CASE REPORT

Fatal Waterhouse-Friderichsen syndrome due to Serotype C *Neisseria meningitidis* in a young HIV negative MSM (men who have sex with men).

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SUMMARY

Waterhouse-Friderichsen syndrome (fulminant meningococcaemia) is a fulminating infection, often leading to mortality in a matter of hours. In the past 3 years there has been a rise in cases of *Neisseria meningitidis* in the men who have sex with men (MSM) population in the USA and sporadic cases over the world especially in those who are HIV positive. We describe a case of a 34-year-old Caucasian man who presented with acute fulminant meningococcaemia, which proved fatal over a period of hours. This case report emphasises the need for further vigilance, prompt diagnosis and treatment of fulminant meningococcaemia without signs of meningitis especially in the MSM population and the need to expand vaccination recommendations of Meningococcaemia in the MSM population across the USA in both HIV positive and negative males.

undertaken. The patient did not respond to the resuscitative measures and eventually died.

The blood cultures that were drawn in the emergency room grew *Neisseria meningitidis* Serogroup C within 24 h. The autopsy report obtained from the medical examiner's office showed disseminated coagulopathy with purpuric skin rash and petechial haemorrhage with haemorrhagic adrenal glands. Microscopic examination of the brain did not show acute or chronic meningitis. Microscopic examination of the adrenals revealed acute haemorrhage consistent with Waterhouse-Friderichsen syndrome related to fulminant meningococcaemia.

GLOBAL HEALTH PROBLEM LIST

- *N. meningitidis* causing meningitis and Waterhouse-Friderichsen syndrome in the MSM (men who have sex with men) subgroup of the population.

CASE PRESENTATION

A 34-year-old Caucasian homosexual man presented to the emergency room with the c/c of pain in upper and lower limbs, nausea, vomiting and generalised weakness of 1-day duration. The patient had 10 episodes of vomiting. No fevers or chills. He also had petechial rashes on the upper and lower limbs. He was alert and oriented when he presented to the emergency room, but soon became lethargic and had to be intubated. The patient's blood pressure started dropping and the patient soon became critical with massive haemoptysis and haematemesis.

The patient's white cell count on admission was 7600/mm³ with a platelet count of 34 000/mm³. His serum bicarbonate level was 9. His blood work showed International Normalised Ratio of 2.5, fibrinogen of 76 mg/dL which pointed towards fulminant disseminated intravascular coagulation (DIC). Blood cultures were drawn. As his course worsened, his arterial blood gas showed a pH of 6.83.

The patient was given broad spectrum antibiotics including vancomycin, meropenem, fluconazole and doxycycline and also received one dose of ceftriaxone. He was subsequently admitted to the intensive care unit and was given fluids and vasopressors. He was also given three units of fresh frozen plasma, one unit of platelets, 10 bags of cryoprecipitate and 5 mg of intravenous vitamin K. During the course of the night he became bradycardic and went into asystole and then resuscitative measures were

GLOBAL HEALTH PROBLEM ANALYSIS

There is a recent rise in the incidence of meningococcal meningitis due to *N. meningitidis* in the USA in the MSM population.¹ Most cases are seen in HIV positive males and present with classic signs of meningitis along with disseminated meningococcaemia. It is important to note that HIV negative patients of the MSM population are at an equally high risk of infection with *N. meningitidis*. Also cases may present just as Waterhouse-Friderichsen syndrome without any signs of meningitis and thus it is necessary to recognise this high-risk population and the symptomatology of Waterhouse-Friderichsen syndrome with appropriate intervention as the disease progresses over a matter of hours. While the state of New York has come up with directives for vaccination of this new high-risk group,² there is a need to expand the vaccination directives to other places and also in the MSM population who are HIV negative.

N. meningitidis is the second most common cause of community-acquired adult bacterial meningitis.³ The typical initial presentation of meningitis due to *N. meningitidis* consists of the sudden onset of fever, nausea, vomiting, headache, decreased ability to concentrate, and myalgias in an otherwise healthy patient. Fulminant meningococcaemia (Waterhouse-Friderichsen syndrome) is one of the most devastating manifestations of *N. meningitidis*. It tends to strike young, previously well individuals and progresses over a matter of hours to death. Between

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2005 and 2011, the incidence of meningococcal disease in the USA was 0.3 cases per 100 000.⁴

In the fall of 2012, an outbreak of meningococcal disease was detected in MSM in New York City^{1 2 5 6} with 22 reported cases between 2010 and March 2013 of which 12 occurred in HIV-infected individuals⁷ and 7 cases were fatal.¹ In 2013 three cases of invasive meningococcal disease were reported in Germany, three cases in France and one case in Belgium.⁸ In our case, a 34-year-old MSM, HIV negative male presented with fulminant meningococcaemia (Waterhouse-Friderichsen syndrome) with acute onset of symptoms without any signs of meningitis with fatal progression of the disease within a matter of hours despite treatment. Fulminant meningococcal disease may present with initial symptoms such as leg pain, cold hands and feet, rash and may quickly progress to shock, DIC and purpura fulminans. This deadly infection can be seen in crowded environments such as dormitories, boarding schools and kindergartens. Military barracks is also one of these environments. Turhan *et al*⁹ reported two young male recruits who were diagnosed with meningococcaemia and meningitis due to *N. meningitidis* W135 even after being vaccinated with bivalent (A/C) meningococcal vaccine previously. Therefore quadrivalent *N. meningitidis* vaccination application is really important for individuals under risk such as military personnel, students staying on college and university campuses and MSM even if they are HIV negative.

Learning points

- Early identification of symptoms and treatment of fulminant meningococcaemia.
- Identification of new high-risk groups which now includes men who have sex with men (MSM) in HIV-positive and HIV-negative subgroups.
- Modifying recommendations of meningococcal vaccination in MSM even if they are HIV negative and expansion of the vaccination recommendations outside the state of New York to other states in the USA of America and around the world.

New York state Department of Health recommends quadrivalent meningococcal vaccination in (1) All HIV-infected MSM, (2) MSM, regardless of HIV status who have close/intimate sexual contact with men met through online websites, digital applications or at a bar.² There is a need to expand these guidelines outside the state of New York as more cases are appearing in other parts of the USA and worldwide.

Contributors All the authors were involved in the discussion. Abhinav Agrawal contributed in the management of the patient and writing of the case report. SJ participated in the management of the patient. Abhishek Agrawal reviewed current guidelines. ME performed reviewing of the manuscript and gave expert opinion.

Competing interests None.

Patient consent Obtained.

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