INTRODUCTION

Botulism is a rare but serious and potentially fatal illness. The botulism intoxication is caused by the exotoxin of Clostridium Botulinum which has a generalised effect on the neuromuscular junction involving both striated and smooth muscles. Botulinum toxin causes its major effect by blocking neuromuscular transmission in autonomic and motor nerve terminals. The types A, B and E account for human cases. Since the discovery of the toxin about 100 years ago, five clinical forms of botulism have been described: 1) classic or foodborne botulism; 2) wound botulism; 3) infant botulism; 4) hidden botulism; and 5) inadvertent botulism. A clinical pattern of descending weakness is characteristic of all five forms. Almost all human cases of botulism are caused by one of three serotypes (A, B, or E). Classic and wound botulism were the only two forms known until the last quarter of this century. Wound botulism was rare until the past decade. It is caused by local production of toxin by Clostridium botulinum after wound infection. Although it is a rare variant of botulism, it is increasingly being reported in drug users who inject subcutaneously. Infant botulism, first described in 1976, is now the most frequently reported form especially in the USA.
botulism is the most severe and debilitating form, caused by ingestion of the toxin. In our country many of the reported cases are food borne and mostly from home canned vegetables because preparing such food is seen quite often in rural areas of Turkey. Food-borne botulism may manifest as an outbreak but there are also single cases. Onset is within 12-36 hours after ingestion, signs of gastroenteritis with diarrhea, nausea and vomiting usually precede muscle weakness which often is generalised. Ocular and bulbar signs such as diplopia, ptosis, dysarthria and dysphagia are common. Weakness decends usually symmetrically to involve muscles of the trunk, respiratory system and limbs. Autonomic manifestations include dry mouth, dilated, fixed or poorly reactive pupils, blurred vision, constipation, ileus and urinary retention. Identification of the toxin in the patient’s serum confirms the diagnosis. Electrodiagnosis is mostly helpful. Nerve conduction studies show normal amplitude and latency of sensory action potentials. A small compound muscle action potential elicited by a single shock further declines with repetetive stimulation at a small rate. Repetetive stimulation at 20-50 Hz is the most specific test, showing an incremental response in most patients. Single fiber EMG has shown increased jitter and blocking and some reduction in fiber density. Specific therapy is the antitoxin therapy which should be administered early because it is unlikely to be effective after 3 days of exposure. In this condition only supportive therapy can be applied.

CASE PRESENTATION

A forty year old female patient who had nausea and vomiting besides blurred vision and generalised weakness of all four extremities was admitted to the emergency room. Her complaints had begun 2 days after she ate some of the home canned green beans she had prepared before. The day after the meal she had a fierce nausea and vomiting and two days later she complained of having blurred vision, diplopia, weakness and paresthesias in all extremities, difficulty of swallowing and breathing. She realised that she couldn’t shut her eyes properly and that her fascial muscles were weak. In neurological exam, she had bilateral ptosis, bilateral total external ophthalmplegia, bilateral mydriasis and fascial diplegia. She also had dysphonia and dysphagia. She had quadriparesis involving all extremities by 3/5. All deep tendon reflexes were abolished and she had generalised hypotonia. Nerve conduction studies of sensory nerves were in normal limits whereas M amplitudes were quite low in some of the motor nerves. Electromyographic studies showed that there was no voluntary motor activity in fascial muscles while there was a decline in the recruitment pattern of extremity muscles. In the repetetive stimulation of the trapezius muscle which was performed in the first week, all findings were in normal limits (Stimulations were performed with 2Hz, 3Hz and 20 Hz frequencies). No blink reflex was obtained in the first week. Regarding the story, the neurological signs and electrodiagnostic tests, a diagnosis of botulinum intoxication was made and as the clinical signs were progressing rapidly and in the descending fashion, antitoxin therapy was initialised and the patient was given A, B and E type antitoxin each given in 600 000 U/day, I.V and in slow infusion form.

Follow-up

A week after the antitoxin therapy, there were no changes in the neurological examination of the patient. The clinical improvement began gradually after this first week and in her first month’s check, her muscle strength was found to be +4/5, her deep tendon reflexes came back and she hadn’t ophthalmoplegia any more while light reflex was intact in both eyes. Her clinical improvement became more clear in the second month’s control and her neurological exam was found to be normal in the third month. Electrodiagnostic tests were repeated in the second week and in the third month. In the second week, the repetetive nerve stimulation test showed a mild decrement (10 Hz stimulation) while it was normal in the third month (3 Hz stimulation) (Fig.1-2 respectively). Blink reflex couldn’t be obtained in the first two weeks whereas in the third month its latency was in normal limits with a slight reduction in the amplitude (Fig.3-4 respectively).
DISCUSSION

Rapid diagnostic approach is very important in life threatening diseases and botulinum intoxication is one of these. The diagnosis is a clinical one, confirmed by electrodiagnostic tests and by testing stool for the organism, C. botulinum, or its toxin in serum and feces 13. Although serologic tests and electrodiagnosis are important tools in making the diagnosis, serology may be negative and electrodiagnostic results may not always be typical. The principal electrodiagnostic feature, an incremental response on high rates of repetitive nerve stimulation, has variable sensitivity and may not always be useful as a diagnostic test given the vagaries of test timing and severity of illness 8. Cox N. and Hinkle R. reported that a high index of suspicion is important for the diagnosis and prompt treatment of infant botulism, because this disease can quickly progress to respiratory failure 14. Unless SFEMG is used electrodiagnostic studies may not be helpful and diagnosis on clinical grounds is more essential than it is believed to be, especially in sporadic cases. Our patient happened to be one of these. Signs of internal and external ophthalmoplegia, dry mouth, descending paralysis, obstipation with weakness, absence of fever and lucid sensorium as cardinal symptoms should always bring botulism to mind 15. We thought in the same way in our diagnostic approach and made a clinical diagnosis firstly, the electrodiagnostic tests supported our clinical diagnosis partially (Findings of low amplitude CMAPs in nerve conduction studies and finding decremental response in the begining are helping but foundation of no incremental response in higher frequencies of stimulation is of no help). The frequency of diagnostic findings in cases of botulism intoxication seem to be quite different from each other in case reports published so far. Some of them claim that serology is more important while the others stress the importance of electrodiagnostic studies. In a presentation of a case of two boys with symptoms of food borne botulism, the diagnosis was made by serology (mouse neutralization test), whereas the EMG showed negative results 15. Graf WD. et al. suggested that electrodiagnosis was not a reliable tool as far as their 11-year review of toxin-confirmed cases had reflected 16. In a report of seven patients with foodborne botulism, the clinical picture was characterized by mild symptoms with a long latency of onset and by involvement of cranial and upper limb muscles; only one patient, a child, developed respiratory failure. Spores of Clostridium botulinum were found in stools in some but not all cases. Conventional neurophysiological tests had low sensitivity; abnormal findings were present only in the patient with severe clinical involvement, in whom compound muscle action potentials (CMAPs) appeared to be reduced. Repetitive nerve stimulation at a high rate showed pseudofacilitation and not true posttetanic facilitation, but single-fiber electromyography (SFEMG) showed abnormalities of neuromuscular transmission in every case 9. Clay SA. et al imply that the EMG pattern of brief, small motor unit potentials, in the context of the clinical syndrome may be diagnostic for acute infantile cases 17, almost alike our case. Graf WD. et al reported a
case where results of electrodiagnosis were negative but enema effluent contained adequate concentrations of organism and toxin to confirm the diagnosis. L. Mulleague et al. wrote that the diagnosis of botulism was based on clinical findings, but EMG remained the most useful discriminatory investigation. They also implied that the diagnosis should be confirmed by toxin bioassay, although occasional false-negatives can occur. Early diagnosis and associated therapy overcome the necessity of intubation and prolonged intensive care. Although the diagnostic approaches may show slight variaties it is certain that the important point is rapid diagnosis and therapy in botulinum intoxication. Our case confirms this very clearly. In this case report we also underline the importance of clinical and electrophysiological follow up in such patients thus the clinical diagnosis is confirmed and it is important supplying information of prognosis and patterns of healing. We performed a follow up of our patient up to 3 month’s time both clinically and electrodiagnostically. Here we performed a blink reflex study as well as the repetetive test. Although we hadn’t had an increment but a decrement pattern at the beginning, our follow up study showed a normal repetetive test finally and although the blink reflex couldn’t be obtained at the beginning, the follow up test showed a normal response. We also followed up a pattern of healing which quite overlapped the electrophysiological improvement. We emphasize the importance of clinical diagnosis and that making it rapidly is life saving.

REFERENCES