Yorkshire S5 7AU, United Kingdom*

After attending this presentation, attendees will have knowledge of cardiomyopathy associated sudden death and keratoderma as well as the law regarding manslaughter in England.

This presentation will impact the forensic community and/or humanity by discussing issues of interaction of sudden “natural” death and the law pertaining to homicide in England.

A 15-year-old girl was involved in an incident in which she was subject to threats, then blows before running away from the incident. She ran uphill away from the incident, collapsed and died. At autopsy a history was given that she suffered from the skin disorder Erythrokeratoderma Variabilis or possibly Papillon-Lefèvre syndrome and was on antihistamines. At autopsy there was significant thickening of the skin, most obvious of the palms and soles of the feet. The hair was abnormal as were the teeth. On internal examination the heart was clearly abnormal. There was a degree of dilatation of the ventricles and on microscopic examination there was obvious fibrosis, with no evidence of acute infarction or inflammation. A diagnosis of Plantopalmer Keratosis associated cardiomyopathy was made.

There are a number of genetic abnormalities associated with Plantopalmer keratosis. A number of heart conditions may be found including Naxos syndrome and Carvajal syndrome.

A charge of manslaughter, with an additional charge of affray was laid against four teenage defendants. The basis of the manslaughter charge was the charge of affray, except for brain edema and generalized haemostasis. Three of the defendants were convicted of manslaughter and affray. The fourth defendant was acquitted.

On appeal, the convictions for manslaughter were overturned. The convictions for affray were upheld. As well as the unique pathologic features, this case has clarified the law of manslaughter in the English legal system, limiting its extent to an act that is dangerous in the sense that sober and reasonable persons would recognize it as foreseeable.

Reference:

1. *R v Carey, C and F* (2006) EWCA Crim 17

Cardiomyopathy, Homicide, Keratoderma