Moya Moya Disease: An Unusual Clinical Presentation

A Somarajan, R Ashalatha, K Syam

Abstract

Moya Moya disease is a rare cause of stroke in adults, and is a rarity secondary to hepatitis C virus infection (HCV) and cryoglobulinemia (CG). We report such a rare association in a young patient who presented with intracerebral hemorrhage.

INTRODUCTION

A variety of central nervous system disorders can occur with HCV infection. Stroke may be the initial manifestation of HCV infection, although hemorrhagic stroke is rather uncommon. HCV associated stroke may be related to the presence of cryoglobulins, which leads on to a vasculitis or a coagulopathy. A Moya Moya like presentation can be secondary to such a vasculitis occurring in intracerebral vessels, which can present as intracerebral bleed in adults. We report this case considering the rarity of such an association and lack of earlier descriptions of the same from India to the best of our knowledge.

CASE REPORT

A 25 years gentleman who was detected to have borderline essential hypertension in 2001 and was on treatment thereafter developed an acute onset left hemiparesis in October 2002 which recovered completely in 5-10 minutes. He did not have any other vascular risk factors nor any positive family history of an acute cerebral insult. He was evaluated for the same as a case of ‘Stroke in the Young’. Laboratory examination revealed significant albuminuria and hematuria with mildly elevated serum creatinine, renal biopsy showed evidence of membranous glomerulonephritis. Other investigations including hemogram, ESR, liver function tests, lipid-profile, C-reactive protein (CRP), ANA (anti nuclear antibody), APLA (antiphospholipid antibody), anti dsDNA (antibody to double stranded DNA), SS-A and B (systemic sclerosis A and B antibody), cANCA, pANCA (antineutrophilcytoplasmic antibody), complement C3 and 4 titres, VDRL, TPHA, HBsAg and HIV-I and II ELISA were negative. CT head, MRI and MR angiography of brain, renal and mesenteric angiography, nerve conduction study, muscle and nerve biopsy were also non-contributory. Although the etiology for the transient cerebral ischemic event remained obscure at that time, he thereafter received full dose of prednisolone for his renal disease, which he continued till May 2003.

In June 2003, he presented with acute severe holocranial headache with vomiting and decline in sensorium. Clinical examination revealed a blood pressure of 140/90mm Hg, with signs of meningeal irritation and features of raised intracranial pressure in the form of bilateral papilledema and lateral rectus palsy. There were no other lateralising signs. A CT head taken showed an intraventricular hemorrhage with...
Subarachnoid extension without any parenchymal component (Fig. 1). He was treated with anti-edema measures and other symptomatic measures and patient improved completely.

A subsequent evaluation with a cerebral 4-vessel DSA showed stenosis of terminal internal carotid artery and proximal middle cerebral artery and anterior cerebral artery bilaterally with prominent basal ganglia collaterals qualifying for a Moya Moya disease (Figs. 2-4). A secondary cause for the same was searched for, his serum cryoglobulins were elevated and HCV RNA and anti HCV antibody were found to be positive. Other workup including ANA, rheumatoid factor, HBsAg, liver function tests and coagulation parameters including serum homocysteine and sickling test were non-contributory. A CSF study was deferred in this patient since he had massive intracerebral bleed. He was initiated on antiplatelet agents in addition to full dose of steroids. The option of treating the prevailing HCV infection with alpha-interferon and ribavarin was suggested which could not be implemented due to financial constraints. At 6 month follow up the patient is remaining asymptomatic.

**DISCUSSION**

Cryoglobulinemia (CG) is a condition characterized by the presence of serum proteins that reversibly precipitate in the cold. CG may be idiopathic (essential mixed CG) or secondary to other diseases such as lymphoproliferative disorders, collagen diseases and chronic infections. It has been reported that 46-54% of patients with chronic HCV infection show detectable cryoglobulins, although most of them remain asymptomatic. A high prevalence of membranous glomerulonephritis has also been noted in patients with chronic HCV infection as has been noted in our patient as well. Cryoglobulinemia associated CNS vasculitis usually leads to ischemic strokes. But this patient with documented cryoglobulinemia and HCV infection presented with a Moya Moya picture with secondary intraventricular bleed.

We report this case to draw attention to the varied manifestations of a systemic disease like cryoglobulinemia associated with HCV infection, which
can be brought to light only with a high index of suspicion. Also in all cases of Stroke in the Young, a detailed workup to pinpoint the etiology is rewarding. To the best of our knowledge such an association has not been reported in our part of the country.

REFERENCES

---

**Announcement**

**ITP Study Group**

At the recently held First National Conference on Idiopathic Thrombocytopenic Purpura, it was decided to form ITP Study Group with a view to study the natural history of the disease in India and also to see the commonly prevailing practice in treating this disease. Based on the information collected in such study, recommendations can be made about the management of ITP in India including a possible role of alternative forms of therapy.

Those who are interested in joining the study group should contact: Dr B C Mehta at (labmed@ghrc-bk.org). It is necessary that those who wish to join the group have easy access to internet. All communications of the study group will be through e-mail and web. Members will have access to the data/information on web.

---

**Announcement**

**Medical and Surgical Endocrinology Update, Hinduja Hospital**

For details contact

Dr. Deepraj Bhandarkar  
Dr. Manoj Chaddha  
mobile : 98210 14939  
mchadha@vsnl.com

---

**DOCTOR 2004 SOFTWARE**

A highly advanced, easy to use, economical and revised medical software package made just for you.

**CLINICAL**: Case sheets and speciality sheets; prescription autodose, autoallergy, contraindication, interaction alert, fonts option (Hindi/Tamil etc.). Allows auto-filling with very little typing needed. Detailed lab, PDR, auto-case summary, certificates, letters; detailed diet adviser.

**ADMINISTRATIVE**: Appointment scheduler; finance billing; salary, room, manpower management; drug store, detailed patient statistics and inventory. Secure, and network ready.

**OTHERS**: Web compatible - send case summary, reports by e-mail etc.

**EDUCATIVE**: Disease guidelines and journal reference; medical photographs and graphs; patient education videos and printouts. Widely used, reliable, saves life, time and money.

No learning required. Hospital pack, and excl. medicine, surgery, OBG, clinic packs available.

Address: MEDISOFT, Achutha Warrier Lane, Cochin, Kerala-682035. Ph.: 09847294414  
E-M: medisoftindia@hotmail.com OR medisoft@doctor.com  
web: www.medisoftindia.com  
Rs. 8000/- only