Splenomegaly in Silent Endocarditis

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A 33-year-old woman with Williams syndrome had a fever of unknown origin that persisted for more than four months. The patient visited a hospital and underwent systemic investigations. Computed tomography detected splenomegaly (96 × 58 mm), and positron emission tomography revealed diffuse uptake in the spleen (Figure 1), for which a definitive diagnosis was uncertain and the patient was referred to us for further examination. At the first visit, her general condition was relatively good and the patient had no symptoms other than fever. A systolic murmur was auscultated but it was considered to be due to the underlying disease of Williams syndrome. However, immediately after hospitalization, she complained of a dry cough, although her lungs were clear on radiography. Our differential diagnosis included primary splenic lymphoma, and splenectomy was planned for the pathological examination. Preoperative transthoracic echocardiography incidentally detected severe mitral regurgitation, along with chordae tendineae rupture and vegetation formation at the mitral valve (17 mm). Under a diagnosis of infective endocarditis, the patient was sent for emergent mitral valve plasty. Both blood cultures drawn before the cardiac operation and the resected valve were positive for ampicillin-resistant *Enterococcus raffinosus*; the patient was treated with a 6-week combination of vancomycin and gentamicin. The patient had a good postoperative clinical course without any complications.

This was a case of native-valve infective endocarditis that progressed silently, and splenomegaly was the only clinical sign. An inquiry regarding the details of investigations at the former hospital showed that blood culture and echocardiography were not performed. Splenomegaly is reportedly observed in cases of infective endocarditis¹, and accordingly, we highlight that the disease should be included in the differential diagnosis of splenomegaly, particularly in cases with fever of unknown
origin. The present case was especially characterized by her underlying genetic disorder of Williams syndrome, in which congenital cardiovascular anomalies are frequently observed. Thus, the possibility of infective endocarditis should have been investigated earlier.

*E. raffinosus* is an emerging pathogen that causes various infections, including endocarditis. So far, three cases of *E. raffinosus*-associated endocarditis have been described in the literature, and the present case is the fourth such case. The organism can possess the *van*A gene, providing resistance to glycopeptides; thus, it may become a difficult-to-treat pathogen.

Finally, we again underline that splenomegaly is a potential sign of infective endocarditis that develops silently in a patient.

**Ethics Statement:**

Informed consent was obtained from the patient’s family for the publication of this case report.

**References**


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**Figure legend**

**Figure 1.** Computed tomography and positron emission tomography of the splenomegaly found in the patient with silent endocarditis. The spleen size was 96× 58 mm. No other remarkable findings were noted.
Figure 1. Computed tomography and positron emission tomography of the splenomegaly found in the patient with silent endocarditis. The spleen size was 96 x 58 mm. No other remarkable findings were noted.