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The Balance Between Thrombosis and Bleeding in Antiphospholipid Antibody Syndrome with Poorly Controlled SLE

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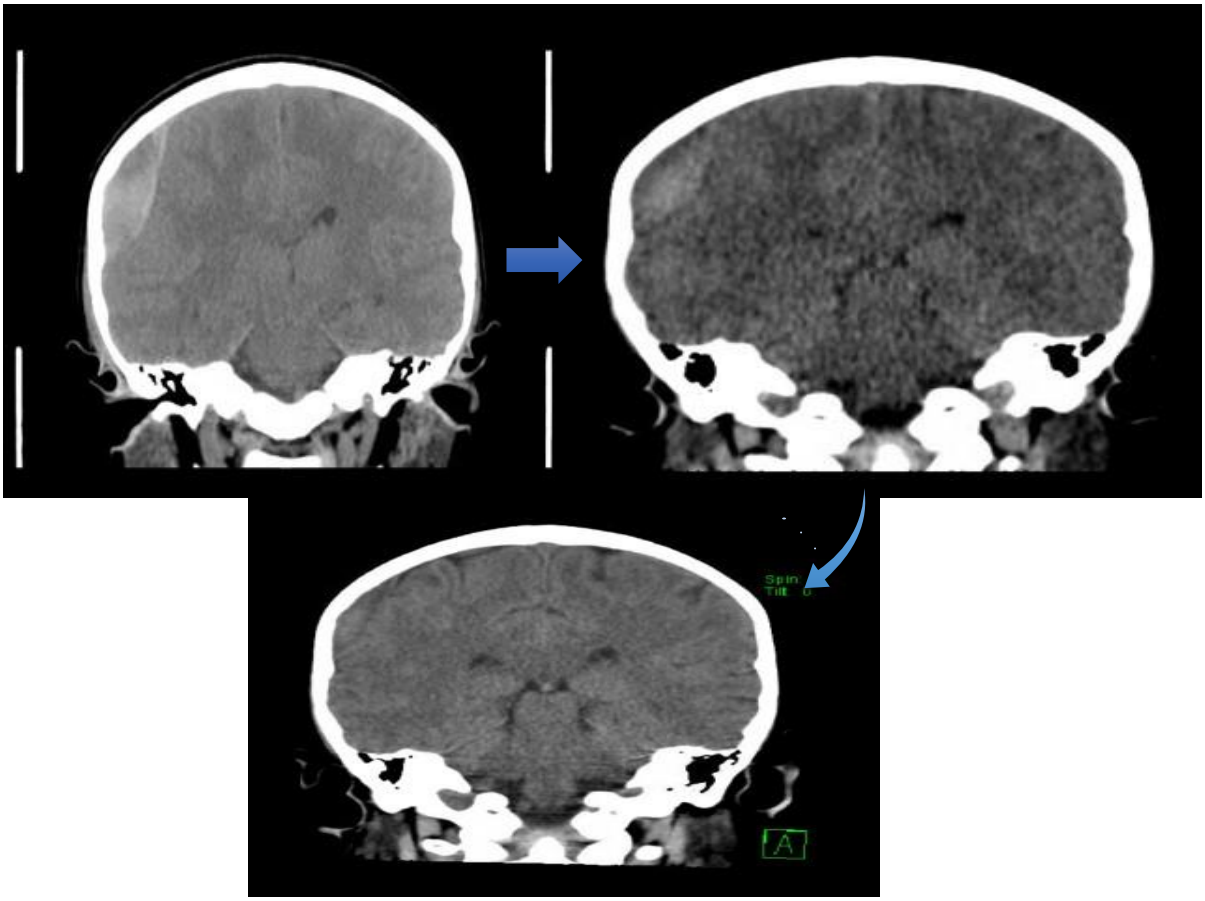
Introduction

- Antiphospholipid syndrome (APS) is one of the major causes of acquired thrombophilia, in which venous or arterial thrombosis, or both, may occur.
- Preventing thrombosis is one of the major aims in the treatment of APS.
- There is still no consensus about the duration and intensity of prophylactic antithrombotic treatment in children.
- Children are at higher risk of bleeding secondary to minor trauma.
- The available studies suggest that oral anticoagulation should be maintained in the long term in patients with APS who have experienced previous thrombotic events.
- Clearly, aggressive anticoagulation carries with it a higher risk of bleeding.
- There is no guidance on how to balance management of a bleed with maintaining anticoagulation in acute presentation of bleed and infection.
- We are reporting a case of secondary APS, given oral anticoagulant treatment and presented with extradural hematoma following possible minor trauma.

Case Presentation

- A 16 years old boy known to have APS (diagnosed according to criteria) secondary to JSLE. He presented at 6 years of age with raynaud's and arthralgia. He had positive Double stranded DNA. He had raised inflammatory markers. He was then diagnosed with JSLE and started on steroids, hydroxychloroquine and azathioprine.
- Soon after he developed his first deep vein thrombosis and was started on warfarin. Subsequent to that he had a second DVT. Warfarin dose was adjusted and his INR range maintained between 2-3. He continued to be steroid dependent and his treatment was changed later to Mycophenolate.
- He presented to the pediatric rheumatology clinic 8 months ago. His disease was poorly controlled with DsDNA of 126 units, ESR of 83 mm/hr, thrombocytopenia of 100, Hgb of 100 g/L. Rituximab discussed and agreed. Some delay occurred because of insurance. He received 2 doses of rituximab (500mg/m2)
- He presented 3 days later with fever and headache. He was found to have fronto-parietal extradural hematoma with mass effect. He was admitted to intensive care and managed under multidisciplinary team care including neurosurgery, pediatric rheumatology, hematology and intensive care.

- Hematoma was managed conservatively. His anticoagulation was discontinued in case surgical intervention was required. He remained in intensive care for 3 weeks.
- Anticoagulation was reintroduced gradually after discharge from intensive care.
- Initially enoxepirine used at small dose. Dose was gradually increased.
- Warfarin was then reintroduced.



- Patient was seen in the pediatric rheumatology clinic one week after discharge . He was doing well with no complaints .
- His labs at the visit showed WBC of 3 , HgB of 90 g/L , platlets of 100 , ESR of 10 mm/hr, INR of 1.9, DsDNA of 76 units and C3 of 0.5 g/L and C4 of 0.05 g/L
- He was started on prednisolone 20 mg BID and azthiaporine 25 mg daily, in addtion to hydroxychloquine 200 mg daily.

Discussion

- The major clinical manifestation of APS is vascular thrombosis. In adult cases, thrombosis on the venous circulation is found in 59%, arterial vessels in 28% and in both systems in 13%.
- The incidence of thrombotic events in pediatric patients with APS 2nd to SLE is not well studied.
- In a cross-sectional cohort study of 59 pediatric patients with SLE, the overall incidence of thrombosis was 17%.
- In adult cases, two systematic reviews of the literature antiphospholipid antibodies (aPL) showed that Lupus Anticoagulants (LA) are the strongest risk factors for both arterial and venous thrombosis, anti Cardiolipin antibodies (aCL) at medium to high titer appeared as possible risk factors of arterial thrombosis, aβ2GPI antibodies posed risk for venous thrombosis.
- Analysis of the aPL antibody profile, rather than of a single test, helps to establish risk of thrombosis.
- In pediatrics, a study of 59 SLE patients showed that 24% were persistently positive for lupus anticoagulant. 57% of those had one or more thrombotic events compared with only 4% of patients who were negative for a lupus anticoagulant.
- Whereas in a cohort of 29 pediatric patients with SLE, 65% had evidence of aPL. The presence of aPL, specifically aCL, was significantly associated with thrombotic events.
- High-intensity warfarin was not superior to moderate-intensity warfarin for thromboprophylaxis in patients with antiphospholipid antibodies and previous thrombosis.
- The low rate of recurrent thrombosis among patients in whom the target INR was 2.0 to 3.0 suggests that moderate-intensity warfarin is appropriate for patients with the antiphospholipid antibody syndrome
- Warfarin use, in multiple studies carried a 3% risk of bleeding.
- Thrombocytopenia in APS is often mild and benign (70-120×103/mm3) and is rarely associated with hemorrhagic complications.
- The prevalence of thrombocytopenia in APS estimated in the literature ranges from 20 to 40%.
- A series of 171 APS patients reported a percentage of 23.4% of thrombocytopenia cases; (17.6%) of them had severe thrombocytopenia (<5x103/mm3).
- We could not find evidence in the litereature discussing the management of such complex case.

Conclusion

- Patients with APS and JSLE have increased risk for thromboembolic events, anticoagulation therapy carries its own risk for bleeding. Managing both together is a challenge as in our case especially following immuno-suppression combined with minor trauma.
- We need to improve our advice to patients in preventing minor trauma as some consider roller coaster to be safe.
- Collaborative work is needed.
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- We need to improve our advice to patients in preventing minor trauma as some consider roller coaster to be safe.
- Collaborative work is needed.

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