Fatal *Strongyloides stercoralis* infection in a patient with chronic inflammatory demyelinating polyneuropathy

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A 50-year-old villager with chronic inflammatory demyelinating polyradiculoneuropathy developed pain abdomen, diarrhea, and vomiting after 8 weeks of prednisolone (40 mg/day) therapy. After 10 weeks, he developed abdominal distension, leucocytosis, thrombocytopenia, liver dysfunction, coagulopathy, and respiratory failure. Stool examination showed larvae of *Strongyloides stercoralis*. He died in spite of antibiotics, metronidazole, ivermectin, vasopressor, and artificial ventilation. The patients on corticosteroid therapy are at risk of fatal septicemia due to *Strongyloides stercoralis* hyperinfection.

Keywords: *Strongyloides stercoralis* infection, CIDP, Ivermectin, Steroid, Outcome

Introduction

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is an important but rare disorder with prevalence of 1–7.7/100 000.¹ Intravenous immunoglobulin and corticosteroids are recommended for its treatment. Intravenous immunoglobulin is unaffordable to most patients with CIDP in the developing countries due to its high cost and these patients are managed with prednisolone.² Patients on corticosteroid therapy are prone to develop number of side effects, such as impaired glucose tolerance, hypertension, osteoporosis, Cushingoid appearance, and secondary infections. Wide arrays of infections are possible, such as tuberculosis, fungal or pyogenic. *Strongyloides stercoralis* is an intestinal nematode which infects a large population in tropical and subtropical areas. Autoinfection permits the parasite to persist in the asymptomatic state in the most patients. In immunocompromised patients, it can lead to life-threatening hyperinfection, disseminated infection, and secondary septicemia. We report a patient with CIDP on corticosteroid treatment who developed *Strongyloides stercoralis* hyperinfection-related sepsis and died.

Case Report

A 50-year-old Indian village businessman was referred to emergency department with acute abdomen. Ten months back, he developed pins and needles in the feet and hands which were followed a

month later by weakness. Over next few weeks, he needed support for walking and consulted the neurology service. Occasionally, he went to field bare footed. He was thin built (37 kg), afebrile with blood pressure of 110/70 mmHg and pulse 80/minute. His mental status and cranial nerves were normal. His lower limb power was Grade 3 on Medical Research Council scale and upper limb Grade 3 to 4. Biceps, triceps, supinator, knee, and ankle reflexes were absent. The pin prick and touch sensations were impaired below knee and distal to wrist joint. Joint position and vibration sensations were impaired in lower limbs. Bedside autonomic function tests, such as R–R interval variation, cold immersion test, and valsalva maneuver, were normal and there was no orthostatic hypotension. His hemoglobin was 15.7 mg%, leukocyte 9400/mm³, neutrophil 60%, eosinophil 6%, erythrocyte sedimentation rate (ESR) 20 mm at first hour, fasting blood sugar 104 mg%, serum bilirubin 0.64 mg%, creatinine 1 mg%, calcium 8.2 mg%, phosphorous 4.4 mg%, alkaline phosphatase 238 IU/l, transaminase 35 IU/l, sodium 132 mmol/l, and potassium 4.8 mmol/l. Electrolytes were normal. Cerebrospinal fluid (CSF) revealed 8 lymphocytes/mm³, proteins 62 mg% and sugar 60 mg%.

Nerve conduction studies were suggestive of acquired demyelinating neuropathy. There were prolonged distal latency of right and left median (6.4/8.6 ms) and, ulnar (6.2/8.0 ms) nerves, and peroneal nerve conduction was not recordable. Conduction block was present in both the median (right 42% and left 53%), ulnar nerves (right 37% and left 33%). Nerve conduction velocities were slow in the median (26.7/

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22.6 m/second) and ulnar (28.4/29 m/second) nerves. The ulnar sensory and sural conductions were unrecordable and median sensory conduction velocity was slow (32.6 m/second bilaterally). Serum electrophoresis showed M-band in fast gamma region and was 14.7% of total proteins and serum immunofixation revealed IgG-kappa M-band. Bone marrow biopsy was normal. The patient was diagnosed as CIDP with monoclonal gammopathy and was prescribed prednisolone 40 mg/day. At 8 weeks follow-up, his power and sensation improved marginally, though he needed help for walking and other activities of daily living.

After 10 weeks of therapy he developed diffuse pain abdomen, diarrhea and 2–3 bouts of vomiting daily. He was treated with metronidazole and ciprofloxacin for 7 days but without response; therefore, he was readmitted. He was febrile (38°C), pulse was 110/minute, BP 106/70 mm Hg, and respiratory rate 24/minute. There was abdominal distension and bowel sounds were reduced. His radiograph and ultrasound abdomen were normal. His blood and urine cultures were sterile. Liver functions were deranged (serum bilirubin 2.8 mg/dl, transaminases 99 IU/l) and had coagulopathy (aPTT 190 seconds, INR 2.25). He had leucocytosis (11 700/mm³) and thrombocytopenia (70 000/mm³). Absolute CD4 count was 81 cell/μl. He was managed with injection ceftriaxone, metronidazole, and amikacin but continued to have diarrhea. His stool examination revealed larvae of *Strongyloides stercoralis*; therefore, tablet ivermectin 6 mg daily was prescribed. On the fourth hospital day, his sensorium deteriorated, respiration became labored and developed hypotension (70/50 mmHg). Antibiotics were changed to ceftazidime and ofloxacin. He was put on mechanical ventilation with inotropic support. The patient died on the seventh day of admission.

Discussion

This patient with CIDP with monoclonal gammopathy who was on prednisolone died of *Strongyloides stercoralis* hyperinfection associated septicemia. The *Strongyloides stercoralis* infestation was detected in stool examination and septicemia was considered due to fever, leucocytosis, organ dysfunction, coagulopathy, and hypotension, although the organism could not be cultured. *Strongyloides* hyperinfection was attributed to intestinal (pain abdomen, diarrhea, adynamic ileus) and pulmonary symptoms (bronchopneumonia and adult respiratory distress syndrome).³ Our patient might have acquired infection in the community as he lived in village and went to field bear footed occasionally.

In the available literature, we could get only one report of CIDP patient in which, a 64-year-old man developed *Strongyloides stercoralis* induced hyperacute

pneumonitis following corticosteroid therapy for 4 years and he succumbed to this infection.⁴ In neurological practice, the other report of *Strongyloides stercoralis* induced respiratory failure is in a patient with polymyositis on corticosteroid therapy.⁵ A review of 151 patients with *Strongyloides* on corticosteroid therapy revealed mainly patients with hematological malignancy (23%), vasculitis (18%), and organ transplant (21%). These patients had nonspecific gastrointestinal symptoms, pulmonary symptoms and sepsis.⁶

Strongyloides stercoralis produces multi-organ dysfunction by two mechanisms: hyperinfection and secondary infection. In hyperinfection, the number of *Strongyloides stercoralis* larvae increases many folds in the intestine and migrate to extraintestinal sites resulting in systemic invasion. Patients with impaired cellular immunity, especially those receiving long-term steroids, HTLV-1, and HIV-infection are at risk of developing the hyperinfection syndrome or secondary septicemia. The mechanism of gram negative septicemia is attributed to transmigration of the intestinal bacteria through the breach caused by invading filariform larvae.⁷ The commonly associated gram negative organisms are *Escherichia coli*, *Klebsiella pneumoniae*, *Pseudomonas aeruginosa*, *Proteus mirabilis*, *Serratia marcescens*, and *Enterobacter cloacae*.⁷ In our patient, there were evidences of multiorgan dysfunctions probably due to septicemia, but repeated blood cultures were negative.

Our patient was on corticosteroid therapy for 10 weeks. Corticosteroid administration results in lymphopenia, loss of jejunal mast cell response to *Strongyloides* antigen, affects female worms to produce more infective larva, and steroids also produces molting signals which enhance the production of infective filariform larva.³ Repeated stool examinations for *Strongyloides* larva are diagnostic. The treatment of choice is ivermectin, but thiabendazole, albendazole, and mebendazole are also effective. Treatment of septicemia is by antibiotics and supportive care. However, the prognosis of these patients is poor and mortality is as high as 50–80%.⁷

Conclusion

The patient on long-term corticosteroid therapy is at risk of *Strongyloides* hyperinfection and secondary septicemia especially in developing countries. This complication should be kept in mind while treating a patient receiving long-term corticosteroids.

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