

Letters to the Editor

Catastrophic antiphospholipid syndrome (CAPS): A rare and fatal encounter

Sir,

Antiphospholipid syndrome (APS) is a rare, multisystem, autoimmune, hypercoagulable disorder caused by production of antibodies against cell membrane made up of phospholipids, which will cause venous and/or arterial thromboembolism that can become disastrous if multi organ failure develops. We report a case of catastrophic variant of APS with good outcome. Patient developed pulmonary and peripheral vessels thrombosis in spite of anticoagulant and antiplatelet agents that was treated by surgical intervention which is reported in very few cases.

A 35-year-old female obese (102 kg) patient, without any bad obstetric history or other co-morbidities except 1st time detected diabetes mellitus, admitted for acute left middle cerebral artery infarct at peripheral hospital. She was not thrombolysed there because of late presentation. She was discharged after 2 days with improved limb weakness and was on subcutaneous low-molecular-weight heparin at home for 3 more days [Figure 1].

She developed breathlessness on exertion 2 days after discharge. She was diagnosed to have bilateral massive pulmonary embolism with respiratory failure, so referred to our hospital for further management.

She was not thrombolysed for pulmonary embolism because of recent cerebral vascular stroke. She was put on Bipap ventilator support and given heparin and supportive treatments. She also diagnosed to have right popliteal vein thrombosis on Doppler examination after admission to our hospital.

Subsequently ventilator requirement increased and she was not maintaining oxygen saturation with 100% FiO₂ and 15 positive end-expiratory pressure. So decided for surgical embolectomy by thoracovascular

surgeon. When surgical preparation was going on, she developed right lower limb femoral arterial thrombosis also. Hence, it was operated first followed by pulmonary embolectomy (thrombectomy). Postoperatively oxygen requirement decreased to minimal on ventilator with good femoral arterial flow also.

However, she developed severe sepsis, septic shock and acute renal failure postoperatively, which was treated accordingly. After stabilization, she was weaned from ventilator support and shifted to the ward. She was discharged on day-22. Considering multiple arterial/venous thromboembolic events, APS work up was done, which came positive. And according to criteria that was a catastrophic variant.^[1]

After 3 months of follow-up, again antiphospholipid antibodies sent, which came positive. So diagnosis was confirmed.^[2] Our patient developed multiple thrombotic events within a span of 1-week. We suspect APS initially and taken prompt actions particularly for pulmonary and femoral arterial thrombosis. We succeeded in tackling both the life threatening events of thrombosis with the help of vascular surgeon. In selected/resistant cases, immunosuppression with

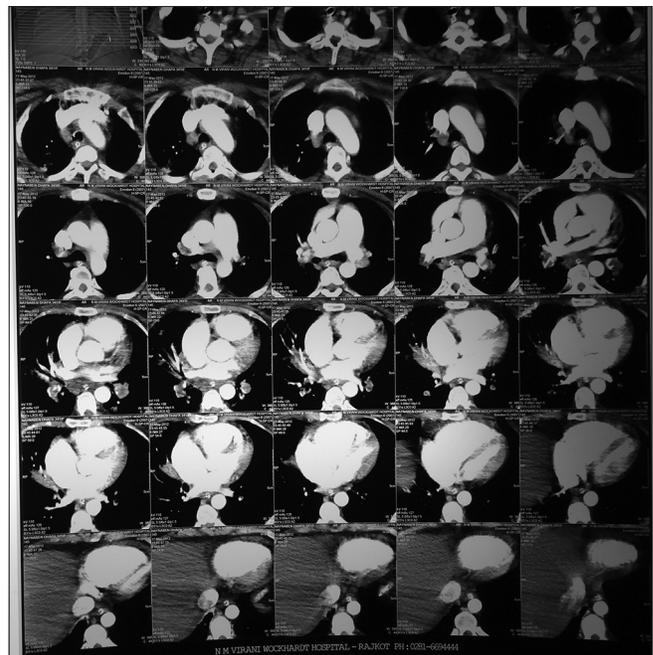


Figure 1: CT Angiography showing Bilateral pulmonary thrombosis

corticosteroids, immunoglobulines, plasma exchange, cyclophosphamide can be added.^[3]

Conclusion

Antiphospholipid syndrome should be suspected and evaluated if more than one thromboembolic event in any patient. Treatment of catastrophic APS requires an aggressive multidisciplinary collaborative treatment strategy for favorable outcome.

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