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AN INTERESTING CASE OF STROKE IN THE YOUNG

SHRIDHARAN
Department of General Medicine,
MADURAI MEDICAL COLLEGE AND HOSPITAL

Abstract : Stroke occurring in the young population is due to causes different and diverse from that of the elderly. Apart from cardioembolic origin which is the most common etiology of stroke in the young, collagen vascular diseases, vasculitis, hematological disorders etc cause stroke in the young. In this report we present an interesting case of stroke in a 22 yr old male who presented with acute onset hemiplegia and status epilepticus, imaging revealed cerebral venous sinus thrombosis, patient dramatically improved with intravenous anticoagulants along with other supportive measures like antiepileptics, anticerebral edema measures. On further nvestigation, he was found to have Protein C deficiency and advised lifelong oral anticoagulation, and antiepileptic.

Keyword :stroke, status epilepticus, anticoagulation

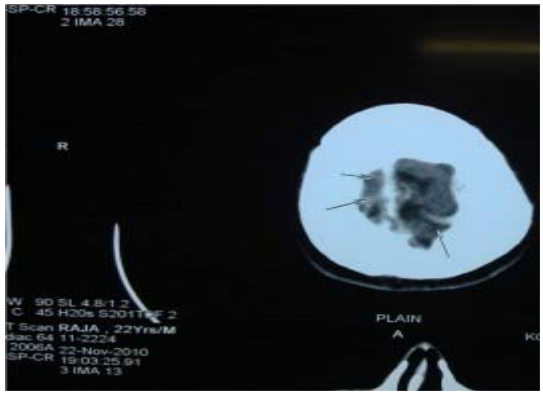
Introduction

Stroke in the young (<45 yrs of age) occurring due to Inherited clotting factor deficiencies (Protein C,S deficiency, activated protein C resistance etc.) is a rare entity. Protein C deficiency usually presents as cerebral venous sinus thrombosis, which may manifest itself as headache, papilledema, seizures, focal neurological deficit etc. Early identification of the condition and prompt institution of anticoagulation ensures good therapeutic outcome.

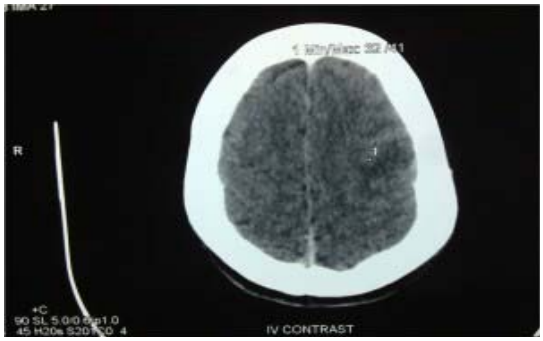
Case report

Mr. R, 22 yr old male, an agricultural worker by occupation was brought to the hospital by his father with h/o insidious onset of weakness of rt upper and lower limb followed in a few hrs by convulsions involving right upper and lower limbs - tonic, clonic type; multiple episodes; each episode lasting for 10-15 min with no regaining of consciousness in between. H/o loss of consciousness following seizures present. No h/o fever, headache, vomiting, diarrhea, dyspnea, palpitation, chest pain, trauma. Patient is not a known diabetic or hypertensive, no h/o TB. Patient is not a smoker or alcoholic. No h/o substance abuse. No significant family history.

On examination – patient was stuporous, afebrile, no pallor, no clubbing, no cyanosis, no lymphadenopathy, no pedal edema. Vitals : pulse 90/min, regular rhythm, felt in all peripheral accessible arteries, no radiofemoral delay. BP 110/70 mm Hg. Temperature 99 F CNS – stuporous, responding to deep pain by movement of left upper and lower limbs. Pupils 3mmequal, reacting to light. Fundus – bilateral papilledema. Right UMN facial weakness present. Muscle bulk equal bilaterally. Hypotonia right upper and lower limbs. Normal tone left upper and lower limbs. Power 0/5 right upper & lower limbs; >3/5 left upper & lower limbs. DTR depressed right side; normally present left side. Plantar reflex – right: extensor, left – flexor. Carotids – equal, no bruit. Other system examination (CVS, RS, Abdomen) – within normal limits. With a *provisional diagnosis* of RIGHT PARTIAL SEIZURES WITH SECONDARY GENERALISATION, STATUS EPILEPTICUS, RIGHT HEMIPLEGIA, patient was *treated* with Inj diazepam 10 mg iv stat followed by Inj Phenytoin 20 mg/kg iv stat, followed by 100 mg iv TDS and we proceeded with *INVESTIGATIONS* his blood sugar 126 mg%, urea 46 mg%, creatinine 1.2 mg% ; serum sodium 136 mEq/l, potassium 4.6 mEq/l; ECG – within normal limits; ECHO – normal study; HIV ELISA negative. Electroencephalogram done on the 2nd day was normal. Complete Hemogram & Peripheral Smear – within normal limits (Hb 13.5 gm% ESR 40 mm/hr TC 7500/cu.mm; all three cell lines normal in number and morphology)CT BRAIN – acute thrombus involving entire superior sagit tal sinus, frontoparietal cortical vein of left side. Infarct involving left perisylvian and frontal subcortical regions



CT Brain showing acute thrombus involving entire superior sagittal sinus, frontoparietal cortical vein of left side



CT Brain showing Infarct involving left perisylvian and frontal subcortical regions

Patient was started on Inj Heparin 5000 units iv tds and Inj Mannitol 100 ml iv tds. Other supportive measures were continued. Patient regained consciousness and his power started to improve.
We proceeded with investigations to ascertain the cause of CVT Antinuclear Ab(ANA) negative, Anti ds DNA negative, Antiphospholipid Ab IgM and IgG – negative. Protein S – normal; Protein C – 20 IU/dl (normal 65 – 135 IU/dl)
Heparin continued for 14 days and switched over to Oral anticoagulant (Acenocumarol 2mg od) –advised to give it lifelong, with periodic monitoring of Prothrombin time/INR. Oral antiepileptic Tab Phenytoin 200 mg HS was advised to be continued. Plan to do EEG at 6 mo intervals to determine the need to continue antiepileptic.

Discussion:
Stroke in the young refers to stroke occurring in persons below 45 yrs of age. Common causes of stroke in the young include –(i) Cardioembolic – rheumatic heart disease, infective endocarditis, mitral valve prolapse etc (ii) arteritis – TB, syphilis, aorto arteritis, takayasu arteritis (iii) collagen vascular disease – SLE, antiphospholipid antibody syndrome (iv) Hematological – sickle cell disease (v) AV malformation (vi) Thrombophilia – protein C,S deficiency, antithrombin III deficiency, hyperfibirnogenemia, cancer, pregnancy, OC pills, nephritic syndrome, myeloproliferative disorders, etc.(1)

Thrombosis of the cerebral venous sinuses, particularly of the superior sagittal or lateral sinuses and the tributary cortical and deep veins gives rise to a number of important neurologic syndromes.(5) Cerebral venous thrombosis may develop in relation to infections of the adjacent ear, paranasal sinuses or bacterial meningitis. Non infectious causes are more common resulting from one of the many
hypercoagulable states(thrombophilia) – dehydration, pregnancy, post partum, oral contraceptive pills, cancer, cyanotic congenital heart disease, factor V Leiden mutation, protein C or S deficiency, primary or secondary polycythemia, thrombocythemia, paroxysmal nocturnal hemoglobinuria, inflammatory bowel disease, hyperhomocystinemia, drugs – tamoxifen, erythropoietin - of which protein S deficiency and hyperhomocystinemia have the propensity to cause both arterial and venous thrombosis.(1) ***Protein C***, also known as autoprothrombin IIa, is a vitamin K-dependent glycoprotein structurally similar to other vitamin K-dependent proteins affecting blood clotting, Protein C is a major component in anticoagulation in the human body.(4) It acts as a serine protease zymogen : APC(activated protein C) proteolyses peptide bonds in activated Factor V and Factor VIII (Factor Va and Factor VIIIa) – which are highly procoagulant cofactors in the generation of thrombin , a crucial element in blood coagulation; A genetic protein C deficiency , in its mild form associated with simple eterozygosity , causes a significantly increased risk of venous thrombosis in adults. If a fetus is homozygous or compound heterozygous for the deficiency, there may be a presentation of purpura fulminans, severe disseminated intravascular coagulation and simultaneous venous thromboembolism in the womb; this is very severe and usually fatal. Protein C deficiency is found in 0.2 -0.4% of the general population and 3-6% of those with cerebral venous thrombosis.
Patients present with headache, focal neurological signs, seizures. The slower evolution of the clinical stroke syndrome, the presence of multiple cerebral lesions not in arterial territories, and an epileptic and hemorrhagic tendency favors venous over arterial thrombosis.(3) With greater degrees of sinus thrombosis, the patient may develop signs of increased ICP and coma. Venous sinus occlusion is readily visualized during MR or CT venography.
Treatment : intravenous heparin regardless of the presence of intracranial hemorrhage has been shown to reduce morbidity and mortality, and the long term outcome is generally good. Heparin prevents further thrombosis, reduces venous hypertension and ischemia.(1) If an underlying hypercoagulable state is not found, oral anticoagulants (vitamin k antagonists) are given for 3- 6 months. Indefinite anticoagulation is continued if thrombophilia is diagnosed. Antibiotics are advised if the venous occlusion is infectious. Anticonvulsants are given if there is seizures at onset. They are continued for 18 months and an EEG taken – if it continuous to show focal sharp waves, antiepileptic is continued, if not the medication may be tapered and discontinued.(2)

Conclusion
Any case of stroke in the young should be thoroughly evaluated to find out the cause of stroke so that specific therapy can be instituted. Cerebral venous thrombosis should be investigated to find out the exact cause so as to determine the duration of anticoagulation, indefinite antico-agulation needed in underlying inherited hypercoagulable disorders to prevent recurrent stroke and other complications.

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