CASE REPORT

Multiple Lambl's excrescences with subvalvular extension, a rare cause of cryptogenic stroke: treated by port-access cardiac surgery

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SUMMARY

A young male patient was evaluated for a cardiac cause of ischaemic stroke. In the work-up, a tumour was detected on the subvalvular apparatus of the mitral valve for which operative removal was scheduled. A port-access procedure was applied as a novel way of treatment. On thoracoscopic intracardiac inspection, however, multiple tumours were seen attached to the chordae tendineae and to the anterior leaflet of the mitral valve. It was decided to not to perform a plasty but to remove the valve in toto and insert a mechanical valve prosthesis. The tumours were diagnosed as multiple giant Lambl's excrescences.

CASE PRESENTATION

Apart from a history of one similar attack 2 months ago, a previously healthy 33-year-old man presented to the emergency department with transient symptoms of dysphasia and dizziness. The patient had always been at sinus rhythm. There was no previous hospitalisation with no conclusive medical or family history. As blood tests showed no infection, coagulopathy or vasculitis, and the patient did not appear to be ill, he was discharged and scheduled for further examination.

INVESTIGATIONS

A CT scan and MRI of the brain showed a recent ischaemic area in the right frontoparietal region and the anterior part of the left thalamus. In order to evaluate a possible cardiac source of embolism echocardiography was performed. Transthoracic

BACKGROUND

- Novel way of treatment; port-access mitral valve replacement.
- Multiple Lambl's excrescences are rarely described in literature. In addition, to our knowledge, *no report* has been described multiple lesions that involved a relatively wide and mainly subvalvular area.

Figure 1 Transoesophageal echocardiography: solid tumour (arrow) attached to the subvalvular apparatus of the mitral valve.

Figure 2 Four small intercostal incisions are being made between the mid-axillary and mid-clavicular line.

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Figure 3 Endoscopic surgery.

echocardiography (TTE) and transoesophageal echocardiography (TOE) revealed a solitary tumour (6×9 mm) attached to the chordae tendineae of the anterior leaflets of the mitral valve (figure 1).

DIFFERENTIAL DIAGNOSIS

Initially, the list of differential diagnosis included also other causes of juvenile stroke as patent foramina ovale, atrial fibrillation, vascular malformation at the base of the brain, coagulation and blood disorders.

TREATMENT

Since the patient had suffered two clinical episodes of ischaemic stroke, he was scheduled for cardiac surgery to remove the tumour through a *port access* (figures 2 and 3). Cardiopulmonary bypass was accomplished by double percutaneous cannulation of the right femoral vein and the right internal jugular veins, while the arterial cannulation was performed through the left femoral artery. Exposure of the heart was achieved by an endoscopic approach through four small incisions (figures 2 and 3) as usual in the port-access procedure.

Once the left atrium was opened, a round-shaped tumour was identified. However, further inspection revealed multiple small-sized tumours situated along the full size of the chordae tendineae of the anterior leaflet of the mitral valve (figure 4). While, it seemed not possible to remove all the tumours selectively, the decision was made to replace the mitral valve after total chordae tendineae excision. The pathological examination of the excised

Figure 4 Multiple small tumours situated along the chordae tendinae.

tumours revealed its origin to be that of giant Lambl's excrescences (figure 5).

OUTCOME AND FOLLOW-UP

The procedure and postoperative recovery were uneventful.

Figure 5 Histology of Lambl's excrescences: tumours situated along the chordae tendinae.

DISCUSSION

Lambl's excrescences are rare cardiac structures, mostly described as filiform fronds usually occurring at sites of valve closure with a potential for embolisation. These tumours may be an incidental finding¹ or can be a rare cause of stroke,^{2–5} or coronary artery obstruction.⁶ Histologically, they are composed of a cellular fibroelastic core covered by a layer of endothelial cells, and have to be distinguished from cardiac myxomas⁷ and fibroelastomas,^{8–9} of which the latter had been reported to be presented as ischaemic stroke.^{10–11}

In general, the larger the tumour, the more likely it is that its presence will be detected, TOE being superior over TTE. However, as this case shows, small tumours, especially those located subvalvular, can be very difficult to visualise by echo.¹² In this patient, only one tumour on the chordae tendineae was observed by TOE, while during surgery multiple smaller tumours were detected.

Multiple Lambl's excrescences are rarely described in the literature. In addition, to our knowledge, no report has described multiple lesions that involve a relatively wide and mainly subvalvular area. This could be due to the fact that the spatial resolution of echocardiography is too low to detect these smaller lesions. Moreover, during conventional open heart surgery, small tumours which are situated just under the valve leaflets may be easily missed, while in the port-access setting, the visual exposure of the chordae tendineae is much better.

What is also remarkable in this patient was the site of these tumours. Lambl's excrescences are mostly described as preferably located at the sites of valve closure,^{1–13–14} and extremely rare at the papillary muscles.¹⁵ To our knowledge, multiple lesions situated along the chordae tendineae of the mitral valve were not described before. The current hypothesis is that wear and tear damage to the endothelium occurs at the site of contact between the valvular margins and the endocardial surfaces. Subsequently, small thrombi evolve and form the filiform fronds of Lambl's excrescences.^{16–17} The diffuse multiple lesions seen in this patient suggest that this hypothesis cannot be the sole explanation to the pathogenesis of these tumours.

Competing interests None.

Patient consent Obtained.

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Learning points

Lambl's excrescences can present as multiple lesions involving not merely the valvular but also the subvalvular regions of the heart. Owing to its small size and uncommon location, these lesions can be difficult to visualise by echocardiography. Therefore, a careful examination during surgery should be performed to evaluate a possible extravalvular involvement. Endoscopic surgery seems to provide a superior visualisation compared with the standard approach.

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