University Journal of Medicine and Medical Specialities

ISSN 2455- 2852 2018, Vol. 4(2)

A RARE CAUSE OF CORTICAL VEIN THROMBOSIS

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Abstract : Cortical vein thrombosis is an uncommon but severe thrombotic manifestation with high potential to cause disability and tendency to recur. Cortical vein thrombosis in males is very rare when compared to females. A detailed evaluation has to be done to find out the cause for the cortical vein thrombosis particularly if they are young, because treatment for the specific cause may help to prevent recurrent cortical vein thrombosis. Protein C deficiency is one of the rarer cause of cortical vein thrombosis.

Keyword : Cortical vein thrombosis , Proetein C deficiency

INTRODUCTION: Cerebral venous thrombosis (CVT) has a wide spectrum of clinical manifestations that may mimic many other neurological disorders and lead to misdiagnosis. 85% of the cortical vein thrombosis are due to acquired causes.Remaining 15% of the cortical vein thrombosis are due to inherited thrombophiliac states,out of which Protein C deficiency is one of the important cause. Protein C deficiency is a rare genetic trait characterised by inherent tendency for thrombosis.

CASE REPORT: A 34 years old male patient Mr.Chandrasekharan admitted with history of 3 episodes of generalised tonic clonic seizures.history of head ache was present.no history suggestive of any cranial nerve involvement or weakness was present.

On examination patient was conscious,oriented. Blood pressure was 130/80mm of mercury.Higher mental functions were within normal limits.No cranial nerve involvement and no motor weakness.fundus was normal.No pyramidal signs were present.Cardiac and other systems were within normal limits.

All routine investigations such as blood count ,renal function tests,liver function tests,electrolytes,blood sugar were normal.CT Brain showed a hypodense region in the right frontal region,which was suggestive of cortical vein thrombosis.MRI Brain with MRV shomed superior sagittal sinus thrombosis.

On evaluation for the cause for the cortical vein thrombosis,we found out that protein C activity for this patient was very low which was 37%(70-130%).

FINAL DIAGNOSIS: CORTICAL VEIN THROMBOSIS DUE TO PROTEIN C DEFICIENCY. We treated him with Heparin and phenytoin.Now he is on Warfarin 2mg/day and he is under regular follow up.

DISCUSSION: Cerebral venous thrombosis is an uncommon condition, but its clinical presentation is varied and often dramatic. It often affects youngtomiddleaged patients, and more commonly women. Although recognized for more than 100 years, it has only in recent years come to be diagnosed frequently antemortem. With the advent of three dimensional M.R. Flow Imaging it has been shown that the prevalence is more common than reported previously and carries a less serious prognosis. **ETIOLOGY**

Hypercoagulable Conditions. Pregnancy & Puerperium Oral Contraceptives15 Anti-thrombin III deficiency Antiphospholipid Syndrome Protein C & S alteration	**Changes in blood viscosity**. Marasmus Malnutrition Dehydration Congestive heart failure Hyperviscosity syndrome
Changes in vessel wall. Malignancy Infections : local-chronic-otitis media, nasolabial infection Systemic, e.g., gram negative septicaemia, fungal infection.	

CLINICAL PRESENTATION

Clinical presentation can be extremely varied, and symptoms can evolve over hours to a few weeks. In a recent study the most frequent symptoms and signs were headache (95%), focal seizures with or without secondary generalization (47%), paresis (uni or bilateral(43%) and papilledema(41%).Fifteen percent were comatose and a further 39% had some impairment of consciousness at presentation.Twenty percent presented with a isolated intracranial hypertension (BIH) type picture (headache, visual disturbance and papilloedema). Other rarer presentations include thunderclap headache mimicking subarachnoid haemorrhage.

INVESTIGATIONS: Objectives of investigations are a) diagnosis of cerebral vein/sinus thrombosis; b) identification of venous sinus involved; CT BRAIN FEATURES: Haemorrhagic infarcts ;Cerebral edema; Delta sign ;Cord sign; Dense triangle sign MRI BRAIN: Routine use of MRI is likely to throw more light on its prevalence, aetiologic risk factors, course of disease and

efficacy of treatment instituted. MR angiography not only seems to offer an important advantage as a non-invasive tool in diagnostic procedures but also seems useful as a follow-up instrument for documentation of thrombus regression, recanalisation and venous collateralisation.

PROTEIN C DEFECIENCY

General consideration regarding protein C deficiency are as follows. Protein C is a Vitamin dependent plasma protein and one of the most important inhibitor of blood coagulation. Protein C is activated in vivo by thrombin coupled with endothelin cofactor thrombomodulin to form activated protein C. Its a potent anticoagulant enzyme and it has got three mechanism of action: 1)it inhibits thrombin formation by inactivating the coagulation factors Va and V111a 2)it inhibits binding of factor Xa to platelets 3)it stimulates the release of plasminogen activator thereby promoting fibrinolysis. The mode of inheritance of protein C deficiency is thoyght to be autosomal dominant. Diagnosis of protein C deficiency is made by documenting decreased protein C activity.

TREATMENT: Current evidence shows that patients with cortical vein thrombosis without contraindications for anticoagulation should be treated either with body weight-adjusted subcutaneous low molecular weight heparin nor with dose-adjusted intravenous heparin with an at least doubled activated partial thromboplastin time. Concomitant intra cerebral haemorrhage related to cortical vein thrombosis is not a contraindication for heparin therapy. There is insucient evidence to support the use of either systemic or local thrombolysis in patients with cortical vein thrombosis. If patients deteriorate despite adequate anticoagulation and other causes of deterioration have been ruled out, thrombolysis may be a therapeutic option in selected cases, possibly in those without intracerebral hemorrhage or impending herniation from large haemorrhagic infarcts. Oral anticoagulation may be given for 3 months if cortical vein thrombosis was secondary to a transient risk factor, for 6–12 months in patients with idiopathic cortical vein thrombosis and in those with mild thrombophilia.Indenite oral anticoagulation should be considered in patients with two or more pisodes of cortical vein thrombosis and in those with one episode of cortical vein thrombosis and severe thrombophilia.

CONCLUSION: Whenever a patient comes with features suggestive of cortical vein thrombosis immediate clinical and radiological skills should be applied to confirm the diagnosis so that early anticoagulation can be instituted so as to prevent morbidity and mortality. A detailed evaluation to find out the etiology is always warranted. If any inherent thrombophilia as a cause is made out as in our case life long oral anticoagulation therapy should be given with monitoring.

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