Cerebral infarction in a 17-year-old boy

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A 17-year-old boy presented with a history of sudden onset headache, vomiting, weakness of the right half of the body and inability to speak for 24 hours. There was no history of trauma, seizure, fever or any such episode in the past. There was a significant history of a syncopal attack 15 days prior to the episode during which the patient had suffered a minor laceration on the forehead.

Examination revealed an aphasic agitated boy with a pulse rate of 40 per minute and a blood pressure of 180/70 mm Hg in the right arm in supine position. All peripheral pulses were palpable. Cardiovascular examination revealed a grade III/VI pansystolic murmur at the apex. The first heart sound was variable in intensity and the second heart sound was wide and variably split. There was no third or fourth heart sound. Central nervous system examination revealed a conscious boy with pure motor aphasia. The pupils were normally reacting and cranial nerve function was normal except for right upper motor neuron type of facial nerve involvement. Fundus examination was normal. Motor power was 1/5 in the right upper and lower limbs. The tone was increased in the right half of the body and the right plantar response was extensor. Deep tendon reflexes were brisk in the right upper and lower limbs and an ill-sustained ankle clonus could be elicited in the right leg. Chest and abdominal examinations were non-contributory.

What is the most likely diagnosis in this patient based on clinical history and examination?

Sudden onset of dense hemiplegia involving the right half of the body along with motor aphasia suggests stroke involving the left middle cerebral artery territory. Hypertension and bradycardia could be a part of the Cushing’s reflex associated with raised intracranial tension. A grade III/VI pansystolic murmur at the apex suggests mitral regurgitation which makes a cardio-embolic origin of the stroke most probable.

What are the likely factors predisposing to cerebral infarction in this patient?

Cerebral infarctions in young people usually have an underlying cause and should prompt extensive workup for its identification. Infarcts in the middle cerebral territory are usually cardio-embolic in origin. Associated mitral regurgitation makes this even more likely. As the patient is young, a search for an underlying hypercoagulable state is also mandatory.

What minimum emergency investigations would you carry out in this patient?

a) Non-contrast CT scan of the head
b) 12 lead electrocardiography
c) Chest Radiograph
d) 2 D echocardiography and colour Doppler

Non-contrast CT scan of the head revealed a large wedge-shaped hypodense area in the left temporo-parietal region, which was causing compression of the left lateral ventricle and also midline shift. Electrocardiogram (ECG) showed a junctional bradycardia at a rate of 38 per minute. No P waves were discernible. Subsequent 24-hour Holter test showed complete heart block. Chest X-Ray revealed cardiomegaly of the left ventricular type. An echocardiographic examination using colour Doppler revealed an 8-mm patent foramen ovale, severe mitral regurgitation and a left ventricular ejection fraction of 57%. There was no obvious blood clot in any chamber. There was no vegetation on any valve.

What could be the most likely reason for the complete heart block?

That the patient had a syncopal attack 15 days prior to the infarction from which he had an uneventful recovery suggests that the heart block was not a consequence of the infarction, instead it could have predisposed him to the infarction. Cerebral infarction in complete heart block is rare and only two cases have been reported in the past.1,2 The complete heart block in this patient is most likely congenital. Congenital complete heart blocks may very well present for the first time in adolescence.

What is the pathophysiology and presentation of isolated congenital complete heart block?

Isolated congenital complete heart block is an example of passive autoimmunity, with the conduction defect caused by injury from active transplacental transport of maternal IgG antibodies into the foetal circulation. The incidence of isolated congenital heart block is 1 in 15000 to 20000 live births. The conduction defect is usually discovered...
incidentally in otherwise normal children. Stokes-Adams episodes are uncommon in young patients but pose tangible hazards. The diagnosis of congenital heart block is straightforward, based on nothing more than a standard scalar ECG. The QRS complexes are of normal duration because the block is above the bifurcation of the bundle of His. All patients with isolated congenital complete heart block should receive permanent pacemaker implantation.

What are the other investigations required for the complete workup of this patient?

Complete blood counts, serum electrolytes, renal function tests, liver function tests, urine examination, lipid profile, screening tests for coagulation, antinuclear antibodies, anti Ro and La antibodies, anticardiolipin antibodies, lupus anticoagulant, serum and urine homocysteine, protein C, protein S, anti-thrombin III (AT III), ultrasonography of the kidneys and, suprarenals, and Doppler of renal and carotid vasculature would form the battery of investigations in this patient.

The results of the investigations in our patient were as follows: Haemoglobin 14.5 g%, TLC 8600/mm³, polymorphs 66%, lymphocytes 30%, eosinophils 3%, basophils 1%, ESR 10 mm in the first hour by Wintrobe’s method, platelets 200,000/mm³, sodium 145 mEq/L, potassium 4.5 mEq/L, urea 25 mg%, creatinine 0.8 mg%, bilirubin 0.9 mg%, SGOT 32U/L, SGPT 26U/L, alkaline phosphatase 6KAU/L, total cholesterol 200 mg%, HDL 45 mg%, triglycerides 140 mg%, prothrombin time 11 seconds, partial thromboplastin time 29 seconds, antinuclear antibodies-negative by immunofluorescence, protein C 90% (normal = 58-148%), protein S 100% (normal = 58-148%), AT III 90% (normal = 80-120%). Ultrasonography of the kidneys and, suprarenals, and Doppler of renal and carotid vasculature were normal. Lupus anticoagulant was positive as suggested by a prolonged Russel viper venom time test on two occasions 8 weeks apart. Anticardiolipin antibodies were however negative. Anti Ro and La antibodies could not be done due to financial constraints.

What is the significance of antiphospholipid antibodies in ischaemic strokes?

Though antiphospholipid antibodies may be found in 10% of all patients with acute stroke, they are found in as many as 50% of young patients with stroke. Cerebral infarction in association with isolated positive lupus anticoagulant has also been described and it is proposed that lupus anticoagulants may be stronger risk factors for thrombosis than anticardiolipin antibodies.

What are the cardiac manifestations of antiphospholipid antibody syndrome?

The cardiac manifestations of antiphospholipid antibody syndrome include valvular disease (mitral regurgitation being the commonest valvular lesion), myocardial dysfunction, coronary artery disease, and intracardiac thrombosis. However, heart block has not been described. Mitral valve involvement is common in patients with antiphospholipid syndrome and the severe mitral regurgitation in our patient can be explained on this basis. Patients with severe valvular heart disease and antiphospholipid antibodies have an increased risk for developing thromboembolic events, as was the case in our patient.

What is the significance of a patent foramen ovale (PFO) in patients with ischaemic strokes?

PFO has been increasingly recognized as an important source of paradoxical embolism in patients less than 55 years of age with cryptogenic strokes. Also, in the presence of a hypercoagulable state, as was the case in our patient, authorities now recommend that closure of this defect be done in order to prevent recurrences.

What would be the ideal management in this patient?

The ideal management in this patient would include conservative medical management of the raised intracranial tension, physiotherapy, lifelong anticoagulation, antiplatelet therapy, permanent pacemaker implantation, closure of the PFO and possibly mitral valve repair.

A temporary transvenous pacemaker was initially inserted for the complete heart block in this patient and seven days later a permanent pacemaker was implanted. Acetylsalicylic acid 150 mg/day and warfarin 5 mg/day comprised the medical management of the patient. The patient was discharged after 15 days with minimal residual aphasia. At six months of follow-up, the patient had only mild residual neurological deficit. Surgical closure of PFO is planned but presently the patient has inadequate finances to undergo the surgery.

References