International Journal of Research in Medical Sciences
Hakeem SC et al. Int J Res Med Sci. 2022 Dec;10(12):2950-2952
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012

DOI: https://dx.doi.org/10.18203/2320-6012.ijrms20223103

Case Report

An unusual presentation of catastrophic antiphospholipid antibody syndrome in the background of sepsis

Sai C. Hakeem[1]*, Akash T. Oommen[1], Sandeep Surendran[2], Sumanth Madan[2]

[1]Department of Internal Medicine, [2]Department of Rheumatology, Amrita Institute of Medical Sciences and Research Center, Kochi, Kerala, India

Received: 04 October 2022
Revised: 04 November 2022
Accepted: 10 November 2022

***Correspondence:**
Dr. Sai C. Hakeem,
E-mail: saichandrah@gmail.com

ABSTRACT

A 59-year-old female presented with complaints of gangrenous changes over right ring finger and reduced renal output. She was in hypotension and had to be started on ionotropes. She also had acute kidney injury and was initiated on hemodialyis. Inspite of culture directed antibiotics and amputation of the necrotic region, her condition worsened. Considering the acute multisystem worsening, i.e., less than a week, concomitant autoimmune etiology was considered. Antiphospholipid antibodies were positive. Her tissue biopsy was suggestive of vasculitis. Hence the diagnosis of catastrophic antiphospholipid antibody syndrome (CAPS) was made. Quick recognition and appropriate treatment play a cornerstone in treatment of CAPS. She was pulsed with methylprednisolone and also treated with intravenous immunoglobulins and anticoagulants. She showed remarkable improvement and responded to the treatment. CAPS should always be kept as a differential in case of multisystem acute deterioration even in the background of sepsis. The treatment is a big challenge to physicians given the associated mortality rate if not briskly treated.

Keywords: Catastrophic antiphospholipid antibody syndrome, Sepsis, Gangrene

INTRODUCTION

Antiphospholipid antibody syndrome (APS) is an autoimmune multisystem autoimmune disorder of hypercoagulation. APS is predominantly seen in females.

The diagnosis of APS is based on the clinical features and laboratory criteria. Catastrophic APS (CAPS) is an accelerated form of APS resulting in multiorgan failure.[1]

The early diagnosis and rapid initiation of treatment is the cornerstone in management of CAPS as it has a very high mortality rate of more than 50%. The progression is very rapid and patients usually present in an already evolved state.[2]

We presented a case report of definitive CAPS. The patient presented with gangrene of right ring finger. There was rapid progression in the disease with multiorgan involvement.

CASE REPORT

A 59-year-old lady, known case of type diabetes mellitus and systemic hypertension came to ER in view of blackish discolouration of right ring finger along with erythematous lesions over bilateral lower and upper limbs. She also complained of reduced urine output and fever since last 2 days. On examination, her blood pressure was 80/50 mmHg. Local examination revealed necrosis of right ring finger (Figure 1). She was admitted in ICU for further

management. She was started on ionotropes in view of shock.

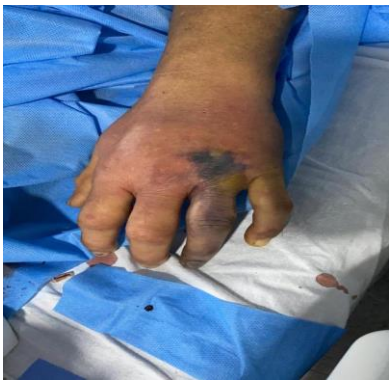

Figure 1: Gangrenous changes of the left index finger.

Her initial labs revealed elevated inflammatory parameters (procalcitonin 19, CRP ~ 270, ferritin 1227), deranged renal parameters (creatinine-4, urea-210), deranged hepatic parameters (bilrubin–9). In view of sepsic shock with multiorgan dysfunction syndrome, she was empirically started on Injection Meropenem and supportive measures. In view of reduced urine output with worsening renal parameters, she was initiated on haemodialysis.

Inspite of above measures, her sepsis was worsening and her erythematous lesions became necrotic (Figure 2).

Figure 2: Worsening of the gangrene.

The need for source control was the need of hour. Emergency amputation of right index finger was done and debridement of skin lesions was also done. Surprisingly, inspite of source control, her sepsis was worsening. Her blood culture was sterile. Her urine culture revealed *Klebsiella* and *Enterococcus*. Her tissue culture revealed *Klebisella* and she was treated with culture directed antibiotics. 2D echo was done to rule out infective endocarditis and there were no vegetations.

Given her multisystem involvement, with rapid progression of disease, the possibility of autoimmune disorder was considered. Her ANA was negative. Her beta-2 glycoprotein and anticardiolipin were elevated. Her lupus anticoagulant was negative. Her biopsy report showed fibrinoid necrosis, perivascular inflammatory infiltrates and multiple luminal thrombii suggestive of vasculitis. The patient developed paroxysmal atrial fibrillation and was cardioverted. Given the multi system involvement and rampant progression, and the fulfilment of criteria the diagnosis of definitive catastrophic antiphospholipid syndrome was made. Here the kidney, heart and skin were involved within 1 week and the antibodies were positive and there was also a positive tissue diagnosis.

The patient was pulsed with intravenous methylprednisolone 1 gram per day for 3 days, followed by intravenous immunoglobulin 2 gram/kg over 5 days. She was stared on heparin with warfarin bridging. She improved remarkably. Her ionotropes were tapered off and her blood pressure stabilised. Her inflammatory parameters normalised (CRP-6, procalcitonin-0.4). Her renal parameters normalised (creatine-0.5) and she was maintaining adequate urine output.

Unfortunately, the patient contracted covid infection and developed COVID pneumonia and succumbed to her illness.

DISCUSSION

Antiphospholipid antibody syndrome is a multisystem autoimmune disorder of hypercoagulation. APS is predominantly seen in females. The diagnosis of APLS is based on the clinical features and the laboratory criteria. Catastrophic APS is an accelerated form of APS resulting in multiorgan failure.[1]

In antiphospholipid syndrome, clinical characteristics are vascular thrombosis and pregnancy morbidity. Vascular thrombus is characterised by ≥1 clinical episodes of arterial, venous, or small vessel thrombosis, in any tissue or organ. Pregnancy morbidity is characterised by ≥1 unexplained deaths of a morphologically normal fetus at or beyond the 10th week of gestation, or ≥3 unexplained consecutive spontaneous abortions before the 10th week of gestation, with maternal anatomic or hormonal abnormalities and paternal and maternal chromosomal causes excluded. Laboratory criteria is characterised by Lupus anticoagulant positivity on ≥2 occasions at least 12 weeks apart and anticardiolipin antibody of IgG and/or IgM isotype, in medium or high titer (>40 GPL or MPL, or > the 99th percentile), on ≥2 occasions, at least 12 weeks apart and anti-β2-glycoprotein-I antibody of IgG and/or IgM isotype, in medium or high titer (> the 99th percentile), on ≥2 occasions, at least 12 weeks apart. Definite APS is present if at least one of the clinical criteria and one of the laboratory criteria are met.

The diagnosis and treatment of CAPS is challenging. Many patients present in advanced stage. The mortality rate of CAPS is also very high. In CAPS, the

autoantibodies against phospholipids activate the immune cells, platelets and endothelial cells hence leading to a procoagulant and proinflammatory state. These antibodies activate complements, inhibit anticoagulants and impair fibrinolysis.[3]

Table 1: Criteria for definitive CAPS.

S. no.	Criteria
1	Evidence of 3 or more organ involvement
2	Development of symptoms within 1 week
3	Confirmation by histopathology
4	Laboratory confirmation of antiphospholipid

The cornerstone in treatment of CAPS is to reduce the proinflammatory and procoagulant state. The routine anticoagulant therapy is not sufficient in the treatment of CAPS. Heparin is generally used for anticoagulation. Immunosuppression can be achieved by high-dose steroids, cyclophosphamide, IVIG and rituximab. A triple therapy strategy consisting of steroids, anticoagulation and either intravenous immunoglobulin or plasma exchange or both has shown good outcomes in management of CAPS and has reduced the mortality rate in CAPS to 30%.[4]

Patient presented with shock and multiorgan dysfunction. Her inflammatory and septic markers were elevated. Given the presentation, infective etiology was considered. Inspite of targeted antibiotics, there was no clinical improvement. As part of source control, amputation of gangrenous finger was done. However, there was no betterment, even after source control. This led to thinking of possibility multisystemic autoimmune involvement. Given the multisystem involvement, and rapid progression of disease in less than 1-week, histopathological confirmation and laboratory confirmation, the definite diagnosis of catastrophic antiphospholipid syndrome was made. Treatment of the critically ill patient was challenging given the background of septic shock, she was treated with methylprednisolone, intravenous immunoglobulins and heparin. She showed miraculous improvement. Her multiorgan dysfunction normalised and she became clinically better.

Hai et al published a care report of bilateral lower limb gangrene in peripartum period of a 25-year-old female.[5] This case report emphasis on need of early diagnosis and quick treatment of CAPS. Time is organ in CAPS, so a quick diagnosis helps to salvage the multiorgan dysfunction. The possibility of catastrophic antiphospholipid syndrome should always be considered even in background of severe sepsis.

CONCLUSION

Sepsis and catastrophic antiphospholipid syndrome have almost similar clinical picture. The possibility of catastrophic antiphospholipid syndrome should always be considered in background of severe sepsis. Early diagnosis and prompt treatment plays a key role in management of catastrophic antiphospholipid syndrome.

ACKNOWLEDGEMENTS

Authors would like to thank the head of the department, Dr. M. Gopala Krishna Pillai, for his support.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Asherson RA, Cervera R, de Groot PG, Erkan D, Boffa MC, Piette JC, Khamashta MA, Shoenfeld Y; Catastrophic Antiphospholipid Syndrome Registry Project Group. Catastrophic antiphospholipid syndrome: international consensus statement on classification criteria and treatment guidelines. Lupus. 2003;12(7):530-4.
2. Asherson RA, Cervera R, Piette JC, Font J, Lie JT, Burcoglu A, et al. Catastrophic antibody syndrome. Clinical and laboratory features of 50 patients. Medicine (Baltimore). 1998;77:195-207.
3. Nayer A, Ortega LM. Catastrophic antiphospholipid syndrome: a clinical review. J Nephropathol. 2014;3(1):9-17.
4. Rodríguez-Pintó I, Espinosa G, Erkan D, Shoenfeld Y, Cervera R; CAPS Registry Project Group. The effect of triple therapy on the mortality of catastrophic anti-phospholipid syndrome patients. Rheumatology (Oxford). 2018;57(7):1264-70.
5. Hai A, Aslam M, Ashraf TH. Symmetrical peripheral gangrene: a rare presentation of antiphospholipid syndrome. Intern Emerg Med. 2012;7:71-3.

Cite this article as: Hakeem SC, Oommen AT, Surendran S, Madan S. An unusual presentation of catastrophic antiphospholipid antibody syndrome in the background of sepsis. Int J Res Med Sci 2022;10:2950-2.