Superior vena cava syndrome as a rare complication to lipomatous atrial septal hypertrophy (LASH)

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A 66-year-old woman with a history of breast cancer was admitted due to dyspnoea and excessive weight gain. Severe oedema of the upper body, resting dyspnoea, and extreme obesity (BMI 40 kg/m²) was noted (Panel A). Superior vena cava syndrome (SVCS) was suspected. A contrasted computer tomography (CT) scan revealed an obstructing tumour in the right atrium (RA) extending to the SVC (Panel B). Transoesophageal echocardiography showed a 5.0 × 4.1 cm tumour in the interatrial septum sparing the fossa ovalis (Panel C, arrow) protruding into the RA. The patient deteriorated within hours and was rushed to surgery. Macroscopically the cardiac tumour was large, mucinous and non-papillary, measuring 6.5 × 6.5 × 1.0 cm (Panel D) and integrated into the atrial septum, involving almost the complete circumference of the SVC and extending down to the roof of the RA. The patient deteriorated within hours and was rushed to surgery. Macroscopically the cardiac tumour was large, mucinous and non-papillary, measuring 6.5 × 6.5 × 1.0 cm (Panel D) and integrated into the atrial septum, involving almost the complete circumference of the SVC and extending down to the roof of the RA. Histopathology showed a combination of brown and mature adipose tissue with fibrous septae and hypertrophied myocytes with no evidence of malignancy. The tumour was diagnosis as lipomatous atrial septal hypertrophy (LASH) (Panel E). The aetiology is uncertain, but believed to occur as a developmental aberrancy in the atrial septum, thus always sparing the oval fossa (Panel C, arrow). It accounts for <1% of benign cardiac tumours. Treatment is usually conservative, but in patients suffering from intractable arrhythmia or SVCS, surgery may be unavoidable.

This image focus illustrates a case of what is usually considered a benign condition—LASH—leading to a lethal complication with obstruction and severe symptoms of SVCS.

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