Kardiologia Polska 2017; 75, 3: 280; DOI: 10.5603/KP.2017.0053 ISSN 0022–9032

STUDIUM PRZYPADKU / *CLINICAL VIGNETTE*

Left ventricular tumour masquerading as hypertrophic cardiomyopathy: how one can be misled by transthoracic echocardiography

Guz serca imitujący kardiomiopatię przerostową — jak można się dać zwieść echokardiografii

Joanna I. Ateńska-Pawłowska[1], Maciej Sterliński[2], Artur Oręziak[2], Marcin Sobiech[1], Michał Lewandowski[1]

[1]2nd Department of Coronary Artery Disease, Institute of Cardiology, Warsaw, Poland
[2]Department of Heart Rhythm Disorders, Institute of Cardiology, Warsaw, Poland

Cardiac fibromas are common benign primary cardiac tumours in children. The incidence of these tumours is rare in the adult population but increases in patients suffering from Gorlin's syndrome. These patients present with multiple basal cell carcinomas, ovarian fibromas, medulloblastomas, as well as skeletal abnormalities and congenital malformations. We present a case of a 41-year-old male with a longstanding history of heart failure in the course of hypertrophic cardiomyopathy (HCM) treated with implantable cardioverter-defibrillator (ICD), as primary prevention of sudden cardiac death, for seven years. At this point he was admitted to our clinic because of atrial and ventricular lead failure. The Heart Team decided on the transvenous lead removal strategy. During transoesophageal echocardiography performed intraoperatively a pathological mass in the right atrium was first encountered. Previous multiple transthoracic echocardiography (TTE) performed with poor echocardiographic window using only standard views as well as electrocardiogram (Fig. 1) did not drown suspicion of any pathology other than HCM. After ICD-VR re-implantation (intentional downgrading of CIED) a chest computed tomography (CT) was performed, which revealed the presence of a heart tumour with degenerative calcifications arising from interventricular septum heading downwards creating a flat base under both ventricles (Fig. 2A), the existence of which become apparent also in TTE (Fig. 2B). On the grounds of the morphology of the tumour, which was strongly suggestive of a fibroma as well as a bifid third rib, also discovered by means of CT (Fig. 3), Gorlin's syndrome was suspected. On physical examination craniofacial anomalies such as macrocephaly and frontal bossing as well as plantar pitting became apparent. Moreover, the patient admitted to having basal cell skin carcinomas in different locations resected twice in his life. Further evaluation showed ectopic intracranial calcifications including of the falx cerebri, bridging of the sella turcica, and jaw bone remodelling on skull radiograph along with mild lytic lesions on hand and foot radiographs. These findings met four major and three minor criteria warranting the final clinical diagnosis of Gorlin's syndrome. As consulting cardiac surgeons considered tumour resection not feasible, the patient has ben treated conservatively with no evidence of tumour growth during one-year follow-up. The ICD therapy has been continued as cardiac fibromas had been often associated with malignant arrhythmia and increased risk of sudden cardiac death. Although echocardiography is generally accepted as an initial diagnostic test for the presence of a cardiac tumour, this imaging method remains highly operator-dependent. In the case of poorly echogenic patients or suboptimal acoustic window a cardiac mass can easily be missed. This case highlights the importance of careful examination of every patient and the necessity of acquiring non-standard views because pathologies can be overlooked during routine echocardiography (Fig. 4).

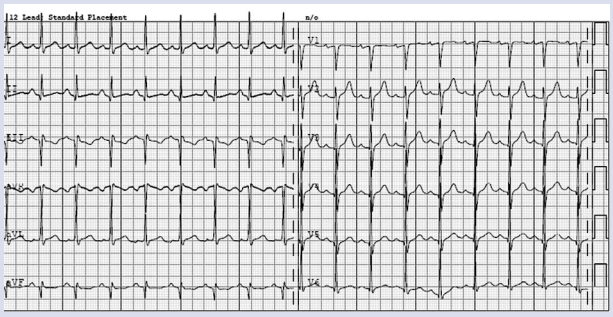

Figure 1. Electrocardiogram on admission

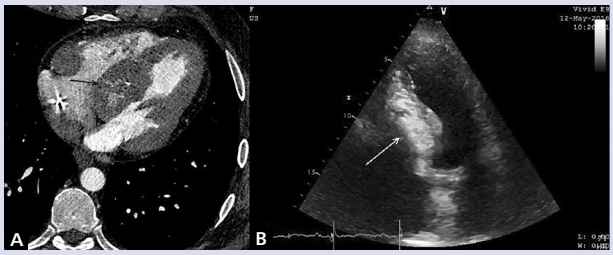

Figure 2. **A.** Computed tomography; pathological mass arising from interventricular septum; **B.** Three-dimensional transthoracic echocardiography focused on the tumour; in panels A and B the arrows point to the tumour

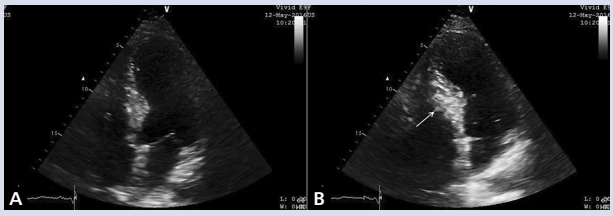

Figure 4. Transthoracic echocardiography; **A.** Standard apical four chamber view; **B.** Non-standard view after tilting the probe when the tumour becomes apparent (arrow)

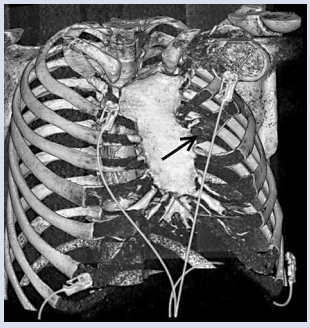

Figure 3. Computed tomography — rib cage reconstruction; the arrow indicates the bifid rib

Address for correspondence:
Joanna I. Ateńska-Pawłowska, MD, 2nd Department of Coronary Heart Disease, Institute of Cardiology, ul. Puławska 255/40, 02–740 Warszawa, Poland, e-mail: atenska@gmail.com

Conflict of interest: *none declared*