CASE REPORT

Lipoma of superior vena cava: a rare occurrence

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Accepted 19 June 2017

SUMMARY

Most benign primary cardiac tumours are myxomas; non-myxomatous tumours are less common but comprise a wide variety. Cardiac lipoma is a rare non-myxomatous variety. A 70-year-old Caucasian woman with right breast cancer status postpartial mastectomy underwent surveillance MRI of the breast and was found to have a possible right atrial (RA) mass. She also reported frequent headaches and palpitations. She underwent a transoesophageal echocardiogram which showed a 2.6×1.6×1.6 cm echogenic mass at the superior vena cava (SVC) and RA junction. She was anticoagulated for a possible thrombus without resolution. Surgical excision was undertaken in view of ongoing symptoms and partial occlusion of the SVC. Intraoperatively, a 2–3 cm smoothly textured lobulated mass was found and histopathology showed adipose tissue consistent with lipoma. The postoperative course was uneventful, and the patient was discharged in stable condition.

BACKGROUND

Primary cardiac tumours are rare and broadly divided into benign and malignant categories. Benign cardiac tumours are either myxomatous or non-myxomatous.¹ The majority of benign tumours are myxomas; non-myxomatous tumours are less common and include rhabdomyoma, fibroma, haemangioma and lipoma.² Cardiac lipoma (CL) is a rare non-myxomatous benign tumour.

CASE PRESENTATION

We report a rare case of superior vena cava (SVC) lipoma in a 70-year-old Caucasian woman with a history of breast cancer. The patient underwent a partial mastectomy and radiotherapy for her breast cancer, T1N0M0, 1 year previously and was being seen by her oncologist for follow-up when she complained of frequent headaches and palpitations. She denied pain or swelling in her breast, focal neurological symptoms, facial congestion,

upper extremity swelling, chest pain or shortness of breath. On physical examination, there was no distension of head or neck veins, facial plethora or upper extremity oedema. A surveillance breast MRI was performed which incidentally showed a filling defect at the junction of the right atrium (RA) and SVC (figure 1).

The history was further explored for predisposition to SVC thrombosis. She denied placement of intravenous ports during treatment of her breast cancer which did not include chemotherapy. She also denied recent central venous catheterisation or blood clots in her extremities.

INVESTIGATIONS

The patient was evaluated with a transoesophageal echocardiogram (TEE), which showed a 2.6×1.6×1.6 cm partially obstructing round, echogenic mass at the SVC and RA junction (figure 2). There were no valvular vegetations; the TEE was otherwise unremarkable.

Cardiac MRI might have provided useful information, especially on T1-weighted and fat suppression sequences, but it is not available in our community. Venography, which might have reconfirmed the filling defect, was of no extra value at that point.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of this mass included SVC thrombus in addition to a primary cardiac tumour at an unexpected location. When the differential, including the possibility of thrombus, was discussed with the patient, she opted for a trial of anticoagulation consistent with her desire to avoid surgery. Checking D-dimer and venous Doppler of extremities, if negative, might have enhanced the post-test probability of cardiac tumour, thereby avoiding a trial of anticoagulation. We did not perform those tests. On the other hand, if this was proven to be a primary tumour, without evidence of impending obstruction, that is, SVC syndrome, surgery was not urgent even if ultimately required. We opted for a 3-month trial of oral anticoagulation followed by a repeat TEE. The risks and benefits were explained to the patient including the necessity of surgery if the mass did not resolve with anticoagulation.

TREATMENT

A trial of oral anticoagulation with rivaroxaban 20 mg daily was given. No significant change was noted on repeat TEE at 3 months. The patient continued to have frequent headaches, which raised concern for possible intermittent transient obstruction of SVC by the mass. Evidence of SVC

Figure 1 MRI of breast. (A) Axial image of breast showing a filling defect in the superior vena cava. (B) Sagittal image showing a filling defect at the junction of the right atrium and superior vena cava.

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To cite: Wahab A, Chaudhary S, Munir A, et al. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-220641

Figure 2 Transoesophageal echocardiogram (TEE). (A) Bicaaval view showing the lipoma (arrow). (B) Short axis view of SVC and aorta showing lipoma (arrow). (C) 3D view of the lipoma (arrow). (D) Bicaaval view postsurgery showing complete resection of the lipoma. Ao, ascending aorta; LA, left atrium; RA, right atrium; SVC, superior vena cava.

syndrome, however, remained absent throughout the course. A consultation with cardiothoracic surgery was obtained for excisional biopsy for tissue diagnosis and for the possible development of complete SVC occlusion.

The patient was scheduled for elective cardiac surgery with cardiopulmonary bypass and TEE guidance. For adequate exposure of the mass, a bicaaval approach for the cardiopulmonary bypass was chosen. Due to its unique location, there was a possibility of damage, tear or resection of part of the SVC during attempts at complete excision of the mass. Moreover, there was a risk of damage to the sinoatrial node and, therefore, cardiac arrhythmias, particularly of atrial origin. Owing to high operative risk and delicacy of vascular structure, vascular surgery was also involved during the surgical excision. During operation, there were no cardiac arrhythmias, and complete excision of the mass was successful. Intraoperatively, a lobulated 2–3 cm glistening, smooth-textured, non-pedunculated mass was found at the junction of the RA and SVC with extension into the SVC lumen. The excised mass was sent for surgical pathology, which showed mature adipose tissue consistent with lipoma (figure 3).

OUTCOME AND FOLLOW-UP

Her postoperative course was uneventful, and the patient was discharged home in stable condition a few days later.

DISCUSSION

The true incidence of primary cardiac tumours is unknown as is the incidence of CL, a specific primary cardiac tumour type. CL has been reported as 0.19 new cases per 100 000 per year in a population-based study with a 5:1 male-to-female ratio.³ It can occur in the subendocardium, subepicardium or on valves.² In contrast to myxoma, which occurs in the left atrium, lipomas mostly tend to arise from the ventricular cavity but can occur intravascularly as well. The inferior vena cava has been mentioned as a common location for intravascular lipomas; however, SVC is a very uncommon site. More commonly SVC is involved in malignancies originating in surrounding structures such as bronchogenic carcinoma.

Figure 3 Histopathology of superior vena cava lipoma. (A) Magnified low-power (10×) view showing tumour of adipose tissue with focal adjacent myocardium. (B) Magnified medium-power (20×) view showing same tumour with relatively uniform-sized adipocytes without necrosis or significant atypia. (C) Magnified high-power (40×) view showing adipose cells, without multinucleated lipoblasts (found in liposarcoma). (D) Medium-power view (20×) showing some adjacent trapped myocardium with hypertrophic changes and eosinophilic myocardial fibres with large nuclei.

To our knowledge, there are only six other cases of SVC lipoma reported in the literature (see table 1).

The age of presentation is variable; it is also not uncommon to find a CL incidentally on imaging as in our patient. The clinical presentation of SVC lipoma depends on size, presence or absence of obstructive or embolic features as with other cardiac tumours.

Our patient's SVC lipoma was partially occlusive. In such a location, SVC lipoma can lead to venous thrombosis due to sluggish flow.⁴ Although most patients remain asymptomatic and are diagnosed incidentally on CT or MRI, lipomatous masses involving the SVC can act as arrhythmogenic substrates leading to the generation of atrial tachycardia originating from SVC.⁵

Mordant *et al* proposed that it is almost impossible to make a distinction between lipoma and liposarcoma before surgical resection, which should be considered even if the patient is asymptomatic.⁶

When found on imaging, the differential of an incidental SVC mass includes a thrombus, vegetation and cardiac tumours of primary or secondary origin. Several imaging studies can help in the diagnosis of lipomas. Chest X-rays may show a widening of the superior mediastinum in cases of SVC lipomas. Thoracic CT scans can show a non-enhancing tumour with fat attenuation. MRI with fat suppression sequencing can further confirm the fatty nature of the lesion. Superior venacaval venography can demonstrate a filling defect in the SVC with development of collaterals.⁶ In addition to the defining gross appearance, transthoracic and transoesophageal echocardiography can also aid in the diagnosis of a lipoma by revealing useful tissue information like echogenicity, calcification and vascularity. Lipomas are echogenic masses without necrosis, haemorrhage or calcification. CT and MRI are highly specific in identifying fat and should be considered in cases of diagnostic uncertainty.⁷ Our patient was incidentally found to have an SVC lipoma on breast MRI, which was limited in detecting the characteristics of this lesion, in contrast to a cardiac MRI with fat suppression sequencing.

Table 1 Case reports of SVC lipoma including this case

Author	Age/gender	Clinical presentation	Diagnosis	Treatment, size of lipoma and postoperative complications
Vinnicombe <i>et al</i> ³	42 years old, F	Lethargy, mild oedema of face and hand	CXR: superior mediastinal widening CECT scan: extent of mass, fat attenuation, intraluminal extension DSV: extent of mass	Surgical excision due to SVC obstructive features 10×5×5 cm mass Uneventful postoperative course
Tanyeli <i>et al</i> ⁹	48 years old, M	RUE oedema and numbness	CECT: SVC mass of fat density CEMRV: an intraluminal mass of the SVC	Surgical excision due to symptoms 5×2 cm mass PE 2 weeks postoperative
Bravi <i>et al</i> ⁴	63 years old, M	Abdominal pain, right shoulder and lumbar pain Clinical presentation vague No direct signs or symptoms of SVC occlusion	CECT: subocclusive SVC mass and thrombosis of IVC involving the PV system Echocardiogram: SVC filling defect of intermediate echogenicity Repeat CECT after anticoagulation: persistent SVC mass with fat density and resolution of PV thrombosis MRI on FSS: confirmatory	Surgical excision of SVC mass due to thrombotic complication Uneventful recovery No recurrence
Mordant <i>et al</i> ⁶	55 years old, F	Incidental finding on CXR: superior mediastinal enlargement	CECT: intraluminal non-enhancing tumour with occlusive involvement of distal RSV, RBV and the SVC up to RA Superior vena cavography: total occlusion of RSV, RBV and SVC up to the azygous vein along with abnormal collaterals MRI: fatty intravascular lesion	Complete surgical excision 9×5 cm mass No postoperative complications No recurrence
Thorogood and Maskell ¹⁰	73 years old, M	Admitted for resection of rectosigmoid CA Preoperative CXR: superior mediastinal widening in right paratracheal area, incidental finding Multiple subcutaneous lipomas on examination No evidence of SVC obstruction	Postoperative CT chest: fat density within the lumen of SVC and the RBV MRI, FSS imaging: confirmatory for fatty nature of mass	No surgical resection being asymptomatic
Concatto <i>et al</i> ¹¹	58 years old, M	Incidental finding on unenhanced CT chest: hypodense elongated lesion with fat density within SVC No symptoms	MRI: confirmatory for fatty nature of mass	Surgical resection, pathology confirmed diagnosis
Wahab <i>et al</i>	70 years old, F	Incidental finding on breast MRI Symptoms of headaches and palpitations	TEE: partially occluding echogenic mass at SVC-RA junction extending into SVC Postanticoagulation TEE: unresolved mass	Complete surgical excision Uneventful postoperative course

CA, carcinoma; CECT, contrast-enhanced CT; CEMRV, contrast-enhanced magnetic resonance venography; CXR, chest X-ray; DSV, digital subtraction venography; FSS, fat suppression sequence; IVC, inferior vena cava; PE, pulmonary embolism; PV, portal vein; RA, right atrium; RBV, right brachiocephalic vein; RSV, right subclavian vein; RUE, right upper extremity; SVC, superior vena cava; TEE, transoesophageal echocardiogram.

Learning points

- In cases of superior vena cava (SVC) lipomas, the differential diagnosis needs to be considered in view of the specific clinical context and includes a thrombus in addition to other cardiac tumours. Overwhelming odds favouring a thrombus over a cardiac tumour in a patient with a prior malignancy led us to consider a trial of anticoagulation. Retrospectively, a D-dimer might have excluded this possibility and avoided the risks associated with anticoagulation and led to definitive treatment earlier.
- Generally, lipomas are asymptomatic but if a patient is symptomatic or there is evidence of an obstruction or suspicion of malignancy, for example, liposarcoma, it should be excised.
- Being a benign tumour, SVC lipoma is excisable and the prognosis is excellent. The recurrence of cardiac lipoma is uncommon.

Contributors AW is the primary author of this case report. He wrote the first draft of the case report and did a preliminary research of the literature. He initially wrote the draft and handed over to other authors for proofreading. He planned and organised the data. SC is the second author who contributed to the case report. He added few important articles after searching the literature and also helped in writing the discussion part. AM is the cardiologist who was actually involved in patient's management and was managing the patient throughout the course. He was aware of the minor details. The case presentation was completed with his help and was reviewed by him later on after writing it, and he viewed the technical aspects of the case with cardiology and cardiothoracic surgery being the main relevant fields. He also provided the echocardiogram pictures and also labelled them. SJS reviewed and edited the final draft. She is the programme director. She reviewed the language aspect of the case report and did fine tuning of the article. She gave the final shape to the article. She also helped in doing the technical aspects of using references.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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