A Case of Cephalic Tetanus Referred as Rabies

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ABSTRACT

Tetanus is an entirely preventable sporadic neurologic disorder caused by an exotoxin, tetanospasmin, secreted by Clostridium tetani and characterized by increased muscle tone and spasms in persons with inadequate immunization status. The diagnosis is mostly clinical and there is no the gold standard for the recognition of disease. It is fatal if left untreated and the outcome is better if recognized and treated early. Full-blown picture of generalized tetanus is hardly a difficult situation for the recognition but when it manifests as localized tetanus without features of generalization, diagnosis can be missed in busy hospitals. This is especially true for the cases of cephalic tetanus when Bell’s palsy precedes the development of trismus and generalization and the history of trauma is not prominent or is overlooked. This communication deals with a review of literature on cephalic tetanus and a typical case of cephalic tetanus referred to us as rabies that made full recovery after 10 days of treatment with diazepam, metronidazole, and stat dose of human tetanus immunoglobulin. The history of injury was trivial which was forgotten and the initial presentation of facial palsy of lower motor neuron type was taken as Bell’s palsy. Moreover, the laryngeal and pharyngeal spasms were much more prominent and used to be precipitated by trivial stimulation such as a puff of air or drinking which were taken as aerophobia and hydrophobia leading to the anecdotal diagnosis of rabies.

Key words: Aerophobia, Cephalic tetanus, Facial palsy of lower motor neuron type, Rabies.

INTRODUCTION

Tetanus is a sporadic neurologic disorder caused by an exotoxin, tetanospasmin, secreted by Clostridium tetani, characterized by increased muscle tone and spasm in nonimmunized or incompletely immunized persons. Sometimes it may also occur in fully immunized persons who fail to maintain adequate level of immunity. Although it is entirely preventable by immunization, the burden of disease is large worldwide. Clinically three forms of manifestations have been recognized. Generalized tetanus is the most common form of the disease characterized by increased muscle tone and generalized spasms. Neonatal tetanus occurs as the generalized form and usually fatal if left untreated. Local tetanus is an uncommon form in which manifestations are restricted to muscles near the wound.

Cephalic tetanus is a rare form of local tetanus 1,2 manifested as trismus and dysfunction of one or more cranial nerves, often the seventh nerve. Most of the cases follow an acute head injury such as a puncture wound, laceration, abrasion or ear infection. The incubation period is a few days and the mortality is high.

Here we report a case of cephalic tetanus, first presented as Bell’s palsy and remained unrecognized in various health institutions and was mis-diagnosed as a case of rabies when aerophobia was noticed and the treatment was delayed due to the lack of correct diagnosis.

CASE REPORT

A 35 years male from western Terai presented with complaints of deviation of angle of mouth towards left for 25 days, trismus and generalized stiffness of whole body for last 10 days.

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He was apparently well till one month back when his wife noticed deviation of angle of mouth towards left and inability to close right eye. Then he started to have insidious onset gradually progressing stiffness of neck, back and extremities and became unable to open mouth after 10 days of deviation of mouth. He was admitted in a hospital and was diagnosed as a case of multiple cranial nerve palsy involving VII, IX, and X cranial nerves. Computer tomography (CT) scanning of brain showed bilateral fronto-temporal cerebral atrophy. Subsequently he developed dysphagia and difficulty in respiration associated with drooling of saliva and stiffening of whole body. He also started to have episodes of spasms and developed aerophobia and hydrophobia. He was further referred to another hospital where he was diagnosed as a case of rabies on the basis of aerophobia, hydrophobia and the findings of CT scan of brain. Then he was referred to our centre.

There was no history of altered sensorium and convulsions throughout the period of illness. Past history was unremarkable except for the history of dog bite 25 years back followed by complete antirabies vaccination. He gave a history of a blow with a metallic rod in his nasal bridge causing abrasions and lacerations happened one month back, i.e. five days prior to facial deviation. No medical help was sought because the wound was taken as minor.

Clinical examination revealed conscious oriented patient with normal vitals. Right facial palsy associated with trismus and generalized stiffening of whole body along with abdominal rigidity and episodes of spasms provoked by trivial stimuli such as a puff of blowing air were noticed. Investigations showed: random blood sugar – 102 mg/dl, hemoglobin – 14.5 gm%, total count – 9700/cmm, neutrophil – 74% and lymphocytes – 26%; cerebrospinal fluid (CSF) examination shows: watery colour with clear appearances, protein – 146 mg%, sugar – 51 mg%, total cells - 9/cmm with 30% neutrophils and 70% lymphocytes, culture - negative, Indian ink staining – negative; human immunodeficiency virus 1 and 2 serology (HIV 1 & 2) – negative, hepatitis B surface antigen (HBsAg) – negative, antibody against hepatitis C virus (Anti-HCV) – negative; liver function test was unremarkable.

He was diagnosed as a case of cephalic tetanus with multiple cranial nerves palsy and generalization. He was kept alone in dark room and treated with Human tetanus immune globulin (HTIG) 4000U intramuscularly stat and first dose of tetanus toxoid was given. Injection metronidazole 500 mg intravenous three times daily was started and spasms were controlled with injectable diazepam. On the third day of treatment he was able to take talk normally but the facial palsy persisted. However, abdominal rigidity and spasms were disappeared and aerophobia was absent.

Figure 1. A patient with cephalic tetanus showing right facial palsy of lower motor neuron type (Published with permission).
DISCUSSION

Cephalic tetanus, a rare variant of localized tetanus that involves the cranial nerves, has an incidence of 6%. Patients without appropriate immunization are at greater risk of contracting the disease. According to Centre for Disease Control (CDC) data, most of the cephalic tetanus reported in United States follows craniofacial cutaneous injuries such as laceration and puncture or infection such as acne and otitis media. Cephalic tetanus has been reported to be associated with dental caries, tooth extraction, root canal treatment, periodontal abscess, cheek trauma and tongue laceration. The incubation period is shorter (1-2 days) than in generalized form (7-10 days).

Cephalic tetanus invariably results in nerve palsies as well as increased facial muscle tone and spasms. The muscles innervated by motor nuclei of brainstem and often the cervical cord are mostly affected. The tetanospasmin reaches these nuclei along the axonal pathways and causes disturbance in myoneural conduction and axonopathy involving the nerves. The seventh cranial nerve is the commonest one, followed by the sixth, third, fourth, and twelfth in decreasing order of frequency. It has been thought that the paralysis is due to high local concentrations of toxin in the brainstem while lesser concentrations cause spasm by abolishing inhibition.

Cephalic tetanus mostly presents with facial pain, unilateral facial palsy, trismus and facial stiffness of healthy half of the face. Pharyngeal spasms leading to swallowing difficulties and laryngeal spasms causing asphyxia are the dangerous forms of symptoms and may lead to death. In 42% of cases, the cranial nerve deficits precede the onset of trismus. In such cases, cephalic tetanus is easily misdiagnosed. With its predilection for the seventh cranial nerve, it commonly mimics Bell’s palsy. Rarely facial palsy is bilateral. The initial rigidity and spasms of the muscle supplied by ninth and tenth nerves may be followed by palsies of these nerves. Occasionally the seventh, eighth and ninth nerves are affected from the very beginning. However the presence of trismus, which is invariable, leads to a correct diagnosis. About two-third of the patients progress to generalized tetanus and they have bad prognosis, although the overall mortality is 15-30%.

Electrophysiological studies showed that paralysis is of lower motor neurone type with denervation potentials, hyperirritability, loss of motor units, and marginally increased distal latencies being the features recorded. It is concluded that facial palsy in cephalic tetanus is mainly due to a functional block of conduction in the course of the peripheral nerve whereas the disturbance of neuromuscular transmission probably has little importance in these cases.

Cervical pachymeningitis, subarachnoid haemorrhage, cervical tuberculous arachnoiditis and dystonia are the diseases to be ruled out. The consideration of alteration in sensorium, fever, signs of meningeal irritation, signs of raised intracranial tension, nature of onset of clinical features, weight loss and other constitutional features will be helpful in differential diagnosis.

This case has a lot of interesting features and represents the scenario of modern medicine where uncommon presentation of uncommon disease is more emphasized in first than the uncommon presentation of common disease. First of all the development of isolated unilateral Bell’s palsy with forgotten trivial injury in nasal bridge without trismus led to misdiagnosis and subsequent anecdotal events. The next, when the full-blown picture of cephalic tetanus with generalization was developed, the finding of CT scan of brain become more confusing and the misinterpretation of appearance of spasms on air blowing taken as aerophobia and drinking water taken as hydrophobia led to the diagnosis of rabies.

The history of laceration in nasal bridge followed by the onset of Bell’s palsy after four days of injury and subsequent development of trismus after 10 days and the pharyngeal spasm and consequent stiffening of whole body along with abdominal rigidity developed on 20th day and history of spasms on minimal stimulation such as blowing with a puff of air leaves no doubt on the diagnosis. Moreover, the response to treatment confirms our diagnosis.

However, few uncommon features such as the findings of CT scan of brain, hydrophobia, aerophobia and the CSF findings remain mysterious. Although, few white blood cells (WBC) in CSF after repeated generalized spasms is explainable, the findings of CT scan of brain remain unanswered. One possible explanation could be the sequel of acute demyelinating encephalomyelitis (ADEM) developed after exposure to anti rabies
vaccine taken 25 years back. But, again there is no history of any alteration in sensorium in the past and the development of asymptomatic ADEM is not known. Moreover, it could be just incidental finding because we do not know how many people with normal mental examination have cerebral atrophy. The presence of aerophobia is explainable if we interpret it as a spasm precipitated by trivial stimuli.

The main purpose of reporting this case is to highlight the importance of high degree of suspicion of tetanus when there is facial palsy of lower motor neuron type. When indicated detail history of injury, whatever trivial they could be, should be explored and any history of chronic ear infection and dental caries should be taken. It is not uncommon to miss the history of trivial injury in busy clinic like in our case, which was initially diagnosed as Bell’s palsy. In addition to this, another lesson that can be learnt from this case is that the diagnosis of fatal disease should be taken only after careful consideration. Making a diagnosis of rabies is almost equivalent to writing a death certificate because rabies itself is a fatal disease and once the diagnosis of rabies is made, the patient is less likely to be reexamined and reviewed in detail because of fear of acquiring the disease.

REFERENCES