Recurrent Arterial Thrombosis In A Young Male: Sticky Platelet Syndrome

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Citation

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Abstract

We report a case of a 17 yr old male who presented with multitude of thrombotic events of the coronary, cerebral and peripheral arterial circulation. When the evaluation for the commoner causes of prothrombotic states were negative, platelet aggregrometric studies revealed hyperaggregable platelets. Patient was diagnosed to have sticky platelet syndrome and started on Aspirin to which he responded remarkably and remained asymptomatic for three years. Given the simplicity in diagnosis, treatment and follow up, internists should consider Sticky Platelet Syndrome (SPS) in their differential for recurrent arterial thrombosis.

CASE REPORT

A 17yr old male was admitted for claudication pain in his right calf of 2 days duration in August 2003. Past history was significant for a history of chest discomfort one week back for which he had taken antacids without relief. The discomfort spontaneously disappeared after 4 hours. He did not seek medical help during that event. He was nonsmoker, non alcoholic, not an IV drug abuser and denied high risk sexual behaviour. He was born to a non-consanguinous parentage and there was no history of coronary heart disease, peripheral vascular disease, dyslipidemia or prothrombotic states in the family. General examination was unremarkable with no evidence of xanthelasma, tendon xanthomas, earlobe crease, dislocated lenses or mental retardation. Palpation of arterial pulses showed decreased voulme of pulse in the right popliteal, posterior tibial and dorsalis pedis on comparison with the left side. Capillary return was delayed in the right toes. Rest of the system examination was normal. Shortly after his admission he developed severe pain in his right foot and toes and started developing signs of impending gangrene of the right toes. The patient was immediately anticoagualated with heparin and was taken up for emergency thromoembolectomy. A thrombus was found in the right femoro-popliteal junction. His routine blood counts, biochemistry, liver functions, renal functions, lipid profile were normal. ECG showed evidence of recent anterior wall myocardial infarction. Chest X-ray was normal. Echocardiogram showed akinesia of apex and anterior wall of left ventricle and a left ventricular clot measuring 2 X 1 cm. A coronary angiogram revealed normal anatomy with no malformations. Patient was treated with Aspirin, Beta-Blockers and ACE-Inhibitors. In addition he was also started on a warfarin (titrated to an INR between 2.5 – 3) in view of a suspected prothrombotic state. A detailed investigation (blood drawn prior to initiating warfarin) for prothrombotic states was done as shown in Table. 1.

Figure 1

Table 1: Investigations for prothrombotic states

Test	Subject's value	Normal
1. Antiphospholipid antibodies	Negative	
2. Anti-cardiolipin antibodies IgG	1 GPL	0 – 15
3. Anti-cardiolipin antibodies IgM	3 MPL	0 – 15
4. Serum Homocystiene	6 micromoles/L	0 – 12
5. ANA	Negative	
6. ANCA	Negative	
7. Bleeding time	2 min.	2 – 9.5
8. Clot retraction	Normal	
9. Sucrose lysis test	Negative	<10% hemolysis
10. Euglobulin lysis time	110 minutes	90 – 360
11. Protein C	0.9	0.7 – 1.4
12. Protein S	0.7	0.7 – 1.4
13. Anti-thrombin III	26 mg/dl	22 – 39

*Testing for Factor V leiden could not be done due to financial constraints.

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DISCUSSION

Patient was discharged with advise to continue warfarin, enalapril and periodically monitor INR. However he was readmitted a month later (Sep 2003) with features of right sided hemiplegia and Broca's aphasia. CT brain revealed the presence of infarct in the left frontal cortex. Patient's INR was 2.7 indicating compliance with Warfarin. The patient's blood was tested for platelet aggregation in serial dilutions of ADP and epinephrine. The platelets (>70%) formed clumps at increasing dilutions of ADP and epinephrine and suggested increased plaletlet aggregability. So a diagnosis of Sticky Platelet Syndrome (SPS) was made. Aspirin and physiotherapy were initiated and Warfarin was stopped. The patient and his parents were counselled regarding the disorder and the importance of compliance with medications particularly aspirin. On a follow up visit after 4 weeks he was recuperating well with near normal power in his limbs. A repeat ECHO was normal with no evidence of LV clot. The patient continued to do well with aspirin and was on regular follow up for nearly 3 years (July 2006). He was readmitted in December 2006 for a massive infarct in the right cerebral hemisphere. History from parents revealed non-compliance of medications for a period of 2 weeks. Unfortunately this time he succumbed to the thrombotic event.

Hyper-aggregable platelets have been described in association with a number of acquired disease entities whereby the cause-and-effect relationship is unclear[1]. In contrast sticky platelet syndrome (SPS) is an autosomal dominant platelet disorder associated with arterial and venous thromboembolic events[2]. It is characterized by hyperaggregability of platelets in platelet-rich plasma with adenosine diphosphate (ADP) and epinephrine (type I), epinephrine alone (type II), or ADP alone (type III)[2]. Clinically, patients may present with angina pectoris, acute myocardial infarction (MI), transient cerebral ischemic attacks, stroke, retinal thrombosis, peripheral arterial thrombosis, and venous thrombosis, frequently recurrent under oral anticoagulant therapy. Clinical symptoms, especially arterial, often present following emotional stress. Combinations of SPS with other congenital thrombophilic defects have been described[2]. The precise etiology of this defect is at present not known, but receptors on the platelet surface may be involved. Normal levels of platelet factor 4 (PF4) and beta-thromboglobulin in plasma suggest that the platelets are not activated at all times; they appear to become hyperactive upon ADP or adrenaline release. In vivo clumping could temporarily or permanently occlude a vessel, leading to the described clinical manifestations. The syndrome appears to be prominent especially in patients with unexplained arterial vascular occlusions[2]. Low-dose aspirin treatment (80 to 100 mg) ameliorates the clinical symptoms and normalizes hyperaggregability. Given the simplicity in diagnosis, treatment and follow up, internists should consider Sticky Platelet Syndrome (SPS) in their differential for recurrent arterial thrombosis.

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