CASE REPORT

Carney complex: fourth time excision of recurrent atrial myxoma via left thoracotomy

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Accepted 9 April 2014

SUMMARY

We report a case of a 51-year-old male patient with a known history of Carney complex, who was operated on for the fourth time due to recurrent left atrial myxomas. Despite surgical challenge, the operation was uneventful. The patient had a difficult postoperative period and needed further treatment for complications. After a prolonged recovery, he is currently well and remains myxoma-free up until now (36 months postsurgery) with regular follow-up using cardiac CT and transthoracic echocardiogram.

BACKGROUND

Carney complex presents mainly by myxomas, spotty pigmentation and endocrine overactivity. Carney *et al*¹ first described it in 1985.

Kirschner *et al*² identified that mutations in the PRKAR1A gene are responsible for the occurrence of Carney complex.

In this particular case, the recurrence of atrial myxomas requiring a redo excision (via left posterolateral thoracotomy) for the fourth time was picked up through regular follow-up with cardiac CT.

CASE PRESENTATION

A 51-year-old man was admitted to our cardiac centre in September 2010 with a known history of PRKAR1A mutation positive Carney complex. He had recurrent left atrial myxomas and underwent excision on three occasions in 1995, 1999 and 2000. Previous surgeries were performed via median sternotomy and on each occasion myxomas recurred at different sites: interatrial septum, mitral valve annulus and then in relation to the left atrium incision line. As a part of Carney complex he also manifested with type 2 diabetes, Cushing syndrome, hypogonadotropic hypogonadism, recurrent cerebrovascular accidents, bilateral adrenal masses and fatty liver. He was regularly followed up with biannual echocardiogram and annual CT. In 2006, he was noted to have a recurrence of myxoma at the left lateral atrial wall. Decision was made to continue follow-up in light of his anticipated risky surgery and postoperative protracted recovery. Due to its increase in size to 3 cm, the decision was made to operate on him in September 2010.

INVESTIGATIONS

Cardiac CT with contrast revealed a filling defect consistent with the left atrial myxoma seen in the left atrium adjacent to the mitral valve. Its diameter is 27 mm. This represents significant increase in

size from the CT of September 2008. The last CT was performed in 2010 confirming a 3 cm size myxoma attached to the lateral wall of the left atrium (figure 1).

TREATMENT

Redo-left atrial myxoma excision was performed through a left posterolateral thoracotomy. We adopted the left side to provide the best access to the myxoma involving the lateral wall of the left atrium. The left femoral vein was cannulated with a long transvenous cannula and the descending thoracic aorta was used to establish cardiopulmonary bypass. The patient's body temperature was cooled to 32°C and a left ventricular apical vent was inserted. Once there was no evidence of any ejection, the patient was put head down. The left atrium was opened parallel to the atrioventricular groove. Two left atrial pump suckers were introduced to clear the field. The myxoma was identified as a pedunculated mass and excised with a rim of the lateral left atrial wall.

OUTCOME AND FOLLOW-UP

Postoperatively, he developed several complications: sepsis which led to acute respiratory distress syndrome requiring reintubation with prolonged ventilation needing tracheostomy. Moreover, renal failure evolved which in turn necessitated haemofiltration. Furthermore, he was noted to have a distended abdomen; a CT scan was performed showed evidence of ischaemic bowel. He underwent a laparotomy; the necrotic small bowel loop was resected with the laparotomy incision left open for a second look operation after which the abdomen was closed. His condition gradually

Figure 1 A CT scan with contrast performed in 2010 showing a filling defect in the left atrium (arrow).

To cite: Azzam R, Abdelbar A, Yap KH, *et al*. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-201827

Figure 2 A CT scan with contrast performed in 2013 showing a clear left atrium with any filling defects.

improved before he was discharged to the district general hospital for continuing care, from where he was discharged home after 17 days.

At his regular follow-up (up to 36 months) he remains well and myxoma-free (figure 2).

DISCUSSION

Carney complex was first described by Carney *et al*¹ in 1985. The main clinical presentations are myxomas, spotty pigmentation and endocrine overactivity. Although myxomas (mesenchymal tumours) can occur at any age, they are encountered more between the third to sixth decades of life.³

Echocardiography (transthoracic or transoesophageal) is a satisfactory diagnostic imaging for myxomas. It can show precisely the site and the size of the tumour. Alternatively, cardiac CT or MRI can be used.³

Surgical excision remains the gold standard treatment in such cases that should be performed once the diagnosis is made. This urgency to operate is based on the high rate of embolisation and the danger of sudden cardiac death.⁴

Dense adhesions encountered in reoperations for recurrent myxomas render the surgery of higher risks compared with complications that would be induced merely by the myxoma.⁴

In our case, the reason for performing the surgery on the fourth occasion was because of the nature of the rapidly growing tumour, and the concern that the patient would develop obstructive symptoms. The left thoracotomy approach was chosen because the mid-sternotomy approach was used in

the previous three cardiac operations. This carried a very high surgical risk as well as the site of the myxoma making the aforementioned approach preferable.

Following myxoma resection, a six monthly imaging is essential for Carney complex follow-up. This can be done by echocardiography in most cases, or alternatively cardiac CT scan or MRI.⁵ This will help in the early diagnosis of recurrence, and therefore avoid complications such as embolism, stroke, intracardiac obstruction and ventricular outflow tract obstruction.³

Learning points

- ▶ Left thoracotomy is a valid approach for redo myxoma resections associated with dense adhesions.
- ▶ Cardiopulmonary bypass with moderate hypothermia with left ventricle venting can be safely used in similar cases.
- ▶ Regular follow-up imaging is mandatory to detect early recurrences before being symptomatic.
- ▶ Multidisciplinary team discussions to tailor decisions for similar cases carrying high surgical risks.

Acknowledgements The authors would like to thank Mr Ragheb Hasan for his support to them for producing this manuscript.

Contributors All the authors contributed to data collection, writing and editing of this manuscript.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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