Libman-Sacks Endocarditis

Synonyms: verrucous endocarditis

In 1924 Libman and Sacks published an account of atypical, sterile, verrucous vegetations on the heart in systemic lupus erythematosus (SLE). \[1\] Clusters of verrucae, looking like mulberries, were described on the ventricular surface of the posterior leaflet of the mitral valve and found at postmortem. The leaflet and the chordae tendinae are often adherent to the endocardium of the ventricular wall.

The disease is often missed at echocardiography. The spectrum of the disease has changed since the original description, as steroids and other treatments have changed the course of the disease. Life expectancy, whilst limited, is much higher than it was. The left side of the heart is affected more often than the right but even the endocardium can be affected. Regurgitation is more frequent than stenosis.

Although the condition is classically associated with SLE, it can also occur in antiphospholipid syndrome (APS), \[2\] whether primary or secondary. High antibody titres are associated with high risk. \[3\]

Epidemiology

Libman-Sacks’ vegetations can be found in approximately 1 of 10 patients with SLE. They are associated with lupus duration, disease activity, antiphospholipin antibodies and APS manifestations. \[4\] The incidence of positive findings on echocardiography is variable but detection is higher with transoesophageal ultrasound and the frequency is higher in those with positive antiphospholipid antibodies. \[5\] Thickening of the leaflets is more common than finding vegetations. Abnormalities on ultrasound may be found in around a third with APS.

The presence of these findings, as with the disease, is 5 to 9 times more frequent in women than in men. The typical patient is a young woman, although it can rarely occur in children.

History

- Most patients with Libman-Sacks endocarditis are asymptomatic.
- If valves are severely affected there may be features of the valve disease. Mitral valve disease is more common than aortic valve disease. Regurgitation is more frequent than stenosis and involvement of the tricuspid or pulmonary valves is unusual.
- Systemic embolism may occur with effects depending upon the destination of the emboli but brain and kidney are likely victims. Emboli can cause blockage of the peripheral circulation.
- The vegetations of Libman-Sacks endocarditis are sterile but secondary infective endocarditis can occur.
- There may or may not be typical features of SLE with the characteristic butterfly rash, fever and arthritis or features of APS, including recurrent miscarriage.

Examination

- Physical signs reflect the pathology and can be found in the various articles on mitral valve disease or aortic valve disease, infective endocarditis and, if the valve disease is severe, congestive heart failure.
- There may also be left ventricular hypertrophy, causing displacement of the apex beat.

Investigations

- Echocardiography should be employed, although not all lesions will be detected. Results with transoesophageal echo are superior but it is an invasive procedure.
- Blood culture is important to exclude infective endocarditis (IE), which may coexist.
- Investigations for SLE, including antiphospholipid antibodies and other autoantibodies. False positive serology in the form of VDRL is common in SLE and anticardiolipin antibodies increase the risk of cardiac abnormalities. \[6\]
- FBC may show raised neutrophils and some anaemia.
- CXR may show cardiomegaly and pulmonary congestion if disease is severe. Calcification of lesions is uncommon.
- If valvular disease seems severe then cardiac catheterisation may be required with a view to valve replacement.

Management
Historical

- Emanuel Libman[14] and Benjamin Sacks [15] (or Sachs) described an unusual type of endocarditis in 1924, non-bacterial, with verrucous vegetations adherent to the endocardium. Dermatologists had earlier described the disease associated with facial rash and fever, but Libman and Sacks presented the first complete clinical picture, with and without skin lesions.

- Libman was at the time assistant pathologist at Mount Sinai hospital in New York. He was an eccentric generalist, who founded the cardiology department, and made important contributions in bacteriology and teaching. He diagnosed the fatal endocarditis of Gustav Mahler, and said of Franklin D Roosevelt in 1944: "It doesn't matter whether Roosevelt is re-elected or not, he'll die of a cerebral haemorrhage within 6 months." After the event, he was asked how he knew: "I only saw him in the newsreels, but I've seen that special wasting look many times. He couldn't last 6 months."

- APS (Hughes' syndrome, or as he first named it - antiphospholipid syndrome), was described by Graham Hughes in 1983, characterised by venous and/or arterial thrombosis, recurrent pregnancy loss, and the presence of antiphospholipid antibodies. Graham Hughes, rheumatologist at St Thomas' Hospital, was the first non-American to be elected to the American Lupus "Hall of Fame" in 1989, and set up the first dedicated Lupus outpatient facility outside the USA in 1997, the 'Louise Coote Lupus Unit' based within St Thomas' Hospital.

- The association between Libman-Sacks endocarditis and APS was first noted in 1985.

Prognosis

All patients with SLE have a shorter life span; cardiovascular disease is a common cause of death.

Complications

- Systemic emboli may occur but they are probably not very common.
- The risk is much higher with mitral stenosis and subsequent atrial fibrillation.
- When strokes occur it is difficult to know if they were due to systemic emboli or the underlying pathology of SLE or APS.
- Valvular disease can lead to heart failure.
- Maternal SLE with anti-Ro/SS-A (Sjögren's syndrome antigen A) autoantibodies is associated with congenital heart block in the baby in about 1 or 2%. It is usually complete but can be 1st or 2nd degree. The rate of recurrence is around 16%.
- Fluorinated steroids that do not cross the placenta may be beneficial.[13]

Further reading & references

- Ren X; Libman-Sacks Endocarditis; emedicine. September 2008.
14. whonamedit.com; Emanuel Libman; Brief biography
15. whonamedit.com; Benjamin Sacks; Brief biography

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