

Infant botulism mimicking an acute abdomen

Pseudo-addome acuto in un lattante affetto da botulismo infantile

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INTRODUCTION

Botulism is the acute, flaccid paralysis caused by a neurotoxin produced by *Clostridium botulinum* or, rarely, an equivalent neurotoxin produced by atypical strains of *Clostridium butyricum* and *Clostridium baratii*. In the infant, clinical symptoms are usually aspecific such as poor feeding, weak suck, feeble cry and drooling; neurologically it can manifest as a symmetric, descending, flaccid paralysis beginning with the cranial nerve musculature. We report a clinical case of botulism presenting initially with abdominal distention, therefore mimicking acute abdomen.

CASE REPORT

SA was a 3.5 month old male referred to us from a general hospital where he was diagnosed with an acute abdomen. He presented with clinical signs of irritability, poor feeding, stipsis and abdominal distention lasting two days. He was born after an uneventful pregnancy with vaginal delivery and Apgar 9 at 1 min, weighing 3,9 kg at birth. He was breastfed and showed normal growth and development before the onset of the disease. His parents reported giving him a pacifier with honey several times during the day. On admission the infant showed feeble cry, mild generalized hypotonia and poor attention. Vital signs were: RR 30/min, HR 126', O2Sat 98%, T 36.8°C, capillary refill 2-3 sec. and cold extremities. His weight was 5.6 kg. His pupils were mydriatic and had poor reactions to light. Abdominal distention and meteorism were present. Upon admission a biochemical lab test revealed hyponatraemia and hypochloraemia. A bolus of 100 ml of Normal Saline was started iv resulting in normalization of the altered exams.

On rectal examination faeces were found and a surgical emergency was eliminated. A complete work-up for sepsis was started and then the patient was put on ceftriaxone iv. Head CT scan and EEG were performed and no cerebral lesions were found. The meteorism and abdominal distention got worse during the following two days; a progressive flaccid paralysis became clinically evident thereafter. Vital signs worsened: respiratory rate was 60' and transcutaneous O2Sat was 90% on air. An arterial blood gas revealed a pCO2 of 56 mmHg. At this point infant botulism was considered in the differential diagnosis in view of worsening clinical condition and the negative sepsis work-up. The patient was intubated and ventilated, and 50 mg/kg of human botulism immune globulin (BIG-IV) was initiated. The stool and a sample of the honey given to the infant were examined by the Istituto Superiore di Sanità (ISS) to confirm the presence of *C. botulinum*. The following day the patient presented with fever and a chest X-ray was performed, revealing bilateral pneumonia. *Klebsiella pneumoniae* grew in the blood culture and the patient was given meropenem iv. Two weeks later the infant progressively improved and after three weeks the patient was extubated. *C. botulinum* was found in the patient's stool but not in the honey. The patient was discharged one month after admission.

DISCUSSION

Infant botulism is a rare disease due to the absorption of a toxin produced by *Clostridium botulinum*. In some children with unstable bowel flora, production of the toxin occurs in the large intestine as there is no other bacterial competition [1]. It typically affects babies under one year

of age. The only food it is associated with is honey to which, however, only 20% of cases can be linked. Infant botulism can present clinically in an acute or subacute form. The toxin produced by *C. botulinum* reduces the release of acetylcholine at the cholinergic synapses and neuromuscular junctions and the initial signs can be quite aspecific: poor feeding, weak suck, feeble crying, change in stool pattern with decreased frequency of bowel movements, onset of symptoms related to bulbar nerve palsies, such as feeble papillary response to light, lack of facial expression and difficulty in swallowing, can be followed by a cardio-respiratory arrest.

As soon as infant botulism is diagnosed intensive monitoring of the patient's nutrition and respiratory functions must occur. Botulism immunoglobulins (BabyBIG) have to be given as they are the only treatment that can eliminate the toxemia associated with infant botulism [1]. As botulism is a life-threatening condition the diagnosis must be prompt: early treatment with BabyBIG within 3 days after hospital admission decreases the length of the hospital stay from 5.7 weeks to 2.6 weeks [2]. Our patient presented stipsis, abdominal distention and meteorism, irritability and poor feeding, preceding the weakness of the bulbar palsies.

In the differential diagnosis we included either acute abdomen or a sepsis or a cerebral event but the presence of mydriasis and feeble pupillary reactions should have soon suggested the possibility of a *Clostridium* infection. As the patient worsened despite the antibiotics, we reconsidered the diagnosis of infant botulism, in view of the presence of mydriasis and feeble pupillary signs. Stool examination confirmed our diagnosis. During the period from 1984 to 2006 only 26 cases of infant botulism were reported in Italy [3]. This is a very small number if compared with the 80-100 cases reported annually in USA [1].

This different occurrence may reflect different environmental distributions of *C. botulinum*, different cultural practices in the feeding of infants with honey, different levels of exposure to dust or soil, and different susceptibility of patients attributable to other presently unknown factors. On the other hand, since the initial symptoms of the disease are often similar to those of several other diseases, differential diagnosis is very difficult and rarely suspected by the physician [4]. Since 1992, approximately in 5% of the 681 patients in the USA who were started with BabyBIG on suspected clinical diagnosis, the disease was not confirmed [5]. Other diagnoses to consider with infant botulism are SMA type 1, sudden infant death and metabolic disorders [6]. Since most paediatricians are unfamiliar with the clinical manifestations of infant botulism, diagnosis can be easily missed and the disease is possibly underestimated and underreported. It is important that clinical history and neurological examination include infant botulism in the differential diagnosis. An altered function of several cranial nerves should direct the physician to suspect this infection. An altered pupillary light reflex may be easily missed. Hence careful, prolonged examination is required, as repetitive muscle activity reveals the reduced availability of acetylcholine [4]. The presence of multiple cranial nerve palsies is important to identify infant botulism, and in our case feeble cry, poor suck, weak gag, difficult swallowing (drooling), and a face with no expression were all indicative of an early bulbar palsy. The use of antibiotics in such patients should be avoided as it can cause the lysis of intestinal clostridia, with a massive release of toxin that may worsen the clinical conditions of the patient.

Key words: infant botulism, acute abdomen, ocular sign, botulism incidence.

SUMMARY

Botulism is the acute, flaccid paralysis caused by a neurotoxin produced by *Clostridium botulinum*. In the infant, clinical symptoms are usually unspecific such as poor feeding, weak suck, feeble cry, drooling, followed by a symmetric, descending, flaccid paralysis beginning with the cranial nerve musculature. The initial symptoms of the disease are often similar to several diseases and therefore differential diagnosis is very difficult and rarely

suspected by the physician. Since 2004 only 22 cases of infant botulism have been reported in Italy. Since most paediatricians are unfamiliar with the clinical manifestations of infant botulism, the diagnosis can be easily missed. Hence the disease may well be underestimated and underreported. We report a clinical case of botulism presenting initially with abdominal distention, thereby mimicking acute abdomen.

RIASSUNTO

In Italia, il botulismo infantile è una malattia rara. I dati dell'Istituto Superiore di Sanità di Roma riferiscono solo 22 casi negli ultimi 20 anni. Negli Stati Uniti, l'incidenza è di gran lunga maggiore, un dato che lascia ritenere che forse nel nostro Paese non tutti i casi vengano diagnosticati. Il botulismo "non diagnosticato" rientra probabilmente nel numero di casi di morte improvvisa (in culla) del lattante. La diagnosi di questa patolo-

gia nel lattante non è semplice in quanto, come nel caso da noi descritto, la sintomatologia può essere subdola e del tutto aspecifica e l'esordio può mimare un quadro clinico di addome acuto. L'interesse del nostro caso scaturisce inoltre dal fatto che esso ha presentato tutti i sintomi e le complicanze possibili riportati in letteratura, risultando quindi estremamente esemplare oltre che di estrema gravità.

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