

Shanghai Fever: A Fatal Form of *Pseudomonas Aeruginosa* Enteric Disease

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Background: Outcome of pseudomonas enteric fever is unpredictable as multiple systemic lethal complications occur abruptly. **Case characteristics:** A 9-month-old girl with multiple ileal perforations, leukocoria, ecthyma gangrenosum, hemiplegia and a perforated ulcer in the soft palate. Blood culture suggested *Pseudomonas aeruginosa* infection. Operative repair of multiple ileal perforations and multidisciplinary management was provided. **Outcome:** On 10th post-operative day, patient succumbed to multiple organ dysfunction syndrome. **Message:** Early detection and management of complications of *P. aeruginosa* enteric disease is important.

Keywords: *Enteric perforation, Granuloma, Pseudomonas, Vasculitis.*

The prognosis of enteric perforation has improved dramatically with advanced anesthesia and post-operative intensive care unit (ICU) support. However, this is not true for pseudomonas enteric disease, as atypical multi-systemic complications are more pronounced. This condition is highly fatal due to rapid onset of septic shock and multiple organ dysfunction syndrome (MODS). One such distinct and fulminant form of pseudomonas enteric disease entitled as “Shanghai fever” was described early in 1918 [1]. This condition is almost indistinguishable from vasculopathy. Thus, a multispecialty approach and awareness of the entity are imperative for timely diagnosis and management of such cases.

CASE REPORT

A 9-month-old female child was admitted with high fever for three days and painful swelling of left eye associated with increased lacrimation. The fever was continuous, not associated with rigors, and relieved with medications. Left eye condition was sudden in onset, rapidly progressing, associated with severe pain, redness and raised local temperature. The child was irritable since beginning of the fever. She had no history of vomiting, loose stool, burning micturition, trauma to the eye, abscess elsewhere in the body, convulsion, altered consciousness and similar illness in the past. Plain X-ray abdomen showed free gas under diaphragm and an emergency exploration was performed. Multiple ileal perforations (15cm, 22cm, 26cm, 32cm and 40cm from the ileo-caecal junction) were detected. There was no obvious evidence of mesenteric arterial ischemia. All the

perforations were repaired in two layers with Vicryl 4/0. A piece of tissue from the perforated margin was sent for biopsy.

The child was shifted to Pediatric Intensive Care Unit due to delayed recovery from anesthesia and decerebrate posturing. The patient also developed right-sided hemiplegia. A non-contrast computed tomography (NCCT) of brain showed acute infarcts involving almost entire left anterior and middle cerebral artery territories with mass effect (**Fig.1**). On 4th post-operative day, we noticed multiple skin lesion in the right hand



FIG. 1 Non-contrast computed tomography of brain showing acute infarcts with mass effect, uncal and subfalcine herniation involving almost entire anterior and middle cerebral artery territories.

with delayed capillary filling in the index and middle fingers. The color doppler study was inconclusive. Anti-neutrophil cytoplasmic antibody (ANCA) was negative and the skin biopsy was inconclusive. The histopathological reports of perforated margin showed nonspecific granulomatous lesion. The patient was put on injectable Amikacin along with Imipenem as the blood culture showed *P. aeruginosa* sensitive to amikacin. We started tube feeding on 6th post-operative day. The patient developed a perforated ulcer on the soft palate in post-operative period (**Fig. 2**). The child initially responded well to treatment, but died due to multiple organ failure.

DISCUSSION

Shanghai fever comprises a triad of symptoms: fever (100%), diarrhea (96%) and sepsis (81%) [2]. Apart from this triad, bowel perforation due to widespread patchy necrotising intestinal lesions, ecthyma gangrenosum, meningitis, acute otitis media, leukocoria of the eye, and facial paralysis are reported [3]. The disease is rare but often associated with multiple organ system complications with high mortality rate (23%-89%).

The young age, neutropenia and hypogammaglobulinemia play important role in the pathogenesis of Shanghai fever [4]. Early administration of anti-pseudomonal antibiotics and intensive care unit management are important for favourable outcome. Some extra-intestinal manifestations of typhoid fever practically mirror that of the *P.aeruginosa* enteric fever [5]. The reported complications of typhoid fever do not

include necrotizing skin lesions, aphthous ulcer or leukocoria [6].

The similar skin lesion, acute abdomen, central nervous system affection and multiple internal organs involvement are found in Degos disease. Degos' disease is a bizarre vasculitis disorder [7]. In this condition, the multiple infarctive and thrombotic lesions are often misdiagnosed as vasculitis. The high mortality is often related to intestinal perforation or cerebral infarction. Albeit, intestinal perforation and brain infarction are scarcely found in Henoch-Schonlein purpura and Churg-Strauss syndrome (CSS), where vasculitis is the central pathologic concern [8]. CSS is caused by diffuse allergic vasculitis involving small and medium sized arteries and an important differential. Multiple organs, lungs, skin, peripheral nerve, lymph nodes, intestine etc., are mostly affected in the third stage of the disease course.

ANCA, particularly perinuclear (P-ANCA), are found in 30-40% cases of CSS. Peripheral blood eosinophilia [greater than $1.5 \times 10^9/L$] and the characteristic pathological features (eosinophilic pneumonia, granulomatous inflammation and necrotizing vasculitis) are essential for the diagnosis. However, the skin biopsy in our case was not suggestive of vasculitis disorder [9].

There are some heterogeneous groups of diseases where various systemic components are invariably associated with the granulomatous intestinal lesions as found in our case. The corresponding extra-intestinal manifestations might be present in inflammatory bowel disease (IBD), abdominal tuberculosis, histoplasmosis, sarcoidosis, lymphoma, eosinophilic gastroenteritis (EG), and systemic mastocytosis. Thus, it is important to be aware of the representative features of pseudomonas induced Shanghai fever and its complications. Besides, we must have a high index of suspicion and low threshold level for early and timely diagnosis and proper therapeutic approach for the management of septic shock and MODS.

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FIG. 2 Oral cavity showing perforated oral ulcer in the soft palate.

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