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A CASE REPORT OF PURPURA FULMINANS SECONDARY TO PROTEIN S DEFICIENCY, SECONDARY TO ACUTE MENINGOCOCCAL SEPTICEMIA

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INTRODUCTION

*N. meningitidis, i*s a human commensal colonizing the nasopharyngeal mucosa. From 1988-2011, the Philippines reported its biggest meninggococcemia outbreak last 2004-2006 in Cordillera region with 418 cases. With approximately 1.2M cases [1] each year worldwide and fatality rate from 4.1%-20%, [2] it presents with abrupt onset of fever, muscle and joint pains, petechial or purpuric rash. A severe feature of it includes purpura fulminans, with thrombosis and hemorrhagic necrosis & in severe cases with infarction and gangrene of limbs and digits due to disseminated intravascular coagulation (DIC).[3]The strong activation of coagulation results in depletion of hemostatic proteins leading to poorer outcome.

Many pediatric illnesses presents with rash and fever, some are self limiting while some are life-threatening without immediate intervention, like the meningococcemia hence a good history and PE are vital to prevent outbreaks and mortality.

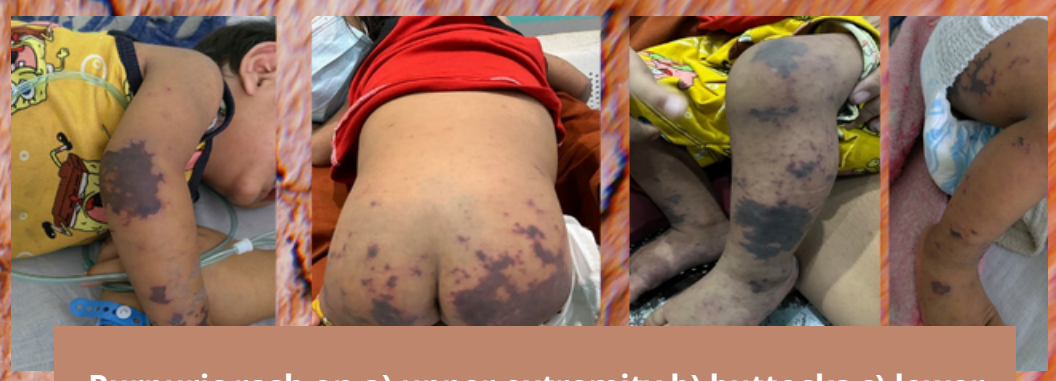
OBJECTIVES

1. To present a case of acute meningococcal septicemia
2. To discuss the pathophysiology of meningococcal disease
3. To discuss the diagnosis and treatment of meningococcal disease
4. To discuss the preventive management of meningococcal infection

CASE SUMMARY

A 1y4m old male was brought in due to rash. He was apparently well until 2 days prior, there was undocumented fever with decrease in appetite and activity with sudden appearance multiple purplish maculo-papular rash on the lower extremities. Mother opted to observe.

1 day prior, still with undocumented fever and purpuric rashes, noted progression of rash which also appeared on the bilateral cheeks and along the trunk and abdomen. Few hours prior, mother also noted progression of purpuric rash on the bilateral upper extremities and back hence decided to consult.



Purpuric rash on a) upper extremity b) buttocks c) lower extremities; d) gangrenous 2nd digit of the right foot

DISCUSSION

Meninggococcal infection causes meningitis and/or septicemia, and can lead to purpura fulminans, that can also lead to shock due to thrombosis, vascular leakage, and cardiovascular failure. In sepsis, there is upregulation of procoagulant pathways leading to thrombin generation. Thrombin then activates protein C, acts with protein S to inactivate factor Va and VIIIa. These antithrombotic mechanisms are dysfunctional in meningococcemia leading to prolonged coagulation and DIC.

Blood cultures are positive in 30-75% of cases while CSF cultures are positive in 70-95% of cases.[4] During the outbreak in Cordillera region, 62% were diagnosed with meningococcal disease based on confirmatory laboratory results, while 38% were diagnosed clinically. Similarly in the presented case, *N. meningitis* was not isolated in blood and skin lesions but meningococcal disease was diagnosed clinically.

Empirical antimicrobial therapy should be initiated immediately upon high suspicion. The treatment for both confirmed and clinically suspected meningococcal disease is ceftriaxone. [8]Close contacts of patients with the disease should receive proper antibiotic prophylaxis.

CONCLUSION

Meningococcal disease presents with abrupt onset of fever, rash and respiratory infections. Isolation of *N. meningitidis* from sterile body fluid confirms the diagnosis but it can also be diagnosed clinically. Ceftriaxone is the drug of choice for while close contacts should also receive proper antibiotic prophylaxis.

References

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