Diagnosis of a rare cause of arrhythmogenic syncope by means of an implantable loop recorder

A 39 year old woman was admitted for evaluation of recurrent syncope. A detailed workup, including invasive electrophysiological testing, was unremarkable. Echocardiographic studies showed normal ventricular dimensions, conserved systolic function, normal left atrium dimension, and normal valvar structures. A rounded mass of heterogenic density was found in the right atrium. The mass arose in the lateral wall and prolapsed through the tricuspid valve during diastole.

The patient underwent surgery for removal of the mass, and he has since remained asymptomatic.

The excised specimen measured 6 × 4 cm, and weighed 40 g. Grossly it was sessile, solid, oval shaped, and had a smooth surface (panel A). Foci of haemorrhage were present throughout the myxoma. The cut surface was grey-white with yellow coloured areas and a stone-like consistency (panel B).

Histological examination revealed that the mass comprised polygonal myxoma cells embedded in a myxoid matrix of acid mucopolysaccharide. No atypias or mitosis were found. Massive bone metaplasia was evident (panel 1), with extramedullary haemopoiesis (panel 3) resembling the appearance of normal trabeculae (panel 2).

Myxoma is the most frequent primary tumour of the heart, arising from the endocardium as a pedunculated mass. Mostly they are found in the left atrium (82%).

Lithomyxoma is a rare pathological entity, first described by Strousse in 1938. Although foci of microscopic calcification and areas of metaplastic bone occasionally occur, massive ossification is a rare event; current literature reports 10 cases of lithomyxoma. Calcification may be considered as a degenerative, age related phenomenon, caused by haemodynamic factors.

In conclusion, potentially life threatening polymorphic ventricular tachycardia in this young woman without structural heart disease and recurrent syncope could only be diagnosed by means of an implantable loop recorder.