ISCHAEMIC STROKE IN YOUNG WOMAN DUE TO PROTEIN-C DEFICIENCY

Selvaraj Pitchai1, Ramu2, Mithun Mathiyazhakan3, Sravankumar Sampati4

1Professor, Department of General Medicine, Sri Venkateswara Medical College Hospital and Research Centre, Puducherry.
2Professor, Department of Neurology, Chennai Medical Hospital and Research Centre, Trichy.
3Postgraduate Resident, Department of General Medicine, Sri Venkateswara Medical College Hospital and Research Centre, Puducherry.
4Postgraduate Resident, Department of General Medicine, Sri Venkateswara Medical College Hospital and Research Centre, Puducherry.


PRESENTATION OF CASE
A 30-year-old previously healthy female patient was admitted in this teaching hospital with history of sudden onset of sensory loss and weakness of left upper and lower limbs. She had headache for 10 days before the onset of weakness. There had been no loss of consciousness, seizures, chest pain or giddiness. She did not have any bleeding diathesis in the past. No history of Diabetes Mellitus, Hypertension or TIA. She had no history of neurological dysfunction in the past. No family history of thromboembolic stroke or CAD. Parents married out of second degree consanguineous marriage and both are alive, and she had five brothers of which one was born dead. Her 10-year-old son is healthy, but his protein-C level was also low 55.3 (70 - 140). Other close relatives were not available for investigations.

Examination
Examination of the patient showed weakness of left upper and lower limbs with 3 - 4/5 power and brisk reflexes and plantar extensor on left side. Cerebellar functions, sphincters and other neurological examinations were largely normal. Carotid on the right side was feeble but no bruit. There was diminished sensation on the left half of the body. Investigations like ECG, chest x-ray, echocardiogram, immunological studies like Anti-DS DNA antibody, ANA were normal. CBC, LFT, serum electrolytes, sugar, urea, creatinine, urine analysis and platelet counts were normal. Lipid profile showed slight elevation of triglycerides. Blood VDRL and HIV were negative. Serum fibrinogen, prothrombin time, PTT, BT and CT was normal. Serum protein-S was normal 76.9 (60 - 140).

Protein-C was very much reduced (22.9), normal being (70 - 140). The absolute concentration of protein-C and antiphospholipid antibodies and antithrombin-III were not measured. CT scan brain was normal. MRI brain showed small acute pontine infarct on ventral pons (posterior circulation stroke) on the right side. MR-Angio showed hypoplastic right vertebral artery and right anterior communicating artery. MRV was normal. Carotid and vertebral arteries Doppler showed mild atherosclerotic changes with soft plaques in right proximal internal carotid artery, but no thrombosis.

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Corresponding Author: Dr. Selvaraj Pitchai, Professor, Department of General Medicine, Sri Venkateswara Medical College Hospital and Research Centre, Puducherry.
E-mail: Selvaraj@gmail.com
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The patient was treated with parenteral anticoagulants initially followed by oral anticoagulants and statins. Patient quickly improved with good power and she was discharged with statins and oral anticoagulants.

Protein-C deficiency mainly causes venous thrombosis. The incidence is 1 in 20000. Asymptomatic protein-C deficiency may be 1 in 200 - 300. So far only few cases of arterial thrombosis have been reported and majority of them are acute MI followed by ischaemic stroke and peripheral arterial thrombosis. Protein-C deficiency increases the risk of stroke or MI when it is associated with other risk factors like smoking, alcohol, dyslipidaemia and hypercoagulable states than by itself. It should be considered as the cause, particularly when young people develop stroke or the cause is unclear. We have seen two cases so far and both of them are around 30 years of age in this institution- one case was admitted with pure dysphasia and another case presented with aphasia with right upper limb weakness.

Even though it occurs as autosomal dominant or rarely as recessive, sporadic cases of protein-C deficiency with arterial thrombosis have also been reported (Dusser et al). In some case protein-C deficiency has produced temporary blindness (Amaurosis fugax) (Smith and Ens al). Kemkes-Matthes et al reported a case of carotid artery occlusion due to protein-C deficiency. Seshia and Israels et al have described a case of acute hemiparesis in a 17-year girl due to protein-C deficiency, but CT-brain was normal in that patient. Low levels of protein-C were first associated with venous thrombosis, (Griffin et al in 1982).

In our patient, the sudden onset of left-sided hemiparesis and hemianesthesia was probably due to a lacunar infarct in the ventral pons (posterior circulation stroke), as evidenced by MRI. In many cases, the lacunar infarct in brain resulting from microembolism from carotid or MCA are not picked up by either CT or MRI scan in the initial stages. In about 30% - 40% of embolic strokes, the source of embolism may not be determined (cryptogenic stroke) even though the patient may be presenting with neurological deficit. In addition, MRA in this patient showed hypoplastic right vertebral artery and hypoplastic left anterior communicating artery in this patient, but MRV was normal.

Posterior circulation stroke usually causes crossed hemiplegia and hemianesthesia. This was not seen in our patient but few cases have reported it. Amaurosis fugax was not seen in our patient but had been reported. Lacunar syndrome can result in pure motor or sensory stroke or both. Lacunar infarcts are caused by occlusion of small penetrating branches in internal capsule, basal ganglia, pons, thalamus, corona radiate or cerebral pedunde. Lacunar syndrome can result in pure motor hemiparesis or pure sensory stroke due to lacunar infarct was first described in 1965 by Fisher and Curry. Lacunar syndrome can be total or partial. The partial motor deficit includes isolated brachial monoplegia, unilateral vocal cord palsy and brachiofacial palsy. Lacunar infarcts generally carry good prognosis, but multiple lacunar infarcts can cause dementia. CT brain or MRI are best diagnostic tools to diagnose lacunar infarcts. Sometimes lacunar infarcts may be missed by both CT and MRI.
and MRI, hence CT or MRI should be repeated few days later since enhancement of lesions occurs after few days.

Lacunar infarction is more common in carotid territory than vertebrobasilar territory in the ratio of 70:30. Atypical lacunar syndromes such as isolated 3rd nerve palsy, foville’s syndrome, isolated eye and other movement disorders like chorea, hemiballismus, dystonia, parkinsonism, aphasia, visual or memory impairment have also been reported. We have also seen one isolated case of dysarthria clumsy hand syndrome and 1 case of isolated dysphasia in the last 3 months in our hospital.

**Conclusion**

Here, we are presenting a rare case of left-sided pure motor hemiparesis and hemianaesthesia of posterior circulation stroke in ventral pons due to protein-C deficiency associated with dyslipidaemia. Besides, the patient’s 10-year-old son also had low level of protein-C (55.3), thereby strongly emphasising a genetic trait in this case. Hence, we conclude that protein-C deficiency associated with dyslipidaemia probably might have caused posterior circulation stroke in this patient, involving one of the paramedian branches of basilar artery in ventral pons, even though other unknown factors may have played a role. In conclusion, therefore, it is important to recognise protein-C deficiency as a significant risk factor in young patients presenting with ischaemic stroke or myocardial infarction of unknown cause. However, the study of protein-C deficiency causing CAD or CVA must involve a large population in multicentres. This case is presented for its rarity namely- 1. A young woman presenting with posterior circulation stroke due to protein-C deficiency; 2. Her son also had asymptomatic low protein-C level; 3. Posterior circulation stroke due to arterial thrombosis; 4. Hypoperfusion due to hypoplastic right vertebral artery and hypoplastic right anterior communicating artery (haemodynamic theory).

**FINAL DIAGNOSIS**

Cerebrovascular accident- acute non-haemorrhagic pontine infarct (posterior circulation stroke) in ventral pons right side due to protein-C deficiency.

**REFERENCES**


