Eur Arch Psychiatr Neurol Sci (1985) 234:404–407

European Archives of Psychiatry and Neurological Sciences

Hemichorea Associated with Varicella-Zoster Reinfection and Endocarditis

A Case Report

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Summary. A 20-year-old woman developed transient right-sided hemichoreatic movements after household exposure to varicella-zoster. Some days before the appearance of involuntary movements a vesicular rash had occurred. About 6 months later an elevated IgG serum titer against varicella virus was found and two-dimensional echocardiography showed signs of an endocarditis. During the following 2 months the IgG value returned to within the normal range and the choreatic movements disappeared almost totally. The possibility is discussed that endocarditis had been caused and maintained by serum antibodies to varicella-zoster virus which cross-reacted with valvular tissue. Embolization to the region of the left striatum and/or postinfectious encephalitis in this region are assumed to be the most plausible causes of the transient hemichorea.

Key words: Varicella-zoster – Hemichorea – Endocarditis

Introduction

Varicella-zoster virus infection may involve not only skin and mucous membranes but also many other organs such as esophagus, stomach, intestines, liver, pancreas, adrenal glands, kidneys, ureters, testes, submaxillary and parotid glands, lymph nodes, spleen and lungs. In addition, there are reports on varicella-zoster infection of the heart and the central nervous system. Heart involvement was that of pericarditis (Kaplan and Tully 1953; Mandelbaum and Terk 1959), myocarditis (Fitz and Milejohn 1956) and endocarditis (case 24 of the Massachusetts general hospital 1963). Neurological sequelae of varicella-zoster infection are encephalitis mostly seen with chickenpox (Appelbaum et al. 1953; Johnson and Milbourn 1970), sensory and motor neuropathies commonly observed with herpes zoster (Thomas and Howard 1972; Reichmann et al. 1978). Rare complications are neurologic hemisyndromes due to vascular disorder (Gilbert 1974; Kolodny et al. 1968; Rosenblum and Hadfield 1972).

The case reported here is considered noteworthy since both the heart and the brain were affected after reinfection of the patient; a transient hemichorea was the leading clinical symptom while cardiac signs were absent.

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Case Report

In December 1982 the father of our patient developed herpes zoster. About 1 to 2 weeks later the patient, a 20-year-old, right-handed woman, developed a vesicular rash which spread over the whole body. She noticed involuntary "shaking" of the right arm 2 weeks later, and 6 weeks later of the right leg. The "shaking" was accompanied by restlessness and was followed by involuntary myoclonic-like movements on the right side of the face. On neurologic examination there were myoclonic-like movements predominantly in the right arm and hand, and the patient complained of paraesthesiae of digits 1 and 2 of the right hand. All other examinations including EEG, ECG, and routine laboratory examinations were normal. The patient received 3 mg clonazepam/day with some effect.

In May 1983, the patient was hospitalized in the Neurologic Clinic of Mainz because her condition had worsened during the preceding weeks. There was mild motor aphasia and a right-sided hemisyndrome with slightly increased stretch reflexes, questionable positive Babinski's sign, impairment of the skilled movements, and mild paresis of the arm and leg. There were choreo-athetotic hyperkinetic movements of the arm and leg muscles and myoclonic-like jerks of the face

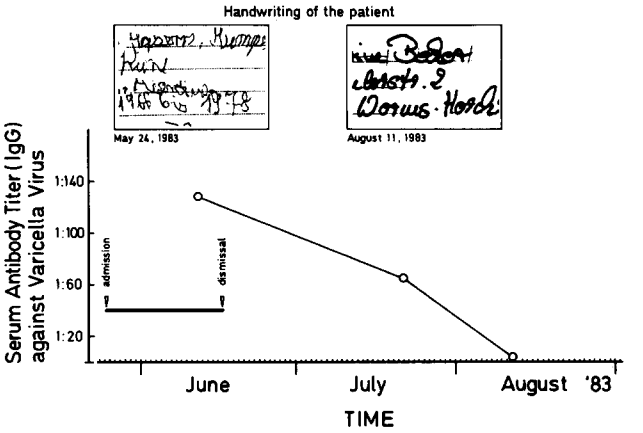
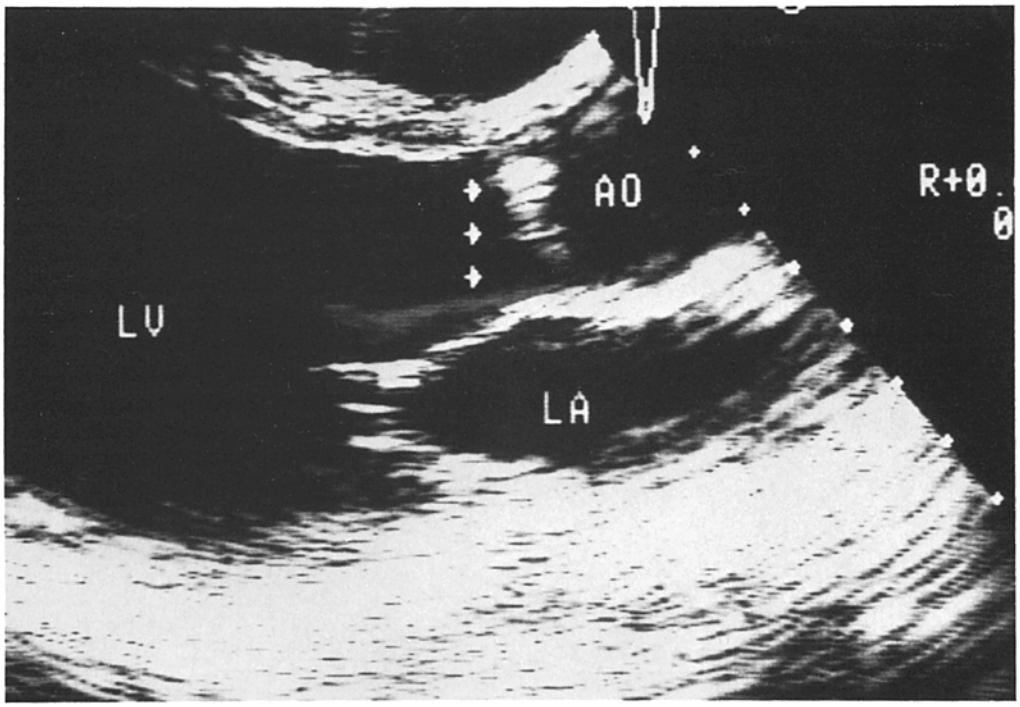


Fig. 1. Course of the serum antibody titer (IgG) to varicella virus compared with the handwriting as a parameter of the choreatic movements. The antibody titer was determined by means of complement binding reaction. The antigen was obtained from M. A. Bioproducts, Walkersville, MD, USA



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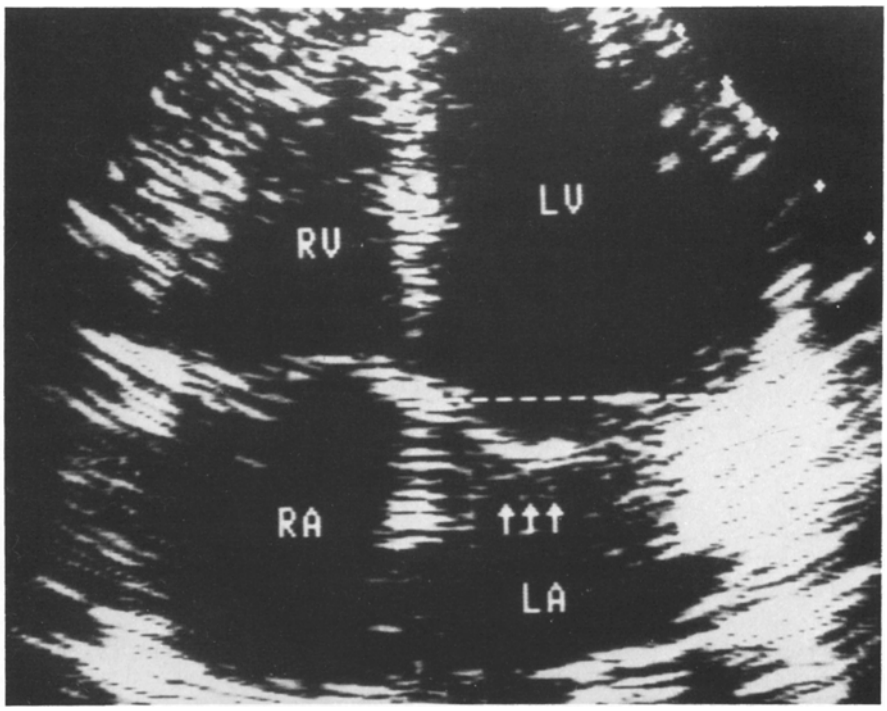

b

Fig. 2 (A). Two-dimensional echocardiogram. Left-sided parasternal longitudinal section. *Ao* = aorta, *LV* = left ventricle, *LA* = left atrium. The *three arrows* point to the aortic valve which is thickened. **(B)** Two-dimensional echocardiogram. Apical view of the four chambers. *RV* = right ventricle, *LV* = left ventricle, *RA* = right atrium, *LA* = left atrium. The mitral valve shows a pronounced prolapse *(three arrows,* – – – – = normal systolic level)

muscles that did not change much during voluntary movement.

Body temperature was never raised above 37°C, and blood pressure was between 90/55 and 110/80 mm Hg. Blood cell counts and the erythrocyte sedimentation rate were normal. The anti-streptolysin titer was below 200 u/ml (negative); anti-mitochondrial (AMA), and anti-nuclear anti-bodies (ANA) were negative. Immunoglobulins G, A, and M were within normal ranges, and serum electrophoresis was inconspicuous. Na^+, K^+, Ca^{2+}, blood glucose and serum enzymes as well as a blood clotting test were normal. Serum cholesterol and triglycerides were not elevated and T_3 and T_4 concentrations were normal, CSF was normal except for a low protein content (18 mg/100 ml). Five aerobic and anaerobic blood cultures, respectively, did not show any growth of bacteria. The IgG antibody titer against varicella virus on June 1983 was, however, increased to 1:128, this titer decreased up to August 1983, to 1:2 (Fig. 1). IgM antibodies to varicella were not found.

X-rays of the head were normal; chest X-rays suggested a disorder of the heart valves which was verified by echocardiography demonstrating aortic regurgitation, soft thickenings of the valves (Fig. 2A, 3C) and a prolapse of the mitral valve (Fig. 2B, 3B). Further examinations including ECG, blink-reflexes to electrical stimulation, pattern-induced visual evoked potentials, and computer tomography (CT) of the brain

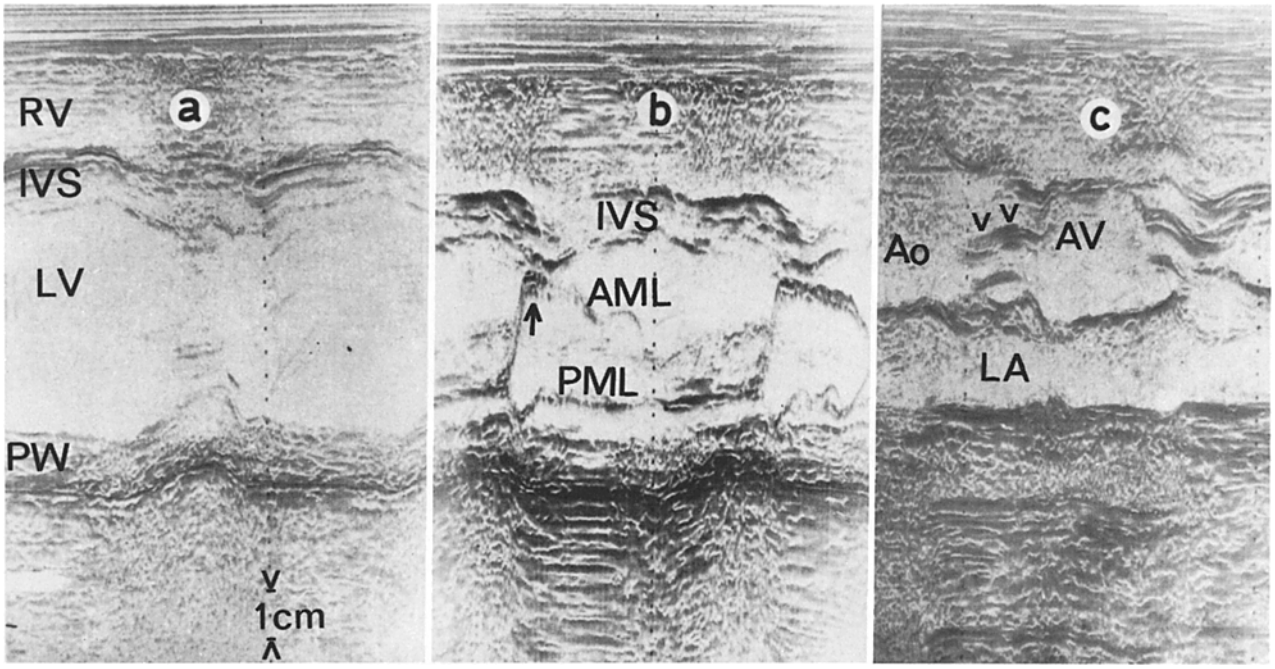

Fig. 3a–c. M-mode echocardiogram. (**a**) M-mode echocardiogram of the left ventricle *(LV)* recorded from the fourth intercostal space. Recorded is the movement of the interventricular septum *(IVS)* and the posterior wall *(PW)*. During contraction interventricular septum and posterior wall illustrated normal movement. Right ventricular *(RV)* and *LV* dimensions are normal. (**b**) M-mode echocardiogram of the anterior and posterior mitral leaflet. During diastole fluttering of the mitral valve (↑), as a sign of aortic regurgitation, can be seen. (**c**) M-mode echocardiogram of the aorta *(Ao)* and the left atrium *(LA)*. The aortic valve is thickened *(vv)*, but opens normally. The left atrium is not enlarged

gave normal results. An EEG from May 24, 1983, was of the α-type with some theta activity over the left temporal region. In a control EEG on August 11, 1983, the theta activity was less clearly expressed.

Under treatment with 200–600 mg carbamazepine/day the involuntary movements were reduced and finally disappeared almost totally. At re-examination on August 11, 1983, the patient was entirely normal.

Comments

The case presented here exhibits three particular facets: Firstly, the leading symptom of our patient were choreo-athetotic involuntary movements in association with an increased antibody titer to varicella-zoster virus; secondly, the choreo-athetotic movements were restricted to one side of the body; and thirdly, there are strong indications that our patient suffered simultaneously from an abacterial endocarditis. As reported by the mother, our patient had had chickenpox during early childhood. Therefore, one has to assume that not a primary infection but a reinfection from the father of the patient with varicella-zoster virus took place. That reinfections may occur in immune adults was recently reported by Arvin et al. (1983). They found that 64% of their immune adults exposed to varicella developed a fourfold or greater rise in IgG antibodies to varicella-zoster virus. The fact that our patient still showed an IgG titer of 1 : 128 about 6 months after the onset of symptoms which clearly declined during the following months (Fig. 1), supports the assumption of a reinfection.

Interestingly the clinical symptoms disappeared while the antibody titer returned to normal values (Fig. 1) which suggests an etiological role of the antibodies directed against varicella virus. One possibility is that the antibodies cross-reacted with neuronal cytoplasm of the caudate and subthalamic nuclei as was found for antibodies against group A streptococci by Husby et al. (Husby et al. 1976) and/or they were cross-reacting with heart valve tissue (Goldstein et al. 1968). Since our patient suffered from an unilateral chorea and in addition signs of an endocarditis (Fig. 2A and Fig. 3) were found by echocardiography, it might be that antibodies cross-reacted against the heart valve tissues. This cross-reaction could have resulted in the formation of local thrombi on their surface which may have led to small emboli in the area of the left subthalamic and caudate nuclei. The slight left temporal theta-focus in the initial EEG supports this view. The inconspicious cerebral CT scan and the missing thrombi on the surface of the heart valves (Fig. 2) do not argue against this assumption, since lesions smaller than 5 mm in diameter cannot be detected with the scanner used, and one has to assume that the most acute phase of the endocarditis had been weeks before hospitalization and ultrasound examination, as is stressed by the elevated IgG titer while no IgM antibody titer could be found.

The clinical symptoms which started in the right arm and spread to the right leg and the right side of the face point to a granulomatous angiitis after generalized herpes zoster infection as described by Kolodny et al. (1968) and Rosenblum and Hadfield (1972). Based on this consideration, the slow progress of the choreatic movements, and the discreet emphasized reflexes of the right limbs, we assume that the neurologic symptoms had been caused by an embolization resulting from the heart with at least one small infarction in the region of the left striatum (Goldblatt et al. 1974). This infarction presumably was followed by a local inflammation causing the spread of the involuntary movements and the pyramidal signs.

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Received November 1, 1984