

Case report

Echocardiographic findings in patients with temporal arteritis: apropos of one case of temporal arteritis-associated verrucous (Libman-Sachs) endocarditis

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ABSTRACT

We report the case of a 70-year-old patient who presented with fever of unknown origin. The initial diagnosis was infective endocarditis as a mitral valve vegetation was found in a transesophageal echocardiogram. Lack of response to empiric antibiotic treatment and further diagnostic work-up established the correct diagnosis of marantic endocarditis due to temporal arteritis. Treatment with steroids and aspirin led to rapid clinical improvement and disappearance of the vegetation. Apropos of this case, we reviewed the records of 25 patients with a new diagnosis of temporal arteritis and analyzed the echocardiographic findings in comparison to those of 40 age- and sex-matched controls. Abnormal echocardiographic findings were present in 13 (52%) out of 25 patients with temporal arteritis and in 5 (12.5 %) out of 40 controls ($p < 0.001$, chi-square test).

Introduction

Temporal arteritis is a chronic inflammatory disease of large and medium sized vessels with an estimated annual incidence of 15-25 new cases per 100,000 persons in high-risk populations (1). Besides its usual presentation with fever, new headache, visual disturbances, jaw claudication and polymyalgia, the disease has been associated with a plethora of clinical signs and manifestations. In this respect, we present a case of marantic endocarditis in a temporal arteritis patient and we report the echocardiographic findings in a series of sequential, non-selected patients with temporal arteritis, compared to normal controls.

Case report

We present a case of a 70-year-old man who was transferred to our hospital

with the diagnosis of "culture-negative endocarditis" for further work-up and treatment. He had a 3-month history of fever (up to 38.5 °C), headache, non-productive cough, fatigue, anorexia, a 3-kg weight loss, night sweats and myalgias.

He was initially admitted to another hospital where he had an extensive work-up that revealed normocytic anemia (hematocrit: 32%) with elevated indices of inflammation (ESR: 72 mm/1st hour and CRP: 142 mg/l, normal < 5). Chemistry panel was within normal limits. Serum protein electrophoresis showed polyclonal hypergammaglobulinemia. Multiple blood and urine cultures were sterile. An abdominal CT scan showed splenomegaly. A transthoracic echocardiogram revealed a 4mm x 4mm vegetation on the anterior leaflet of the mitral valve that was not present in a similar test performed a year earlier. After the patient had been treated with intravenous ceftriaxone (2 gr bid) and gentamycin (80 mg tid) for 2 weeks, without any improvement, he was transferred to our department.

On admission, physical examination revealed a body temperature of 38 °C, a pitting edema of the lower extremities and tenderness on palpation of both shoulders and thighs. Antibiotics were discontinued and a transesophageal echocardiogram confirmed the presence of a vegetation of 11mm x 8 mm on the middle part of the anterior leaflet of the mitral valve causing mild regurgitation (Fig. 1). Multiple blood cultures were negative. There was no evidence of antiphospholipid antibodies. A temporal artery biopsy showed infiltration of the adventitia and medial wall by histiocytes, lymphocytes and polymorphonuclear white blood cells, fragmentation of the internal elastic lamina

and partial obliteration of the vessel lumen, findings compatible with the clinical diagnosis of temporal arteritis. The patient was treated with prednisone (initial dose of 60 mg/day with gradual tapering) and 80 mg/day of aspirin and his symptoms resolved rapidly. A year later he remained in good condition and free of symptoms, while a follow-up transthoracic echocardiogram showed a mild mitral regurgitation and no evidence of vegetations.

Case series

As a PubMed search with the terms “temporal arteritis” or “giant-cell arteritis” and “valvular disease” or “echocardiogram” did not reveal any relevant study, we analyzed the records of the patients with the coding diagnosis of “temporal arteritis” in our department.

Patients and methods

Forty-one of 46 (89%) records were available for review (including our index case patient). Transthoracic echocardiography had been performed in 25 patients (22 women and 3 men, with a mean age of 70.6 years; range: 52-85 years). They were consecutive, unselected patients with newly diagnosed temporal arteritis, who underwent echocardiography because they were included in another prospective study of the effect of temporal arteritis on arterial distensibility (2). None of them had symptoms or signs of cardiac disease nor were they on cardiologic medications. As controls, we used 40 age- and sex-matched patients (26 women and 14 men, with a mean age of 73.1 years; range 55-85 years) who were admitted to the ophthalmology service for cataract surgery and underwent an echocardiogram before the operation. The echocardiograms were performed with the same technical equipment (Hewlett-Packard, Sonos 1000 System; Andover, MA; equipped with a 2.5-mHz transducer) by the same cardiologist. All patients participating in our study were fully informed and provided written consent. The study was approved and authorized by the Scientific Board of Athens University Medical School Clinical-Pathological Division and conforms to stan-

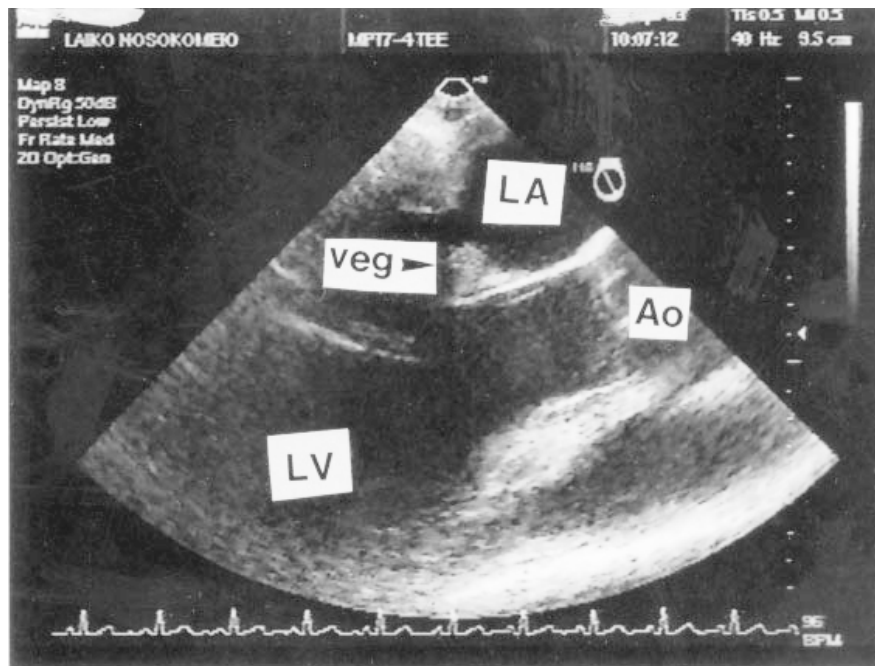


Fig. 1. Transthoracic echocardiogram at 115 degrees showing a vegetation attached to the middle part of the anterior mitral leaflet on the atrial side. LV: left ventricle; Ao: aorta; LA: left atrium, and veg: vegetation.

Table I. Echocardiographic findings in 25 unselected, consecutive patients with temporal arteritis and in 40 age- and sex-matched controls.

| Findings* | Patients (%) | Controls (%) |
|---|--------------|--------------|
| Normal | 12 (48%) | 35 (87.5%) |
| Left ventricular systolic dysfunction | 4 (16%) | 0 |
| Global | 3 (12%) | |
| Regional | 1 (4%) | |
| Mitral valve regurgitation (mild) | 6 (24%) | 3 (7.5%) |
| Aortic valve thickening/calcification** | 4 (16%) | 2 (5%) |
| Aortic valve stenosis | 1 (4%) | 1 (2.5%) |
| Aortic valve calcification combined with aortic valve insufficiency | 2 (8%) | 1 (2.5%) |
| Aortic root dilatation | 9 (36%) | 1 (2.5%) |
| Mild | 8 (32%) | 1 (2.5%) |
| Moderate | 1 (4%) | 0 |
| Aortic and mitral valve regurgitation | 3 (12%) | 0 |
| Tricuspid valve regurgitation (mild) | 1 (4%) | 1 (2.5%) |
| Pulmonary hypertension (mild) | 3 (12%) | 0 |
| Pericardial effusion (mild) | 4 (16%) | 1 (2.5%) |
| Marantic endocarditis | 1 (4%) | 0 |

* The sum of echocardiographic findings exceeds the number of subjects as one individual could have more than one valvular abnormality.

** Only cases with significant calcification causing valvular dysfunction were included in the analysis.

dards defined by our University authorities.

Results

Abnormal echocardiographic findings were present in 13 (52%) out of 25

patients with temporal arteritis and in 5 (12.5%) out of 40 controls (p < 0.001, chi-square test) and are shown in Table I. The most common findings were mild to moderate aortic and mitral valve regurgitation, aortic valve

thickening and calcification, and aortic root dilatation. Only cases with significant calcification causing valvular dysfunction were included in the analysis.

Discussion

To our knowledge our report is just the second of temporal arteritis associated with marantic (Libman-Sachs) endocarditis (3). We also report on a small case series of echocardiographic findings in patients with temporal arteritis. Libman and Sachs added verrucous endocarditis to the clinical spectrum of SLE in 1923, but its pathogenesis still remains obscure. In a series of 69 patients with SLE valvular vegetations were found in 30 (43%) (4). In contrast to the high prevalence of sterile valvular vegetations in SLE patients, vasculitis-associated marantic endocarditis is a rather rare condition, with only 3 reported cases in the English literature (3, 5, 6). Hesselink *et al.* (3) reported the first case of marantic endocarditis associated with giant-cell arteritis.

Our analysis showed that abnormal echocardiographic findings were present in 13 (52%) of the 25 patients and in only 5 (12.5%) of the 40 age- and sex-matched controls. The most common findings were mild to moderate aortic and mitral valve regurgitation, aortic valve thickening and calcification. Although inflammation of the aortic root with aneurysmal dilatation and widening of the aortic valve ring and subsequent aortic valve regurgitation is a well-recognised entity in temporal arteritis (7), there is a paucity of data regarding the association of temporal arteritis with valvular pathology.

The association of the various vasculitides with specific echocardiographic findings has not been studied as extensively as one would expect. Valvulitis has been described in Takayasu arteritis (8), polyarteritis nodosa (9), rheumatoid arthritis (10), Wegener's granulo-

matosis (11), relapsing polychondritis (12), Cogan's syndrome (13) and antiphospholipid syndrome (14).

The quite high prevalence of abnormal echocardiographic findings in our study population cannot be explained solely by the advanced age and the associated comorbidities, since it was significantly higher compared to age- and sex-matched controls. Although the relatively small number of patients does not allow definite conclusions to be drawn, our data suggest that the primary disease might play a role in the pathogenesis of the valvular lesions. Macrophages and T lymphocytes have been implicated in the pathogenesis of vascular injury in giant-cell arteritis through the production of proinflammatory cytokines, matrix metalloproteinases and growth and angiogenic factors (1); the documented presence of these cells in the stroma and subendothelial layer of the valve leaflets (15) implies that the cellular substrate for the initiation of valvular destruction exists.

The clinical significance of our findings remains to be determined. Marantic endocarditis is usually asymptomatic but the vegetations can occasionally get infected or produce emboli. Characteristically, in the above mentioned echocardiographic study of SLE patients (4), after a mean follow-up of five years, the combined incidence of stroke, peripheral embolism, heart failure, infective endocarditis, and death was 22 percent. Therefore, periodic evaluation of temporal arteritis patients with echocardiogram is worth considering.

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