Case Report

A case of dystonic spasms in an adolescent: A neurologist's enigma!

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ABSTRACT

A 15-year-old boy presented with episodic abnormal movements of the right upper limb, neck, and jaw for 10 days associated with pain and spasms, following a penetrating injury to his right hand 2 months ago. He was successfully managed for tetanus with antitoxin, antibiotics, wound debridement, and supportive care. He was found to have protective tetanus antibody levels (1.1 IU/mL). To encounter tetanus in a vaccinated individual with an atypical focal presentation is a cause for concern and requires awareness to avoid delay in diagnosis and management.

Key words: Rigidity, Spasms, Tetanus, Tetanus immunoglobulins, Tetanus toxoid, Vaccination

Although tetanus is a rare vaccine-preventable disease, it still occurs commonly in developing countries, especially in lower socioeconomic strata and rural parts. The disease is characterized by muscle spasms and dysautonomia caused by neurotoxins released from *Clostridium tetani.* Treatment aims to control spasms, reduce autonomic instability, and reduce toxin spread through wound debridement, tetanus immunoglobulins, and antibiotics [1].

The present case is unusual in its presentation as despite the vaccination at the age of 10 years, he presented with spasms of his arm and jaw which persisted and progressed for 2 months. Tetanus in such a clinical scenario cannot be ruled out and should be considered a differential. The case emphasizes the importance of rarest yet fatal and treatable infections which can add to the morbidity and mortality of the disease.

CASE DESCRIPTION

A 15-year-old boy presented with episodic abnormal posturing movements of the right upper limb, neck, and jaw for 10 days associated with shock-like spasms and swelling of the dorsum of the right hand. The boy had a penetrating injury to the right hand with a rusted nail 2 months ago. There is no family history of abnormal movements or high-risk behaviors. He had visited three doctors and was diagnosed with seizures and psychogenic non-epileptiform seizures and treated with antibiotics and antiepileptics.

Access this article online	
Received - 08 December 2023 Initial Review - 20 December 2023 Accepted - 05 February 2024	Quick Response code
DOI: 10.32677/ijcr.v10i3.4380	

On examination, the blood pressure was 144/92 mmHg and the pulse was 110/min, along with a swollen dorsum of the right hand. Right laterocollis, shoulder elevation, trismus, and flexed rigid right arm and fingers were observed with brisk deep tendon reflexes in all 4 limbs (Fig. 1).

On evaluation, the erythrocyte sedimentation rate was 38 mm/h, and serum creatine phosphokinase was 1540 U/L. Routine blood investigations and magnetic resonance imaging of the brain and cervical spine were normal.

The patient was clinically diagnosed with tetanus, and 3000 U intramuscular and 500 U perilesional infiltration of tetanus immunoglobulins were administered together with surgical debridement of the wound on the hand and intravenous metronidazole, magnesium, and oral clonazepam. The autoimmune encephalitis panel, viral markers, anti-nuclear antibody, and copper studies were within normal limits, and tetanus immunoglobulin G levels were 1.1 IU/mL by enzyme-linked immunosorbent assay (ELISA). The wound culture was sterile, and the biopsy showed chronic inflammatory changes.

During the hospital stay on day 2, the patient developed laryngospasm and hypoxemia with increased oral secretions. Immediate endotracheal intubation was performed, and the patient was managed with midazolam and atracurium infusions together with higher doses of oral clonazepam and baclofen administered through a nasogastric tube. On the 6th day, the patient was successfully extubated without the need for a tracheostomy. He was discharged on oral clonazepam, baclofen, tolperisone, and magnesium on the 10th day without any active symptoms such as rigidity or spasms. Thereafter, he was revaccinated as per the primary schedule.

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Figure 1: A 15-year-old boy with right laterocollis, right shoulder elevation and flexed arm

DISCUSSION

Tetanus is an acute disease characterized by muscle spasm or rigidity and dysautonomia that are caused by tetanospasmin produced by the Gram-positive bacterium *C. tetani*. It is found to be more prevalent in tropical climates and was reported to cause 58,900 deaths worldwide. In developed countries, the incidence of tetanus is very low [2,3]. In India, the incidence of tetanus cases was 16,579, with deaths in 7332 cases in 2019 [4]. Post-penetrating injury, at-risk groups include neonates, the elderly (>60 years), IV drug users, and diabetes. However, no obvious cause or trauma may be seen in up to 20–25% of cases. Tetanus has occurred rarely in vaccinated individuals, as in our case [5].

The incubation period of tetanus varies from 3 to 21 days [6]. Post-injury, the toxin is transported retrogradely, and thereby, it suppresses the inhibitory control of interneurons at the anterior horn cells and brainstem, causing unregulated activity of motor and autonomic neurons, causing muscular rigidity and autonomic dysfunction [7]. It can present as either generalized tetanus, in more than 80% of cases, or cephalic or local tetanus, which are the rarer types. In our case, it presented with a rarer local type with predominant brachial involvement, which progressed to generalized tetanus at the time of diagnosis. The atypical presentation could have been due to the pre-existing antibody causing a localized presentation.

Diagnosis is purely clinical and commercially available laboratory tests suggest protective antibody level status by ELISA (Ig G >0.1–0.2 IU/mL) [1]. The occurrence of tetanus despite being vaccinated could be due to immune failure or high doses of toxin exposure. Management includes halting and neutralizing toxin production through tetanus toxoid, antibiotics, and wound debridement. Management of spasms is done using benzodiazepines, magnesium sulfate, and neuromuscular blockade. All patients with tetanus should receive active immunization with a full vaccine series as tetanus infection does not confer immunity [5,8]. On diagnosis of tetanus, immunization should commence without delay, and vaccines should be administered at a site different from the tetanus immunoglobulin [9].

Establishing a diagnosis of tetanus in a vaccinated individual should be considered if the clinical scenario suggests. A delay in diagnosis can be devastating and even lead to death. The purpose of this case report is to sensitize our fellow doctors to this uncommon disease which is still active in our world and may occur even in immunized people.

CONCLUSION

Always suspect tetanus in individuals with a triad of rigidity or stiffness, spasms, and dysautonomia, with or without a history of injury. Early treatment with immunoglobulins, surgical debridement, and high-dose antibiotics with anerobic coverage is essential for a good outcome.

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Funding: Nil; Conflicts of interest: Nil.

How to cite this article: Kaur DM, Tomar N, Rehani V, Anand R. A case of dystonic spasms in an adolescent: A neurologist's enigma! Indian J Case Reports. 2024; 10(3):75-76.