Corrosive poisoning mimicking cicatricial pemphigoid: Munchausen by proxy

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Abstract

**Background** Munchausen by proxy (MBP) is a severe form of abuse in which a caregiver simulates or fabricates illness in another person, primarily the elderly and children, which can even result in death.

**Case** We report two siblings who were victims of MBP, one of whom died. A very rare diagnosis, cicatricial pemphigoid was suspected in the 2-year-old girl who was first abused. She was hospitalized twice, for 3 and 4 months, respectively. Her second hospitalization ended with her death. MBP was diagnosed after the second sibling’s admission with similar atypical signs and symptoms. It was realized that a household-cleaning product, sodium hydroxide, was administered repeatedly by oral route to the children by their own mother.

**Conclusions** Physicians must consider the diagnosis of MBP whenever they are confronted with unusual, persistent or recurrent signs and symptoms in a child.

Introduction

Munchausen by proxy (MBP) is a severe form of abuse in which a caregiver simulates or fabricates illness in another person, primarily the elderly and children. This abuse can result in death in severe cases (Fulton 2000). The most common methods of fabricating illness are lying, poisoning either with drugs or other substances, suffocation, specimen tampering and chart falsification (Venneman et al. 2005). We report two siblings who were victims of MBP, one of whom died as a result of administration of an alkaline corrosive. We report these cases, because the victims were poisoned with an unusual agent, a strong corrosive household-cleaning product, which caused atypical signs and symptoms and death of the first victim.

Case report 1

H.G., a 2-year-old girl, was referred to the Division of Pediatric Allergy for persistent conjunctivitis, which was initially diagnosed as Stevens–Johnson syndrome sequela secondary to ampicillin. Her detailed history revealed that she was hospitalized for 2 months with the diagnosis of Stevens–Johnson syndrome and was discharged and followed up as an outpatient. Her birth history was unremarkable. Her grandfather and brother had histories of penicillin allergy. Her mother was a housewife; her father was fabricating linen posters with his mother and brother on the first floor of the house, where they lived.

On admission to the hospital, the child had conjunctival adhesion of the distal portion of the right eyelids due to scarring, purulent discharge and cicatization under the right eyelid, gingivitis and erosions in the oral cavity. There was no skin involvement. The mother told that the preservative-free artificial eye-drop prescribed by the ophthalmologist caused scarring under the right eyelid as soon as she applied it. After consultation with ophthalmology and dermatology clinics, a preliminary diagnosis of recurrent Stevens–Johnson syndrome was made, and various agents, including oral antihistamine and
cyclosporin, were prescribed. But the child experienced unexpected reactions, such as mild stridor, flushing, mucosal bleeding in the oral cavity and small black discolorations on the frontal surface of the chin, immediately after the mother administered these drugs. Reactions also occurred immediately after some foods were given by the mother, which the child had tolerated before. Multiple drug and food allergies were considered in the patient. All medications were stopped. She was fed with hydrolysed formula by nasogastric tube to lessen the risk of any hypersensitivity reactions. Laboratory investigations including serum biochemistry, quantitative immunoglobulin levels, complements and lymphocyte subsets were within normal levels. Viral and bacterial serological tests, autoantibodies, serum-specific IgE levels for cow's milk and egg, skin prick tests for common allergens, and skin patch tests for Behçet's disease were unremarkable. After consultation with periodontologists, severe gingivostomatitis was dressed with serum saline only. Oral lesions were gradually healed with scarring. Adding each food item gradually to the diet was planned to prevent possible adverse food reactions. The patient's general status gradually improved and she was discharged. During her hospitalization, she had three fever episodes lasting 2–3 days with high acute phase reactants, without any bacterial growth in blood cultures.

After 4 months of a symptom-free interval, the patient was readmitted to the hospital with fever, vomiting and purulent discharge from the old healed perioral lesions and mucosal bleeding from the mouth. After admission, she developed respiratory distress and inspiratory stridor. She subsequently required a tracheotomy. Bronchoscopic examination revealed white-coloured plaques at the mucosal surface of the trachea. After consultation with the dermatology clinic, multiple sharply bordered superficial ulcers on the tongue, scarring on the lower lip with synthesis and syllephant on the right eye were found to be compatible with the diagnosis of cicatricial pemphigoid. Although histopathological evaluation of the lesions demonstrated non-specific findings, indirect immunofluorescence investigation was found to be positive for laminin 5, which supported the presumptive diagnosis.

After the tracheotomy, the child did not tolerate oral feeding, and was fed with nasogastric tube for nearly 1 month. A second bronchoscopic examination revealed tracheal stenosis at the proximal site. During this second hospitalization, which lasted 4 months, the patient had intermittent inspiratory stridor and dyspneic attacks, with exacerbation of mouth lesions and fever. The patient was treated with antibiotics, bronchodilators and intravenous immunoglobulin. Systemic steroid was given for the diagnosis of cicatricial pemphigoid. The patient died following massive bleeding from her mouth, which occurred after an acute episode of inspiratory stridor and dyspnea. Autopsy was offered to the family, but the family did not accept post-mortem examination.

Case report 2

Three months after the first sibling's death, F.G., the 6-year-old brother of H.G., had an episode of purplish discoloration and swelling in the lips, oral aphthous ulcers lasting 2 weeks. At that time, the child was examined by a physician in an outpatient clinic and was advised topical treatment for oral ulcers. His birth history was unremarkable. He had penicillin allergy and was hospitalized for an anaphylactic reaction to penicillin 2 years before. Two weeks after this episode, he was admitted to the hospital for further investigation. On admission, physical examination revealed aphthous ulcers and scars due to old healed lesions on the lips and tongue. Eye examination was normal. Biopsy specimen taken from the oral lesions revealed extended erosions, regeneration in the adjacent tissues, inflammatory response in the subepithelial tissue and fibrosis. Direct immunofluorescence investigation demonstrated IgM and C3 deposition in the perivascular tissue. Serum biochemistry, whole blood cell count, urine analysis and acute phase reactants were within normal levels. Immunological and serological tests were planned to exclude autoimmune diseases. The child's oral lesions healed gradually within 10 days and the patient's discharge was planned. However, the patient had a sudden episode of oedema in the lips, generalized maculopapular rash after eating his routine breakfast, which included cow's milk and egg. Physical examination revealed erosion in mucosal membranes and mucosal bleeding foci in gingiva. The following day, the child had a similar reaction in the mouth after gargling with a mouthwash prescribed previously by a dermatology clinic. Although it was not ordered at that time by anybody in our paediatric team, the mother asserted that the medical staff advised her to give the mouthwash again. The child was agitated and complained about a burning feeling in his throat and chest. His lips and tongue were erythematous and oedematous. The room was inspected in the absence of the mother and the child. A household-cleaning product containing sodium hydroxide was found hidden in the reservoir of the bathroom. Endoscopic examination revealed burns in the throat and oesophagus of the child. The hospital's child protection team and legal authorities were informed, and the child was separated from the mother; the lesions gradually healed and did not reappear.
Discussion

Munchausen by proxy was first described in 1977 by Professor Roy Meadow (Meadow 1977). Since that original report, numerous case reports have been published with a wide range of clinical spectrum. Although in the mild cases mothers only report false symptoms, in the severe cases mothers can cause severe physical harm to their children, even resulting in the death of their children, in the continued pursuit of making their children appear ill (Forsyth 2000). MBP is difficult to characterize; any symptom can occur in the course, and it can mimic numerous diseases depending on the agent utilized to poison the child. The correct diagnosis is often delayed and there are undoubtedly more cases of MBP than reported (Galvin et al. 2005). MBP is typically not diagnosed unless there is a high clinical index of suspicion. There are numerous reasons for the delay in the correct diagnosis. First, hospital staff trust the child’s mother, as nobody thinks that a mother would give harm to her child. On first impression, the majority of perpetrators appear as devoted mothers who will not leave the child’s bedside for even short periods. They easily gain admiration of the staff. This received attention and admiration feed their psychological needs, leading to a vicious circle. Second, physicians do intensive work-up for rare diseases with genuine desire to help the child, which may lead away from the real diagnosis and facilitate the perpetrator's manipulations (Hettler 2002). Third, prolonged and multiple hospitalizations can cause complications such as nosocomial infections, which can further confuse the medical staff.

Typical warning signs of MBP are presented in Table 1 (Meadow 1982; Jones et al. 1986; Forsyth 2000; Fulton 2000; Zylstra et al. 2000; Hettler 2002; Schreier 2002; Galvin et al. 2005; Venneman et al. 2005).

Separating the child from the perpetrator for a sufficient length of time can be diagnostic with the discontinuation of the symptoms and rapid healing. As cases of reabuse in siblings have been reported, siblings and future children should be followed closely by medical personnel.

In our paediatric department, mothers are always kept with their hospitalized children to prevent emotional-separation stress in children. The mother in this case appeared to be a devoted mother; she was very helpful in the care of her first child. She gave all oral drugs and foods and applied all oral dressings to her two children during hospital stays. The children’s injuries occurred either at home or in the hospital when they were together with their mother. As we stuck to the diagnosis of cicatricial pemphigoid, a rare chronic autoimmune disease primarily involving the mucous membranes including oral cavity and the eyes with essential lesions of bulla or erosions, we omitted the possibility of MBP in the first child. When we re-evaluated the unusual presentations of the two siblings and their mother’s behaviours from a different point of view, we had a high index of suspicion of MBP, which we demonstrated by evidence in the last attempt of the mother.

In conclusion, physicians must consider the diagnosis of MBP while searching for a rare disease whenever they are confronted with unusual and persistent or recurrent signs and symptoms in a child.

Acknowledgement

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Table 1. Signs of Munchausen by proxy

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<thead>
<tr>
<th>Relevant to the child’s disease</th>
<th>Relevant to the characteristics of perpetrators</th>
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<tr>
<td>• Younger age (particularly infants, children less than 3 years)</td>
<td>• A mother who welcomes even painful medical tests for her child, is constantly at bedside, yet seems less concerned than medical staff about the health of her child</td>
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<td>• Persistent or recurrent illness that cannot be explained</td>
<td>• Having a history of training or involvement in some aspects of health care</td>
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<td>• Unusual symptoms and medical course – ‘never seen a case like before’</td>
<td>• Having good relationship with medical staff</td>
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<td>• Medical problems not responding as expected to therapy</td>
<td>• Dressing child in inappropriately lavish clothing, or excessive number of toys filling the hospital room</td>
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<td>• Symptoms that occur only when the mother (or suspected perpetrator) is present</td>
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<td>• Unexplained illness in the siblings while under the supervision of the mother</td>
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<td>• History of sudden and unexplained sibling death</td>
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References


