Role of pre-operative assessment in the surgical management of leiomyoma extended to the right heart chambers: a compendium of information from isolated reports

F. Roques a,*, B. Sanchez a, B. Bucher c, J. Larivière b

Department of Cardiac Surgery, Fort de France University Hospital, 97200 Fort de France, Martinique
Department of Cardiovascular Anaesthesiology, Fort de France University Hospital, 97200 Fort de France, Martinique
Clinical Pathology Laboratory, Fort de France University Hospital, 97200 Fort de France, Martinique

Received 18 August 2000; received in revised form 10 January 2001; accepted 25 January 2001

Abstract

Recurrent intravenous leiomyoma extending to the right heart chambers is extremely rare. A large range of surgical techniques and approaches (i.e. two-step procedure, hypothermia and circulatory arrest) have been previously described. We report a recent case where the tumour was excised in a one-step procedure under normothermic cardiopulmonary bypass. This report associated to a comprehensive literature review allows us to discuss the role of pre-operative assessment and to propose refinement of surgical techniques according to the anatomy of the tumour.

Keywords: Heart neoplasms; Leiomyomatosis; Uterine neoplasms; Vascular neoplasms; Vena cava inferior

1. Introduction

Intravenous leiomyoma is a rare non-malignant tumour arising from the uterine vein wall or an uterine leiomyoma infiltrating intravenously and spreading into the venous cavity [1]. The tumour growth is slow. Extension through the inferior vena cava (VC) to the right heart chambers can take many years and is extremely rare. We report a case where the tumour was excised in a one step procedure under cardiopulmonary bypass (CPB). A compendium of this and previous reports allowed us to sort out anatomic information that can be used to develop a therapeutic strategy.

2. Case report

A 53-year-old woman was admitted in our institution for multiple aborted syncopes. Hysterectomy without right omentectomy was performed 25 years earlier.

On trans-thoracic echocardiography, a tumour of the right atrium (30 mm-diameter) was discovered. On trans-oesophageal echocardiography, it appeared multilobular in shape, mobile, prolapsing through the tricuspid valve into the right ventricle. It seemed appended to the free wall of the right atrium. A myxoma was therefore suspected. The patient experienced a syncope during echography and was promptly operated. Following midline sternotomy; the right atrium was manually inspected; the tumour occupied most of the atrium. A canula could not be inserted into the inferior VC via the atrium. Canulation of the superior VC and the left femoral vein was therefore required for CPB. Under normothermic CPB, a vertical atriotomy was performed on the beating heart. A yellowish, hook-shaped tumour appeared extending to the atrium, most of the functional area of the tricuspid valve and the inlet right ventricle. It was only attached by a thin pedoncule to an unseen site, upstream in the inferior VC. Blind traction on the tumour seemed dangerous; a laparotomy was performed. The tumour arose from the right ovarian vein. A 1 cm square venous patch surrounding the ostium was excised and the tumour was divided on site (Fig. 1). Since the upper part of the tumour was not attached to the heart, it could immediately be withdrawn through the open atrium (Fig. 1). The VC was closed using an autologous pericardial patch. CPB was discontinued; the remaining extra-cava part of the tumour (ovarian pedoncule and right appendage) was excised. Histology demonstrated a leiomyoma. No recurrence was shown on CT scan, 2 years later.
Fig. 1. Illustration of the surgical excision of a leiomyoma of the vena cava extending to the right heart chambers. The tumour is divided at the site of attachment (1), allowing excision of its cavo-cardiac part through the open atrium (2).
3. Discussion

Leiomyomatosis extending to the right chambers of the heart is a very rare condition (no more than 30 cases reported [1–9]). A large range of techniques have been utilised for tumour excision: withdrawal (through the iliac vein [2] or the opened atrium [3]) can be performed in a one [4] or two-stage procedure [5], under normothermic CPB [6] or circulatory arrest [4,5,7]. Several factors may explain this variety. First, because of poor documentation on these tumours (insufficient knowledge of tumour anatomy or erroneous pre-operative diagnosis of myxoma), improvisation can occur. Second, surgical strategy may be guided by ‘analogous thinking’ from more experienced surgical situations. Removal of an extension to the VC of a kidney neoplasm for instance often requires a complete circulatory arrest. Similarly, this strategy may be chosen for leiomyoma. In our case, 10 min of CPB time were required to withdraw the tumour, suggesting that circulatory arrest and associated risks can be avoided.

The information gathered from a comprehensive literature review can be summarised as follows:

- The diagnosis of leiomyoma extending to the right heart must be raised when facing a middle age woman with a right heart tumour, especially in case of previous hysterectomy. CT scan provides the best diagnostic information, since it can demonstrate an intravenous tumour with ovarian v internal iliac vein as well as VC and right atrium involvement.
- Removal of the tumour has always been considered easy and safe when a double approach (thoracic/abdominal) was used.
- The tumour has always been found attached to the vascular bed at one site only (the ostium of the internal iliac vein [2,3] or the ovarian vein [7]).
- Since the portion of the tumour located in the atrium is most of the time larger than the other segments, it has never been possible (except in one case [2]) to remove it by pulling downward from an incision in the VC. An approach from the atrium under CPB is therefore mandatory.
- Upward traction of the tumour in a blind fashion for removal through a single atrial opening must be avoided since it may lead to tears in the VC and life-threatening retroperitoneal haemorrhage [8].

CT scan or MRI information required in order to determine a surgical strategy are:

1. A comparison of the size of the tumour at the atrium level vis-à-vis the diameter of the inferior VC.
2. The site of attachment of the tumour to the VC, and
3. The side and portion of the iliac and femoral vein remaining free of the tumour which can be used in order to provide the best venous drainage during CPB.

Based on the above the following strategies can be proposed:

- If the diameter of the part of the tumour located in the heart is smaller than the diameter of the vena cava, a simple laparotomy will then allow the tumour to be removed by downward traction exercised from the iliac vein or the vena cava [2].
- In all other cases, an abdominal and thoracic approach is mandatory; the tumour divided from the CV, must be withdrawn through the open atrium under CPB with separate superior and inferior VC drainage. Since this is a ‘quick and easy’ procedure a circulatory arrest is not necessary; and a right mini-thoracotomy can be preferred to a sternotomy.

4. Conclusion

A suspected diagnosis of leiomyomatosis in a middle age woman presenting with a tumour of the right heart can be confirmed by CT scan or MRI. Moreover, these exams provide the anatomical information (pertaining to the tumour size and attachment) required in order to refine surgical strategy. Complete surgical excision can be easily achieved under CPB through a minimally invasive normothermic procedure on the beating heart.

References