

Case Report: An unusual case of priapism

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The Case

A 6 year old boy presented with a 12 hour history of painful erection of the penis. This started spontaneously and had been preceded by a one day history of fever which was ongoing. He was able to urinate but with difficulty. There was no previous history of priapism or episodes of limb or joint pain. He had never been admitted to hospital before and had never had a blood transfusion. He was not taking any medications and his immunisations were up-to-date.

On examination he looked well grown for his age but was in obvious pain. There were no signs of jaundice or anaemia. He had a fever of 38.5°C and heart rate of 130bpm. He had an erect swollen penis which was painful to examine. The rest of the examination was unremarkable. A malaria parasite screen and sickle solubility test were both negative. Intravenous chloramphenicol and gentamicin were started together with intravenous fluids and morphine. A surgical consultation was requested but shortly after admission his priapism began to resolve and surgical intervention was not indicated. As he appeared to be improving intravenous fluids were discontinued. However on the second day of admission, his mother reported that his behaviour was abnormal. In particular he would get very agitated whenever he was made to drink fluids and was noted to choke and have involuntary spasms of his head and neck. He remained febrile with fevers up to 39.5°C and so antibiotics were changed to ceftriaxone. Over the next 12 hours he became progressively more agitated with fluctuating periods of confusion where he did not recognise his mother and was aggressive towards her. He was also noted to have excessive salivation and was unable to swallow anything. He was restarted on intravenous fluids and also on a diazepam infusion but a few hours later his conscious level deteriorated and shortly after this he died.

What is the diagnosis?

What are other clinical features of this condition?

How can this disease be prevented?

What are other causes of priapism and how should it be managed?

What is the diagnosis?

This boy was suffering from rabies encephalopathy. Further questioning of mother revealed that he had been bitten by a dog one month prior to presentation. He had not received anti-rabies vaccine as post exposure prophylaxis (PEP).

Rabies is a zoonotic disease and is part of the family of lyssaviruses. It affects the nervous system and leads to an encephalopathy which is invariably fatal. 99% of rabies cases occur in the developing world and it is estimated that there are 24,000 cases in Africa annually although this is likely to be an underestimate. It is transmitted in the saliva of a number of mammals including bats, foxes and wolves but over 90% of human rabies cases follow a bite by an infected domestic dog. Incubation time is usually 1-2 months but

can vary widely from a few days to over a year in some rare cases.

What are the other clinical features of this condition?

Two-thirds of rabies cases present with 'furious rabies', which usually starts with non-specific symptoms of fever, malaise and headache. There may be reported pain or paraesthesiae at the site of the bite. This is usually followed by rapid onset and progression of agitation, delirium and reduced conscious level. Hydrophobia, aerophobia and salivation are characteristic features of this disease. As well as hydrophobia and salivation, our patient also demonstrated aerophobia – when the examining physician gently blew on his face he became excessively agitated and developed phobic spasms. Convulsions may also occur, often shortly before death. Priapism, as in this patient, is an unusual but described symptom of rabies encephalopathy and is thought to arise secondary to autonomic dysfunction which may also cause temperature instability, cardiac dysrhythmias and sweating.

In up to a third of cases of rabies, patients develop a 'paralytic' form of the disease, in which the diagnosis may be much less obvious. Unlike in furious rabies, the level of consciousness in paralytic rabies tends to be preserved initially, and the patient presents with progressive ascending flaccid paralysis involving limbs and respiratory muscles, followed by coma; death in such cases is usually due to cardiac failure. Hydrophobia and aerophobia may or may not be present.² Patients may therefore be misdiagnosed as suffering from other types of CNS infection such as cerebral malaria or meningitis, or as having Guillaine-Barre syndrome.

Once a patient has developed rabies, death is inevitable. Treatment is therefore palliative and the patient should be nursed in a quiet side-room and the family counselled on the prognosis. Intravenous fluids should be started to avoid dehydration and the distressing feeling of severe thirst caused by the hydrophobia. Other supportive treatments include diazepam, which is often best given as a slow infusion with the intravenous fluids; analgesia and antibiotics if the patient is febrile or the diagnosis is unclear.

How can this disease be prevented?

Post-exposure prophylaxis (PEP) with anti-rabies vaccine should be offered to all patients who present with a dog-bite. Other immediate treatments of a bite wound include thorough washing and irrigation of the wound; tetanus immunisation; and antibiotics if indicated. Anti-rabies immunoglobulin is also offered to unvaccinated individuals as part of PEP, but this is not available in most resource-poor settings. There are a number of WHO approved regimes for rabies vaccine the most commonly used is five intramuscular injections of a 0.5ml or 1ml vial given on days 0,3,7,14 and 28. However there is a strong argument for using intradermal regimes which use sixty per cent less vaccine than the intramuscular regime, a saving which is especially important in settings such as Malawi where the vaccine is expensive and supplies are often limited. This regime involves giving two 0.1ml intradermal injections at two sites (deltoid and thigh) on days 0,3,7 and 28. It has been shown to be as immunogenic as the intramuscular regime and is also recommended by WHO.

While PEP can reduce rabies transmission, the only way to effectively prevent human rabies cases is by reducing rabies spread in the dog population through large-scale mass vaccination programmes.

What are the other causes of priapism in children and how should it be managed?

The most common cause of priapism in the paediatric population is sickle cell disease, when treatment must include intravenous hydration and analgesia. Blood transfusion/exchange transfusion may also be indicated if available. Priapism has also been described in patients with other haematological disorders such as leukaemia and Henoch-Schönlein purpura. For persistent priapism, decompression may be attempted by aspirating blood from the corpora cavernosa; if this fails, a surgical shunt is indicated in order to avoid infertility.

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