SAJEM EMERGENCY MEDICINE JOURNAL

CASE REPORT

CATASTROPHIC ANTIPHOSPHOLIPID SYNDROME

Amna Javed

Authors

Dept of Emergency Medicine Shifa International Hospital

Correspondence to:

Amnajaved174.174 @gmail.com

ABSTRACT

Catastrophic antiphospholipid syndrome (CAPS) is a rare presentation but it can lead to life threatening complications so high index of clinical suspicion is required. It is categorized as the most severe form of APS and is defined by involvement of three or more organs, systems or tissues, clinical symptoms occurring rapidly within a week, histopathological confirmation of small vessel occlusion in at least one organ or tissue and laboratory confirmation of presence of antiphospholipid antibodies and exclusion of other causes. Although the catastrophic variant presents in less than 1% of all patients diagnosed as APS, its high mortality requires that physicians are aware of this variant. Here, we present a case of 23-year-old male with no known comorbidities, who presented to our department with cough and haemoptysis for 1 month. Chest x ray showed wedge shaped opacification in right middle and lower zone with raised D-dimers of 1186mg / dL (up to 251 mg/ dL). CTPA was done which showed right pulmonary artery embolism along with right atrial filling defect. All coagulation profile was negative with anticardiolipin IgG >280 and strongly positive lupus anticoagulant.

KEYWORDS

Catastrophic antiphospholipid Syndrome, Haemoptysis, Lupus

INTRODUCTION

Antiphospholipid syndrome is a clinical disorder described in 1980s by several investigators and physicians.[1] Since then new discoveries regarding APS, its pathogenesis, diagnostic evaluation and treatment have been made. Currently APS is recognised as an autoimmune disease presenting with arterial and or venous thrombosis. APS displays a wide spectrum of variable symptoms. Catastrophic APS (CAPS, Asher son's syndrome) is a rare variant of APS defined as an acute insufficiency of at least 3 or more organ or organ system caused by small vein thrombosis detected in histological examination.[2] Risk factor of CAPS include infection, surgery, trauma, stress, malignancy and drugs including ineffective anticoagulation.1 The condition has dramatic clinical course and despite aggressive treatment, there is a high mortality rate of 30%.[1]

CASE REPORT

A 23-year-old male with no known comorbidities, presented to our emergency department with cough and haemoptysis for last 1 month. Cough was sudden in onset, productive in nature with streaks of blood in it, with no aggravating factors. Patient was prescribed intravenous antibiotic for possible lower respiratory tract infection, but cough didn't settle.

On examination BP: 110/70 mm Hg, Pulse: 80 b/min, R/R 18/min, Temp: A/F, Saturation: 94% on room air. On auscultation, there was decreased air entry at right middle and lower zone. Rest of the systemic examination was unremarkable. Figure.1

Chest X ray was done, which showed right middle and lower zone wedge shaped opacification. Baseline investigations were sent which showed a raised D-dimer of 1186mg/dL (up to 250 md/dL). CT Pulmonary Angiography was done which showed right pulmonary artery segmental and sub segmental infarct along with filling defect in right atrium. Figure. 2 & 3 Patient was admitted in critical care unit and started on anticoagulation. Extensive workup was sent for thrombophilia, APLA profile, ANA profile was sent. All were negative except APLA with lupus anticoagulant: strongly positive LA1/LA2 ratio: 2.6 (>2 strongly positive), anticardiolipin IgG >280 (>80GPL U/ml strongly positive). Patient underwent thoracotomy for suspicion of right atrial myxoma and small biopsy from inferior vena cava was also taken along with removal of right atrial filling defect. Histopathology confirmed thrombus in both

CASE REPORT

right atrium and inferior vena cava. Suboptimal opacification of peripheral vessels was also seen with no evidence of deep venous thrombosis. Patient was started on warfarin and discharged home. One-week later patient again presented with massive haemoptysis and was intubated for airway protection. Patient was treated on lines of catastrophic APS with plasmapheresis and pulse steroid due to recurrent pulmonary embolism. Patient was extubated on 3rd day of intubation and was switched to oral anticoagulant from heparin infusion wich had a target APTT of 50-60s. Echo was done which showed EF 55% with small pericardial effusion, mild tricuspid regurgitation and mild pulmonary hypertension. Patients condition was successfully stabilized and was discharged home on oral anticoagulation and steroids.

DISCUSSION

APS is an acquired autoimmune disorder, more common in females, that manifest clinically as arterial or venous thrombosis. Catastrophic APS is most severe form of APS with acute multiple organ involvement and small vessel thrombosis. Criteria for catastrophic APS[2]

- Three or more organ or tissue involvement
- Biopsy confirmation of thrombus
- Positive antibodies
- Exclusion of other diseases with multiple organ thrombosis

It is often difficult to distinguish CAPS from other condition which can lead to multiple organ thrombosis due to overlapping features. The major differences are highglighted in Table.1

Early diagnosis and aggressive therapy are essential to manage CAPS since mortality remains high. The treatment is generally directed at addressing thrombotic events and suppressing cytokine cascade. This typically involve combination of anticoagulation, systemic glucocorticoids, plasma exchange and IVIG.[3,4] Our approach towards management is

- Identify infection that may have precipitated CAPS and give appropriate antibiotic.
- Anticoagulation with heparin for treatment in acute setting. In patient who are stable and without evidence of recurrent thrombi or active bleeding transition to oral anticoagulant like warfarin.
- High dose glucocorticoids (methylprednisolone 0.5-1gram IV daily for 8days) followed by oral therapy with equivalent of 1mg/kg of prednisone per day.
- In severe cases plasma exchange and or IVIG (400mg/kg per day)
- Role of rituximab and eculizumab in refractory cases. CAPS associated with SLE can be treated with cyclophosphamide.[5]

CONCLUSION

Catastrophic APS is a rare but highly life-threatening condition if not recognized and treated early. It can have a variable presentation, but high index of suspicion can

Table.1 Major differences in various disorders[6]

Disseminated Intravascular Coagulation (DIC)	Heparin induced thrombocytopenia (HIT)	Thrombotic microangiopathy (TMA)
Widespread activation of coagulation and fibrinolysis that occur in sepsis or malignancy	Immune mediated thrombocytopenia that occur after heparin exposure. Heparin induced antibodies cause arterial/ venous thrombosis	Systemic syndrome in which small vessel platelet micro thrombi form in various vascular bed, leading to thrombocytopenia, microangiopathic haemolytic anaemia and organ injury
LIKE APS **Raised FDP** **Decreased fibrinogen** **Raised D dimers**	LIKE APS Moderate thrombocytopenia Widespread thrombosis	LIKE APS Thrombocytopenia Organ involvement
UNLIKE APS **Raised PT and APTT**	UNLIKE APS Heparin exposure is must	UNLIKE APS Do not cause large vessel thrombosis
Always associated with systemic disorder and presents with bleeding.	HIT is not associated with APL	Specific lab abnormalities related to underlying pathophysiology.

CASE REPORT

Figure.1

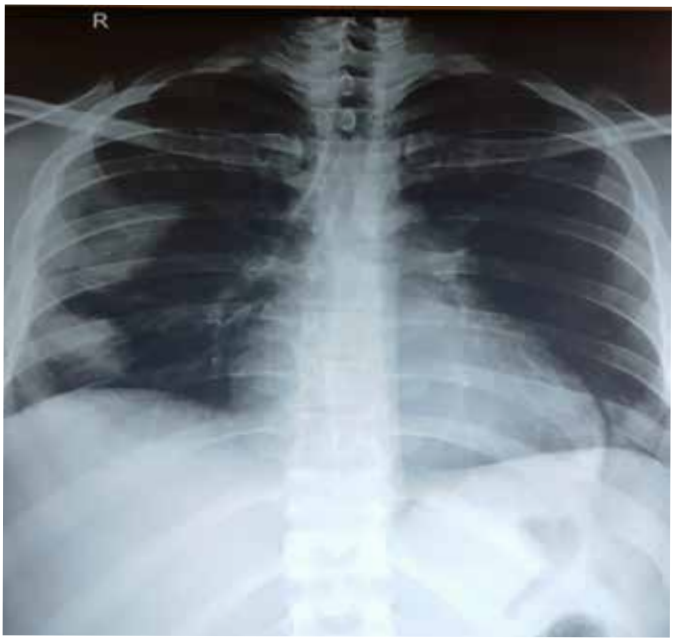


Figure.2

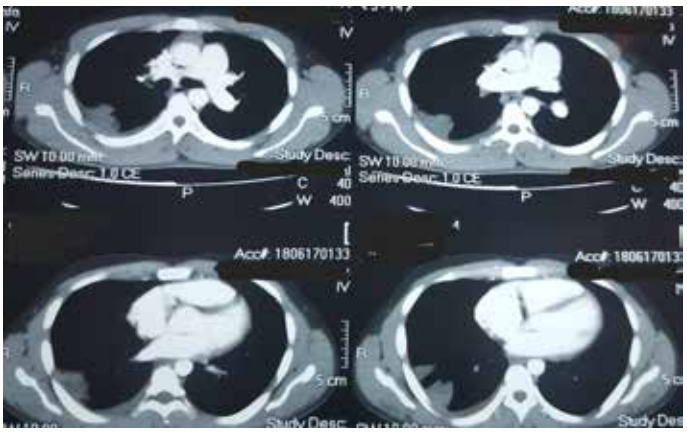
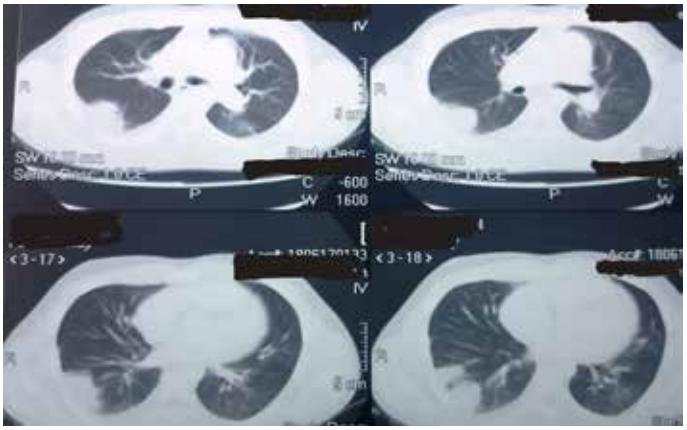


Figure.3



give a clue to the underlying disorder. Lab investigations including anti phospholipid antibody profile along with the other diagnostic criteria can establish the diagnosis. A multimodal treatment approach is required including anticoagulation, steroids, plasmapheresis or intravenous immunoglobulin. Despite aggressive treatment, mortality remains high.

REFERENCES

1. Cervera R, Rodriguez-Pinto I, Espinosa G. The diagnosis and clinical management of the catastrophic antiphospholipid syndrome: A comprehensive review. J Autoimmun. 2018 Aug; 92 :1-11

2. Asherson RA, Cervera R, G de Groot P, Erkan D, Boffa MC, Piette JC et al. Catastrophic antiphospholipid syndrome: international consensus statement on classification criteria and treatment guideline. Lupus. 2003 Jul; 12:530-534

3. Erkan D. Therapeutic and prognostic consideration in catastrophic antiphospholipid syndrome. Autoimmune Rev. 2006 Dec; 6 (2):98-103.

4. Ruffatti A, De Silvestro G, Marson P, Tonello M, Calligaroa A, Favaro M. Catastrophic antiphospholipid syndrome: Lessons from 14 cases successfully treated in a single center. A narrative report. J Autoimmun. 2018 Sep; 93: 124-30.

5. Guillot M, Rafat C, Buob D, Coppo P, Jamme M, Rondeau E et al. Eculizumab for catastrophic antiphospholipid syndrome—a case report and literature review. Rheumatology. 2018 Aug; key228. https://doi.org/10.1093/rheumatology/key228.

6. Erkan D, Zuily S. Uptodate [Internet].Clincial manifestatons of antiphospholipid syndrome. [cited on 2018 Aug 09]. Available from https://www.uptodate.com/contents/-clinical-manifestations-of-antiphospholipid-syndrome.

* ——— *