An unusual case of amaurosis fugax due to papillary fibroelastoma of cor triatriatum

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A 58-year-old woman with no significant medical history presented to us with a transient left-sided visual loss. Apart from previous smoking, she had no risk factors for cerebrovascular event.

On examination, she had a regular heart rate of 64/min and a blood pressure of 120/90 mmHg. Neurological and cardiorespiratory examinations were unremarkable. Chest radiograph, electrocardiogram, laboratory data, and computed tomography head were unrevealing.

A transthoracic echocardiography, and subsequently, a transoesophageal echocardiography (TOE), was performed, revealing a membrane-like echodense structure in the left atrium (LA) with attachment at the septum medially and Coumadin ridge laterally, dividing the atrium into a posterior-superior chamber where the pulmonary veins drained and an anterior-inferior portion which included the LA appendage, suggestive of cor triatriatum. In addition, a mobile echodense mass was attached to the inferior aspect of the membrane (Figure 1C and D). Three-dimensional (3D) echocardiography by TOE confirmed the membrane fanning across the LA towards the inter-atrial septum with two large orifices and further defined the mass as polypoidal and gelatinous in appearance (Figure 1A and B). A presumptive diagnosis of an embolic event due to myxoma of cor triatriatum was made.

The cor triatriatum and the mass were surgically removed (Figure 1E) and histologically, papillary fibroelastoma was observed. Post-operative course was uncomplicated and she has had no further event.

This case highlights the utility of 3D echocardiography in defining complex structure and the importance of being aware of the possible embolic complications related to exceedingly unusual co-existence of two rare entities.

Figure 1 A three-dimensional transoesophageal echocardiography en face surgical view of the cor triatriatum (A). A three-dimensional transoesophageal echocardiography view of the polypoidal mass (B). A five-chamber apical view of transthoracic echocardiography (C). Transoesophageal echocardiography: 90° mid-oesophageal view (D). Surgical photo of the cor triatriatum and the attached mass (E). White arrow, mass; CT, cor triatriatum; Ao, aorta; MV, mitral valve; IAS, inter-atrial septum; LAA, left atrial appendage; PW, posterior wall.

Supplementary data are available at European Journal of Echocardiography online.