Vanishing right ventricular outflow obstruction: an unusual presentation of mediastinal lymphoma

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A 23-year-old patient referred for cardiac evaluation in view of easy fatiguibility, significant weight loss, and a systolic murmur. Examination revealed muscle wasting, lymphadenopathy, and an ejection systolic murmur in the pulmonary area. Mediastinal widening was seen on chest X-ray. Echocardiography showed a large extracardiac soft tissue mass compressing the right ventricular outflow tract (RVOT) and main pulmonary artery (Panel A) with turbulent flow across the RVOT (Panel B) (see Supplementary material online, Video S1–S3) and a peak systolic gradient of 55 mmHg (Panel C). There was no intra-cardiac mass. Computed tomography revealed a large mediastinal mass (Panel D) which proved to be diffuse large B-cell lymphoma (DLBCL), CD-20+, on biopsy. The patient was treated with Rituximab-Cyclophosphamide-Doxorubicine-Vincristine-Prednisolone (R-CHOP regimen) based chemotherapy with excellent clinical and radiological response. Echocardiography 3 months after treatment showed resolution of the mass with normal spectral and colour-Doppler assessment of flow across the RVOT (Panels E and F) (see Supplementary material online, Video S4).

Diffuse large B-cell lymphoma is the most common form of Non-Hodgkin’s lymphoma, accounting for up to 30% of cases. It is an aggressive tumour. Patients typically present with nodal or extranodal disease, exhibiting rapid tumour growth and symptoms depending on the tumour localization. Diffuse large B-cell lymphoma presenting primarily to a cardiologist with RVOT obstruction is extremely rare. The most widely used treatment is R-CHOP. If left untreated, DLBCL has a median survival of <1 year.

Supplementary material is available at European Heart Journal online.