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Atypical Botulism Sparing Palsy of Extraocular Muscles

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Abstract
Recent studies of patients with botulism found ophthalmoplegia as a characteristic clinical sign. Here we illustrate a very rare case of atypical foodborne botulism with multiple bilateral cranial nerve palsies sparing palsy of extraocular muscles. Therefore, the classical diagnostic pentad of botulism (dry mouth, nausea, vomiting, dysphagia, diplopia, fixed dilated pupils) may be of limited sensitivity in single cases.

Key words: cranial nerve palsy, botulism, clostridium botulinum, diagnostic pentad

We report a very rare case of atypical foodborne botulism. Characteristically the symptoms are highly distinctive and include diplegia of the cranial nerves, followed by descending flaccid palsy, respiratory insufficiency and death. Most noteworthy, palsy of extraocular muscles is the hallmark of botulism due to axonal damage of the cranial nerves III, IV and VI, characterized by blurry vision and frank diplopia [1, 2]. Rapid diagnosis, application of botulinum antitoxin and meticulous monitoring of cardio-respiratory functions are mandatory [1, 3].

A 51 years old Turkish female was admitted with subacute dysarthria, dysphagia, nausea, emesis and headaches for 3 days, starting approximately 14 hours after ingestion of repetitive defrosted vacuum-packed beef. Neurological examination demonstrated diplegia of the cranial nerves V, VII, and IX to XII without any evidence of vegetative or motor contribution of the ocular nerves or descending palsy. Cranial MRI, repeated CSF studies and analysis of diphtheria toxin were unremarkable. Although repetitive facial nerve stimulation has a variable sensitivity and may not always be useful as a diagnostic test given the vagaries of test timing and severity of illness [7], electromyography was performed demonstrating no significant changes upon repetitive 30 Hz-stimulation and a slight decrement during 3 Hz. X-ray revealed a paralyzed right sided diaphragm according to a phrenic nerve lesion. Human pathologic neurotoxins of types A, B, E and F are produced by a diverse group of anaerobic spore-forming bacteria, including Clostridium botulinum groups I ands II, Clostridium butyricum, and Clostridium barantii. Till now the neurotoxin detection is based on the mouse lethality assay demonstrating typical botulism-specific symptoms (Fig. 1) [1]. Other sensitive and rapid in vitro assays have not yet been appropriately validated on clinical and food matrices [6]. After skin testing for serum-sickness-like-reactions trivalent botulinum antitoxin was administered to delay progression of symptoms [3]. Simultaneously penicillin therapy was started to eradicate a facultative Clostridium botulinum infection. Complete recovery of nearly all symptoms were observed 12 weeks after therapy. Besides foodborne botulism diplegia of the cranial nerves may also be triggered by diphtheria, myasthenia gravis, Lambert-Eaton-Syndrome and tick paralysis [1, 2]. These differential diagnoses could be excluded clinically, as well as by analysis of CSF and electrophysiological and microbiological findings [4].

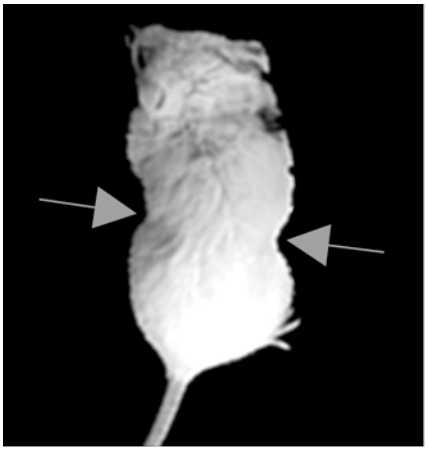

Fig. 1. Mouse bioassay with the typical picture of botulinum toxin induced "wasp-waist-phenomenon". For more information of typical findings in botulism see Lindström and Hannu (2006) [6].

In summary, our observations demonstrate an atypical presentation of botulism with isolated diplegia of the cranial nerves sparing palsy of the extraocular muscles. Recent studies of patients with botulism found ophthalmoplegia as the characteristic sign [5]. Therefore, in every case of isolated paralysis of multiple cranial nerves atypical manifestation of botulism should be considered. The classical diagnostic pentad of botulism (dry mouth, nausea, vomiting, dysphagia, diplopia, fixed dilated pupils) [3] even in combination with other typical symptoms like descending palsy, is

of limited sensitivity for diagnosing botulism and may prolongate diagnosis and therapy.

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