Antiphospholipid Antibodies (APA) and Cerebral Stroke

V.P. Gharpure
V.G. Desai
C.T. Deshmukh
U.B. Nadkarni
M.K. Jain
M.D. Shah

Antiphospholipid antibodies (APA) are a group of complexly related autoantibodies directed against phospholipids found in cell membranes. Association of APA and thrombotic events had been demonstrated in 1946 by Aggeler et al. (1) who documented thrombotic episode with circulating anticoagulant and thrombocytopenia. Appan et al. (2) reported a 5-year-old child having anticardiolipin antibodies (ACA) with multiple thrombosis; Pereira et al. (3) reported a neonate with spontaneous aortic thrombosis due to passive transfer of maternal lupus anticoagulant; and

Roddy et al. (4) reported the first infant with APA and stroke. Since there is no report in the Indian literature, we are reporting a child with ischemic cerebral stroke and raised ACA.

Case Report

A 9-year-old female was referred in February, 1991 with complaints of right sided hemiplegia and right facial asymmetry for one month and inability to talk for 4 days. One month prior to her admission, she developed sudden weakness on the
right side which progressed to complete right hemiplegia. There was no history of convulsions or altered sensorium. She was diagnosed as cerebral vascular stroke and treated with aspirin, dipyridamole and steroids. Her lower limb power improved and she could walk with support in two weeks. However, ten days later, she developed sudden deterioration in right sided weakness and inability to speak. She had no other contributory history.

On examination, she had motor aphasia, right upper motor neurone paralysis and spastic hemiplegia on the right side. There was no evidence of increased intracranial tension. Examination of other systems did not reveal any abnormality.

On investigation, she had microcytic, hypochromic anemia with Hb 8.9 g/dl; ESR was 50 at the end of 1 hour; platelet count was 150×10⁹/L; and total and differential WBC count were normal. CSF and chest X-ray were normal. CFS-TB antigen and Mantoux test were negative. CT scan revealed an infarct in the left thalamus, basal ganglia and posterior limb of left internal capsule (Fig). Repeat CT scan after 4 weeks showed no deterioration. RBC sickling, VDRL, ANA and anti-dsDNA antibody tests were negative. Lipid profile was normal. 2D Echo and ECG were normal. Carotid angiography ruled out intracranial arterio-venous malformation. ELISA test for IgM ACA was positive with titre of 35 MPL units (>3MPL units was significant) but IgG ACA was negative. Activated partial thromboplastin time (aPTT) was 44 seconds, with control of 34 seconds. Abdominal ultrasonography and impedance plethysmography did not reveal thrombosis at any other site. Direct Coomb's test was negative. She was treated with low dose aspirin and physiotherapy. She could walk with support and speak a few works within 3 weeks. By 6 months, she was able to walk without support, spoke small sentences, and could raise her hand above the shoulder. IgM ACA titre fell down to 5 MPL units and IgG ACA titre, now positive, 15.5 GPL units (>5GPL units—was significant). Her lupus anticoagulant test was negative. She had no more thrombotic episodes during one year of follow-up.

Discussion

APA can be demonstrated by false positive VDRL test, and/or prolonged aPTT, and/or ACA by ELISA of which ACA is most sensitive and specific. APA can be positive in other autoimmune disorders like systemic lupus erythematosus, infections and phenothiazine group of drugs. The APA syndrome is diagnosed when one of the above serological tests are positive, in addition to evidence of thrombosis; thrombocytopenia or fetal loss during the course of the disease. Our case had high ACA titres with clinical evidence of cerebral infarct, with decrease in the titres after 6 months, as has been described in the literature. Kushner et al. (5) reported normal PTT in 65% of patients who had APA with ischemic cerebral stroke; Levin et al. (6) reported normal angiography findings in almost 50% patients with APA and ischemic stroke. The present case had normal aPTT and carotid angiography was unremarkable. Administration of anticoagulants, low dose salicylates and antiplatelet agents have been tried to prevent recurrent thrombosis; however, resistant cases may require use of steroids and immunosuppressants. Steroids and high dose IgG infusions have also been recommended.

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REFERENCES


Carbamazepine Therapy for Infantile Tremor Syndrome

M.V. Murali
P.P. Sharma
P.B. Koul
Piyush Gupta

Infantile tremor syndrome (ITS) is characterised by mental and psychomotor changes, pigmentary disturbance of hair and skin, pallor, tremors and subnormal intelligence(1). Since the etiology of this disorder is largely obscure, the therapy has been empirical and supportive. Iron, magnesium, calcium, vitamin B₆ and B₁₂, phenobarbitone and phenytoin have been used without consistent benefit(2,3). Kalra and Marwaha reported considerable improvement of symptoms in a trial of propranolol in eight patients(4).

Carbamazepine, a structural analogue of tricyclic antidepressants and phenothiazines, has been successfully used in treatment of various extrapyramidal movement disorders which include rheumatic chorea, chorea following head injury and torsion dystonia(5,6). We initially tried this drug on a child with ITS who was resistant to propranolol therapy. Dramatic disappearance of tremors in this child prompted us to use carbamazepine as a first line drug in four more children with ITS.

Case Reports

Case 1: An 11-month-old boy was admitted to Guru Tegh Bahadur Hospital, Delhi with the complaints of abnormal movements of limbs of four days duration following an episode of upper respiratory tract infection. The child showed global delay of developmental milestones. He was exclusively breast fed and was not weaned yet. On examination, weight was 7.5 kg and head circumference 45 cm. He was plump but pale with hair being depigmented, thin, silky and easily pluckable. Coarse tremors...