Antiphospholipid Antibody Syndrome With Associated Stroke And Increased Intracranial Pressure.

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Citation

J Patel, X Tang, R Sultan. *Antiphospholipid Antibody Syndrome With Associated Stroke And Increased Intracranial Pressure.*. The Internet Journal of Rheumatology. 2009 Volume 6 Number 2.

Abstract

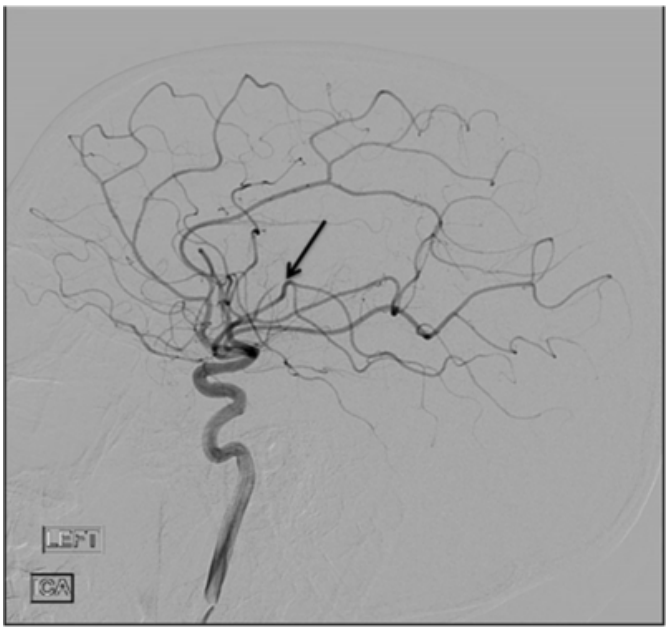
Rheumatologists often evaluate patients with Antiphospholipid antibodies; IIH has been reported in patients with antiphosholipid antibodies with or without thrombo- occlusive events. Exact pathogenesis of IIH in patients with antiphospholipid antibody syndrome is not known. We report a case of 15-year-old female presents with numbness of both hands for 5-6 months. Initial MRI revealed non-specific white matter changes but MRA of head and MRI of neck were not revealing. Laboratory data showed anticardiolipin antibodies and elevated sedimentation rate titer. Subsequently the patient developed unilateral right upper extremity numbness, slurred speech with right facial droop. Cerebral angiography revealed occlusion of the left middle cerebral artery. Lumbar puncture revealed intracranial opening pressure of 47 cm H2o, labs with normal cell count, glucose and protein. Patient's symptoms improved and she was discharged on aspirin. More research is needed to establish the relationship between IIH and anticardiolipin antibodies.

CASE REPORT

A 15 year old Caucasian female developed numbness in her left hand in November 2007 that quickly resolved; subsequently in May 2008 she developed a transient left sided hemiparesis. This was followed by progressive left hand weakness with a loss of fine motor function. She also had some difficulty in speech with episodes of dysphasia; numbness over left hand was persistent. A neurological exam was normal. MRI of the brain showed some non-specific white matter changes, non enhancing MRA was normal. Her lab showed Elevated sedimentation rate (32), low platelets (142), positive anticardiolipin antibodies (IgG-74, IgM-10), positive Lupus anticoagulant (63.8), Negative ANA, normal hypercoagulability profile and unremarkable basic labs.(Table-1)The patient was admitted in hospital for slurred speech, weakness of the right hand and right facial droop which lasted>24 hours. Cerebral angiogram was done. Left cerebral angiogram showed a focal area of occlusion in a branch of the middle cerebral artery(Figure-1), right cerebral angiogram was normal. Patient underwent a lumbar puncture, which revealed increased intracranial pressure of 47cm H2o. It raised the possibility of pseudotumor like syndrome. Her CSF studies were unremarkable. Patient was started on low dose aspirin; her symptom improved and she was discharged home on baby aspirin.

Figure 1

Figure-1. Cerebral angiogram showing occlusion of one of the branches of the left middle



Cerebral artery distally.

Figure 2

Table-1. Laboratory Test Results

Test	Result	Interpretation
Platelets	142	low
PTT-LA	82.8	High
PTT-LA Mix	75.8	High
Drvvt	59.7	High
Drvvt Mix	53.4	High
Drvvt confirm	1.7	High
Anticardiolipin Ab IgG	74	High
Anticardiolipin Ab, IgM	10	High
ANA	Negative	Normal
ESR	32	High
Ceruloplasmin	25.4	Normal
Lyme IgG/IgM Ab	<0.91	Normal
Factor V Leiden Mutation	Negative	Normal
Protein S-Total	117	Normal
Protein S -Free	84	Normal
Protein C Antigen	97	Normal
TSH	1.842	Normal
Thyroxine	8.1	Normal
CSF Glucose	55	Normal
CSF Character	CLEAN AND COLORLESS	Normal
CSF Opening pressure	47	High
CSF Total protein	18	Normal
CSF WBC	2	Normal
CSF RBC	88	Normal

Figure 3

Table-2. Differential diagnosis of Intracranial Hypertension

Idiopathic Intracranial Hypertension
Intracranial mass lesions (tumor, abscess)
Increased cerebrospinal fluid production (Choroid plexus papilloma)
Decreased cerebrospinal fluid absorption (Subarachnoid hemorrhage, bacterial or infectious meningitis)
Obstructive hydrocephalus
Obstruction of cerebral venous flow (jugular vein compression, venous sinus thrombosis

DISCUSSION

Antiphospholipid antibodies are those that prolong phospholipids dependent coagulation assays, can be categorized in to lupus anticoagulant and anticardiolipin antibodies. The antiphospholipid antibodies and stroke study group (APASS), a consortium of 15 medical centers previously reported that the presence of anticardiolipin antibody is an independent risk factor for first ischemic stroke (4). Arterial thromboembolism in the antiphospholipid antibody syndrome most commonly involves the cerebral circulation with stroke being the initial clinical manifestation in 13% and transient ischemic attack in 7% of the patients (6). Based on APASS data, patients with a first ischemic attack and a single positive antiphospholipid antibody test (IgM or IgG)result who do not have another indication for anticoagulation may be treated with aspirin or moderate intensity warfarin (INR, 1.4-2.8)(7). Aspirin is likely to be preferred because of its smaller side effect profile and lack of need for laboratory monitoring. The pathology of vascular events associated with anticardiolipin antibody is not clear. It was suggested that alterations of the natural balance between the coagulation and the fibrinolytic system favor thrombosis. Several possible targets for ACL-Ab action in thrombosis have been implicated, including endothelial cells (8), apoprotein H (9), naturally occurring anticoagulants and protein C and S(10).

Anticardiolipin antibodies have been associated with idiopathic intracranial hypertension (2) but the pathogenesis is not clear. Idiopathic intracranial hypertension may be due to increased blood viscosity induced by ACL-Ab but it's not studied. Idiopathic intracranial hypertension can develop in the presence of known risk factors such as thyroid disorders, Cushing's syndrome or systemic lupus erythematosus. Since our patient did not have these risk factors it is assumed that anticardiolipin antibody did play a role in the pathogenesis of idiopathic intracranial hypertension. The risk of the long term sequel associated with idiopathic intracranial hypertension is progressive visual loss; therefore close follow up of their visual acuity, and visual fields is required. Options of medical treatment are primarily diuretics like aceazolamide and encouragement of weight loss (12) surgical therapies such as CSF diversion via a lumbar shunt or fenestration of optic nerves may be necessary, in cases refractory to medical therapy. (12)Our case illustrates that antiphospholipid antibodies should be considered in the differential diagnosis of idiopathic intracranial hypertension (Table-2). More research is required to

Investigate the mechanism of the relationship between antiphospholipid antibodies and idiopathic intracranial hypertension before we could routinely recommend screening patients with idiopathic intracranial hypertension for antiphospholipid antibodies.

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