A giant interatrial mass: an unusual presentation of primary amyloidosis

Guz w przegrodzie międzyprzedsionkowej serca – nietypowy obraz pierwotnej amyloidozy

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Abstract

We report a patient with primary systemic amyloidosis who had a very unusual form of clinical and instrumental presentation.

Key words: transthoracic and transoesophageal echocardiographic, interatrial septum, amyloidosis

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Case report

We report a patient with primary systemic amyloidosis who had a very unusual form of clinical and instrumental presentation.

The patient was admitted to our department for dyspnoea and atrial flutter. Transthoracic and transoesophageal echocardiographic examination demonstrated an unusual significant increase and thickening of the atrial septal wall and an echo-dense, high increased echogenicity/granular sparkling circular mass inside the interatrial septum while all the other echocardiographic findings were normal (particularly normal biventricular dimensions and function, normal appearance and thickness of left ventricle walls) (Figure 1). Computed chest tomography (CT) showed a giant expansive lesion located along the interatrial septum with granular sparkling appearance, sparing the fossa ovalis (maximum thickness diameter 28 mm × 56 mm), expanding and infiltrating

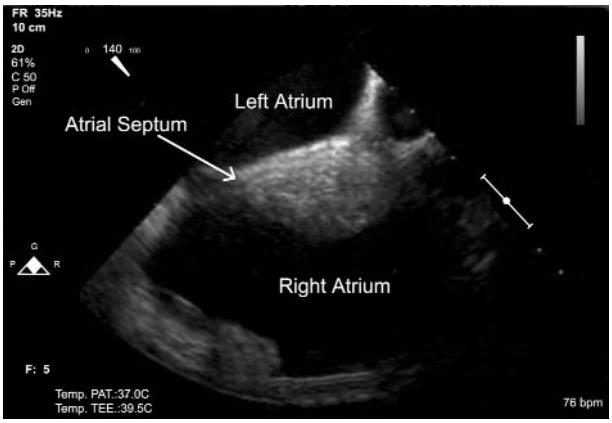

Figure 1. Transoesophageal echocardiographic bicaval view at 114° showing the large amyloid mass infiltrating the interatrial septum

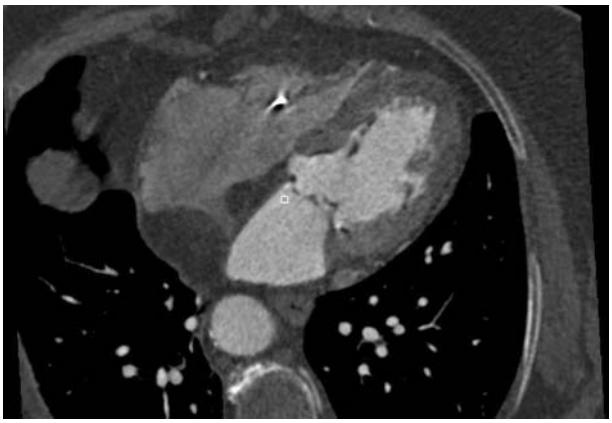

Figure 2. Computed chest tomography showing a thickened interatrial septum with the huge amyloid mass extending to the right atrial wall

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the posterior right atrial wall (Figure 2). The mass also caused compression of the terminal portion of the superior vena cava without clinical and haemodynamic instability. Fine needle umbilical fat biopsies were performed and resulted positive for systemic amyloidosis. Congo-red staining was performed using standard laboratory technique. The final diagnosis is primary amyloidosis.

This is a rare case of a localised amyloid deposit in the heart of a patient with no clinical evidence of a pre-existing systemic disease. Echocardiographic examination is a well known important tool for establishing the presence of cardiac amyloid involvement and may be useful in estimating prognosis in such patients. Obviously echocardiographic images should be interpreted in the context of the clinical picture and other investigations. A thickened interatrial septum has been shown in a minority of patients [1], and a study by Bhandari et al. [2] showed it to be specific for amyloid in the later stages of the disease, with 100% specificity. This case represents a very unusual echocardiographic presentation of primary systemic amyloidosis, and a giant interatrial septum mass was the first sign of cardiac amyloidosis.

References

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