

# Atypical Presentation of Tetanus

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Tetanus is a life- threatening nervous system disease caused by *Clostridium tetani* (*C. tetani*). It is characterized by an acute onset of hypertonia, painful muscular contractions (usually of the neck and jaw muscles), and generalized muscle spasms without other apparent medical causes. However, various atypical forms of clinical presentation were reported

worldwide and need to be clarified. Being aware of these atypical presentations helps to achieve early diagnosis and management of this dreadful condition. Here, we emphasize different experience through many worldwide case reports about atypical presentation of tetanus.

## INTRODUCTION

Tetanus is a deadly however preventable disease caused by a neurotoxin protein produced by *C. tetani* an obligate anaerobic, motile, gram- positive, non- encapsulated and spore forming bacillus. The spores are resistant to heat, desiccation, and disinfectants. [1,2] Hippocrates has named and described the disease in the 4th century B.C. [3] Interestingly, tetanus symptoms were identified retrospectively as tetanus about 5000 years ago. [1] World Health Organization's (WHO) planned to eradicate tetanus by the year 1995, However, it still endemic in the developing world with many fatalities. [4]

## CLINICAL PICTURE

Tetanus is usually recognized by an injury that is infected with soil, rust or manure. Injury can be due to ulcers, burns, animal bites, septic abortion, otitis media, surgeries, child birth, intramuscular injection and even trivial wounds. [5,6,7] The incubation period varies (one day - several months) with average about one week. The shorter the incubation period the more severe is the disease. The life span of the disease is usually two weeks however several months may be needed for full recovery. Four clinical types of tetanus were recognized; generalized tetanus, localized tetanus, cephalic tetanus and neonatal tetanus. [8].

**Generalized tetanus:** is the most prevalent form of tetanus and usually presents by lock-jaw (trismus), risus sardonicus (orbicularis oris muscle spasm) and painful generalized muscle contraction. Arching of the back due to leg and back muscles hyper-extension with flexed arm muscles (opisthotonos) [9,10]. Airway obstruction, sympathetic hyperactivity, autonomic dysfunction with lability of blood pressure and cardiac rhythm was reported [8].

**Localized tetanus:** is a rare form of tetanus that is limited to the site of the original wound. It usually deteriorates to generalized tetanus. If the initial wound is in the head, the disease can progress into the cephalic form of tetanus with affection of the muscles supplied by the related cranial nerves [8].

**Cephalic tetanus:** includes manifestations mimicking that of stroke due to dysfunction of cranial nerves [11]. Cephalic tetanus is sometimes due to localized tetanus in the area of head and scalp. It can be complicated and progress to the generalized form [8].

**Neonatal tetanus:** can result from infection of the umbilical stump in a baby born to unvaccinated mothers [12]. After an incubation period of about seven days, the affected baby presents clinically with failure of suckling, opisthotonos and generalized muscle rigidity. High mortality is usual due to complications such as apnea and sepsis [8].

## ATYPICAL PRESENTATION OF TETANUS

### 1- Cases of tetanus without a clear history of trauma.

Tetanus is usually diagnosed by the typical history of injury and the characteristic symptoms of the disease. When patients have no clear history of trauma, we have to diagnose tetanus on the basis of his symptoms and clinical course. Some cases of tetanus were reported – from Germany and Japan -without a clear history of trauma and/or an obvious point of entry. Based on the symptoms and clinical follow up, Tetanus was diagnosed and treatment was started immediately [13,14].

### 2- Cases of cephalic tetanus presenting with multiple cranial nerves dysfunction.

A 77- years- old patient from Japan presented with trismus, and developed right Horner's syndrome within a week. Symptoms due to

multiple cranial nerve palsies were observed: right inferior oblique muscle weakness, reduced right corneal reflex, right facial palsy, dysphagia, and abnormal tongue movements. Neuroimaging findings were normal. Given the history of head trauma, symptoms and clinical follow up, cephalic tetanus was diagnosed and treated immediately. His condition improved without any complications [15].

### 3- Worsening headache as an initial atypical presenting symptom of tetanus.

A 42-year-old woman from Japan presented with severe headache. She gave a history of recurrent tension headache and migraine. One day earlier, she was working in rice field where she gave a history of minimal unnoticed farm trauma. Treatment of headache was tried before and after admission but with minimal effect. On the 5<sup>th</sup> day of admission, she developed trismus and other symptoms that confirmed the diagnosis of tetanus. Treatment was started promptly and headache was significantly improved by the following day [16]. A similar case of generalized tetanus was reported from a tropical island in Malaysia. The case was presenting initially with headache. She was a woman of 43 years old presented with headaches and generalized weakness of the body that developed into trismus and neck spasm. She gave a history of wound on her foot in a contaminated area. She did not receive anti-tetanus prophylaxis. Once diagnosed as tetanus, she was given immunoglobulin, tetanus toxoid, metronidazole, and sedatives in the proper dosages. Her neurological condition improved and she was successfully discharged with complete recovery after 6 months of follow-up [17].

### 4- A case presenting with a striatal toe: an unusual presentation of tetanus.

A case of a 15-year-old girl from India was initially diagnosed to have a striatal toe. Her condition progressed and she later developed clinical features consistent with tetanus. A history of blunt trauma to nose was confirmed retrospectively. Treatment was started immediately with eventual complete recovery.

Striatal toe is an apparent extensor plantar response, without fanning of the toes, in the absence of any other signs suggesting dysfunction of the cortico-spinal tract [18].

### 5- Dysphagia as an unusual initial and sole presentation of tetanus.

Many cases were reported - from Italy, Switzerland, Germany and Japan - to have dysphagia as an initial and sole presenting symptom in tetanus. Dysphagia can present an early symptom of tetanus, but it is actually rare to be the only and initial symptom of the disease. Dysphagia and tremors were the only initial presenting clinical symptoms of an old man in Italy. During hospital admission, he faced a rapid clinical deterioration of his condition. The classical clinical presenting signs (trismus, opisthotonos, neck spasm) were absent but the diagnosis of tetanus was suspected. Once diagnosed, treatment was started immediately with excellent clinical recovery [19]. Similar conditions were reported from Switzerland, Germany and Japan [20- 22].

#### **6- An unusual case of chronic relapsing tetanus associated with mandibular osteomyelitis.**

A patient from Japan with history of radiotherapy (due to malignant lymphoma of the neck) developed tetanus eight years after the therapy. Radiotherapy was incriminated to result in mandibular osteomyelitis necrosis with formation of infectious focus of tetanus. Treatment with penicillin G, tetanus toxoid and chronic administration of metronidazole was very effective but symptoms of tetanus returned when the administration of metronidazole was stopped because the infectious focus could not be completely eradicated. This was an interesting case report of chronic relapsing tetanus, and demonstrates that tetanus can occur due to mandibular focus but the chronic administration of metronidazole can prevent the relapse [23].

#### **7- An unusual case of tetanus masquerading as an acute abdomen: A case report.**

A case of a 67-year-old farmer from Nepal- with history of sustained laceration injury before about 45 days - presented to the emergency department with abdominal pain and rigidity. He was diagnosed with tetanus based on clinical symptoms. His condition ultimately required mechanical ventilation. The patient was then managed in the intensive care unit and later achieved a gradual recovery[24].

#### **8- A diagnostic challenge of a case of localized tetanus mimicking acute perforated viscus.**

Abdominal tetanus is the rarest presentation of localized tetanus in which other muscles are not involved. Diagnosis is challenging as it mimics

acute abdomen. This case report from Malaysia is about a male patient of 40-year-old who presented to the emergency department with generalized abdominal pain for 3 days. His physical examination revealed a guarded abdomen with epigastric tenderness. Systemic examination was unremarkable. The initial diagnosis was acute abdomen with suspected perforated viscus. On the 3<sup>rd</sup> day of admission, he developed opisthotonos with marked elevation of serum CK levels. A diagnosis of localized tetanus was confirmed and a gradual full recovery was achieved after prompt management [25].

#### **9- Dysarthria as a clinical presentation of a case of tetanus.**

An 80-year-old man from Portugal with history of hypertension and dyslipidemia presented with dysarthria. Cranioencephalic CT suggested pontine and mesencephalic ischemia. Stroke treatment was started. Two days later, the patient displayed dysphagia then the following day he developed an apparent tonic seizure with respiratory distress refractory to diazepam and phenytoin, requiring sedation and invasive mechanical ventilation. Ultimately, he manifested trismus and generalized spasms. MRI did not reveal any brain injury. Tetanus diagnosis was established and promptly managed. He gradually improved over a 70-day hospital stay [26].

#### **10- A case of cephalic tetanus presenting as unexplained ptosis.**

A patient from Singapore with a history of dental procedure was referred for a left bell's palsy. He complained instead of deteriorating unilateral ptosis and dysphagia. Eventually, trismus was also noted. He was diagnosed with cephalic tetanus [27]. Similarly, an old female in Korea - with a history of head trauma- presented with left ptosis followed by facial palsy. Eventually, she developed dysphagia and trismus.

#### **11- Autism and *Clostridium tetani*.**

A significant percentage of individuals with autism have a history of extensive antibiotic use that may disrupt protective intestinal microbiota, giving the chance for colonization by opportunistic pathogens including *C. tetani* that produce tetanus neurotoxin (TeNT). The vagus nerve is capable of transporting tetanus neurotoxin (TeNT) to the CNS bypassing its normal preferential binding sites in the spinal

cord and obscuring the symptoms of typical tetanus. Once in the brain, TeNT disrupts the release of neurotransmitters by the proteolytic cleavage of synaptobrevin, a synaptic vesicle membrane protein. Neurotransmitter release inhibition would explain a wide variety of behavioral deficits apparent in autism that may be improved when treated with antimicrobials effective against intestinal clostridia[29].

#### **12- Periocular discomfort- in a patient who underwent surgery for sleep apnea- as a primary presentation of tetanus.**

A 63-year-old man from S. Korea underwent an endoscopic surgery for the treatment of his newly diagnosed obstructive sleep apnea. After 3 weeks of surgery, he presented with right side periocular discomfort. Four days later, he started to suffer from dysphagia, trismus, ptosis and jaw pain. Based on the previous data, tetanus was diagnosed. Once diagnosed, tetanus was promptly managed. His symptoms improved and the cure was achieved within a month without any complications[30].

#### **13- Rectus sheath hematoma as an unusual adverse event of tetanus.**

A case of tetanus was reported from Japan about a patient with shock complicated with a rectus sheath hematoma caused by rupture of a pseudo-aneurysm of the inferior epigastric artery. A rectus sheath hematoma might be misdiagnosed as unstable blood pressure associated with autonomic hyperactivity, which is usually observed in patients with tetanus. The possibility of the occurrence of bleeding complications should be considered if a patient with tetanus has severe and persistent hypotension[31].

#### **14- Tetanus presenting with respiratory failure: Mayo Clinic experience.**

Mayo Clinic case report of a 65-year-old woman presented with dyspnea, persistent hiccups and pleurisy of 3 days duration. The condition was caused by tetanus due to inadequate secondary immunity. She required intubation for progressive trismus and laryngospasm-associated respiratory failure. Infusion of lorazepam did not control her spasms. Resistant spasms and hiccups improved with fentanyl and cisatracurium treatment. Three weeks later, the patient was weaned from the ventilator with complete recovery. In a review of a 25-year experience at Mayo Clinic, respiratory failure requiring intubation developed in many cases of tetanus.

The earlier the diagnosis and management of such cases the better the prognosis of their conditions [32].

#### **15- Atypical tetanus in a completely immunized 14-year-old boy.**

This is a case report from Australia of rare clinical course of tetanus in a completely immunized 14-year-old boy. His initial symptoms, which included a flaccid paralysis, supported a diagnosis of botulism. Preliminary mouse-test results with combined botulinum antitoxins A, B, and E, obtained from tetanus-immunized horses, supported this diagnosis. The clinical course changed from paralysis to rigor and the negative, more specific, botulinum mouse test with isolated botulinum antitoxins A, B, and E, obtained from nonvaccinated rabbits, excluded the diagnosis of botulism. Tetanus was suspected despite complete vaccination. The final results of a positive mouse test performed with isolated tetanus antitoxin confirmed the diagnosis of tetanus. Adequate treatment was started, and the boy recovered completely.[33]

#### **16- Spastic paraplegia (mimicking myelitis) as an initial presentation of a case of localized tetanus.**

A 35-year-old male from Japan presented with mild paresis of the lower limbs and hyperreflexia involving all his extremities without trismus or opisthotonos. Spinal magnetic resonance imaging (MRI) revealed a high area of intensity at th7-9. Diagnosed was established as a spastic paraplegia. Cerebrospinal fluid (CSF) showed pleocytosis. Constipation and dysuria developed and the bilateral leg spasticity worsened. Methylprednisolone pulse-therapy was administered. However, the symptoms had evolved on the 11th day of hospitalization into optic hyperesthesia and opisthotonos. On the 13<sup>th</sup> day of hospitalization, tetanus was suspected and managed promptly with a successful outcome. He gradually improved over a month. In a case of progressive spastic paraplegia with a subacute course showing a faint abnormality on spinal MRI, the diagnosis of localized tetanus should be suspected. [34]

#### **Conclusion:**

Tetanus still a morbid and mortal infectious disease. Despite it was controlled- for a far extent- in developed countries, yet it remains a complicated health problem in developing countries due to contaminated environment, low



socioeconomic level, late and/or inappropriate management. The various presenting manifestations stress the need to be aware of its multiple atypical presentations. This in turn can facilitate the early diagnosis and management of the disease.

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#### Highlights:

- Tetanus is a life-threatening yet a preventable infectious nervous system disease caused by *Clostridium tetani*.
- Tetanus is usually recognized by an injury that is infected with soil, rust or manure.
- Four clinical types of tetanus were identified; generalized tetanus, localized tetanus, cephalic tetanus and neonatal tetanus.
- Different atypical presenting clinical pictures of tetanus were reported worldwide.
- Awareness of typical and atypical presenting manifestations of tetanus allow early diagnosis and management of the disease.

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