Necrotic Lesions in Infants: Ear, Nose, and Throat Manifestations

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Abstract

Aim: To describe a unique finding of necrotic lesions of the peripheral aspect of the body affecting the nose and ears in sporadic cases admitted in the pediatric ward at our center.

Background: Infants till 4 months are immune to viral infections. However, a severe viral infection may lead to rapid deterioration with secondary bacteremia. Pseudomonas infection is frequently complicated by necrotic lesions of the periphery, namely, ecthyma gangrenosum (EG). This case series includes five cases which were sporadic coming from suburban areas of two districts of Madhya Pradesh.

Case description: All five cases of the age group between 1 month and 10 months had presented with a short history of high-grade fever of 4–6 days, followed by the appearance of rashes. Rashes were maculopapular involving thighs and buttocks with some cases involving the nose and ears. The patients were admitted to the pediatric ward and a complete hemogram, blood sugar, urine analysis, cerebrospinal fluid (CSF) culture, pus culture, antineutrophil cytoplasmic antibodies (C-ANCA) test, and biopsy were done. Antibiotic coverage was immediately started and response was observed.

Conclusion: The skin lesions affecting infants associated with fever and rash are very typical in presentation. In this series, the necrotic lesions are peculiar with the involvement of the nose and ears along with the peripheral region. These cases were primarily diagnosed by presentation and confirmed by biopsy and other investigations. Three cases improved while two patients died. Such cases were not seen prior to the study period nor seen later. Ecthyma gangrenosum was concluded to be the final diagnosis comparable to the clinical picture in most cases both with and without bacteremia.

Clinical significance: Infantile febrile illness with skin lesions are a rare clinical entity that should be reported early. They are primarily diagnosed by clinical presentation. If managed timely, serious complications can be avoided.

Keywords: Case report, Ecthyma gangrenosum, Infantile febrile illness, Pseudomonas aeruginosa.

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Background

Acute febrile illness with skin lesions in infancy is a serious medical condition. Skin lesions affecting extremities and limbs sparing the trunk can be seen in some clinical conditions like ecthyma gangrenosum, and toxic epidermal necrolysis syndrome. This series aims to evaluate the cases of nasal tip necrosis with necrosis of the pinna along with necrosis of buttocks and fingers in infants below 1 year of age that were reported in Netaji Subhash Chandra Bose Medical College from October 2012 to December 2012. These cases came sporadically and we have not seen such cases at our center neither in the preceding years nor the successive years.

Case Description

Total five cases are included in this series. These infants were of age group between 1 month and 10 months. Four infants were male and 1 was female. Common symptom for the presentation was high grade fever (101–103°F) of the short duration of 4–6 days with a cold/cough. After 2–3 days of fever, infants started developing rashes and dark avascular lesions over the periphery of the body. Patients had a toxic appearance with labored breathing along with rash at the time of presentation. The distribution of rashes typically involves an anterior aspect of the thighs and buttocks sparing the trunk. Rashes were maculopapular and reddish in appearance. Initially, they were black patches that sloughed out. One patient had gangrenous fingertips and toes, 1 had nasal tip destruction and 1 had discoloration and ulceration over the bilateral pinna (Figs 1 and 2).

The ulcer had sloping edges with active pus discharge. These ulcers were bone-deep with indurations and redness. None of the cases had seizures or any neck stiffness. Only 1 case had a vacant stare with uprolling of eyeballs. There was no apparent history of any previous drug reaction or any drug administration. Hemoglobin levels were low in all the cases, cerebrospinal fluid (CSF) culture
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Table 1

<table>
<thead>
<tr>
<th>Case</th>
<th>Histopathological Evaluation</th>
<th>Clinical Picture</th>
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</thead>
<tbody>
<tr>
<td>1</td>
<td>Nasal necrosis</td>
<td>Gangrenous lesion</td>
</tr>
<tr>
<td>2</td>
<td>Gastrointestinal lesion</td>
<td>Necrotic lesion</td>
</tr>
<tr>
<td>3</td>
<td>Nasal cartilage necrosis</td>
<td>Nonspecific inflam</td>
</tr>
</tbody>
</table>

Fig. 1: Gangrenous lesion over the tip of the nose – 4-day admission

Fig. 2: Patient with nasal destruction and rashes with an ulcer over cheek and main trunk on day 10

was sterile, total leucocyte count (TLC): 12,000–18,000 with primary neutrophilia, erythrocyte sedimentation rate (ESR): 40–50, blood culture came out to be positive for Pseudomonas aeruginosa in four cases and sterile in one case (Table 1). β-Lactam antibiotics (ceftazidime) with antipseudomonal coverage were started. Systemic corticosteroids (dexamethasone) were given for 7 days along with nebulization. Three patients responded well but two succumbed to death. Regular dressing of the wound with surgical debridement was done.

Discussion

The differential diagnosis kept in mind for these infants include toxic epidermal necrolysis syndrome, pyoderma gangrenosum [ecthyma gangrenosum (EG)], erythema multiforme and staphylococcal scalded skin syndrome (SSSS). No previous history of drug reaction is very helpful in ruling out toxic epidermal necrolysis syndrome since it is primarily a drug-related condition or a graft versus host disease reaction. Differential diagnosis may also include infantile edema with gangrene having autoimmune lymphangitis leading to necrotic lesions of nose and other parts of it which require immunofluorescent studies.

Ecthyma gangrenosum was initially reported by Hitchsman and Kreilich in 1897. It is a necrotic skin lesion caused by P. aeruginosa usually in immunocompromised patients. The skin lesions are of four types as follows: (A) Ecthyma gangrenosum with vesiculate lesion and hemorrhagic cellulitis; (B) in gastrointestinal lesion; (C) in pediatric patient; and (D) infant with pseudomonas infection along with necrosis of upper and lower nasal cartilage. However, literature has supported that it can develop even in immunocompetent patients without bacteremia. When associated with systemic immunocompetency, it is life threatening. Various case reports have been reported with infants with diarrhea and necrotic lesions over the nose in which culture was positive for pseudomonas, but the patient died in 7–10 days. It is a rare disease and the incidence of this disease is uncertain. It is estimated to be affecting 3–10 patients per million per year.

Age of the patients ranged from 1 month to 10 months. This is in correlation with the infantile predilection of these diseases. The patients belonged to suburban areas of two districts of Madhya Pradesh, namely, Jabalpur and Sagar.

All patients were from suburban areas and male–female ratio was 4:1. All patients were born by full-term normal vaginal delivery. Thus, preterm infancy, and illness did not contribute to any of the symptoms.

The duration of the illness was short of around 4–6 days with refusal to feed, vomiting, up rolling of eyes with a vacant stare. Red patches over the body were present in all the cases within 2–3 days of the onset of fever. Four cases had bilateral presentation of the disease while one case had unilateral spread. Pinna was involved bilaterally with necrosis and discoloration of the helical and antihelical curves, but no evidence of diffuse perichondritis (Fig. 3).

All patients were admitted with a preliminary diagnosis of acute viral fever with meningococcemia with septicemia and purpura fulminans with cutaneous vasculitis with one case of aspiration pneumonia. Japanese encephalitis (JE) and dengue fever were considered initial differential diagnoses as per the fever pattern, but on testing for IgM for both, it came to be negative.

Blood culture came out to be positive for P. aeruginosa in four cases and sterile in one case. The clinical picture resembled that of EG. It should be considered as a possible diagnosis even in the absence of bacteremia or any positive biopsy.

All cases had low hemoglobin levels, with TLC, 12,000–14,000, blood sugar, 60–74 gm/dL and urine showed traces of albumin in all cases. Gram stain and culture were negative. Patients underwent tests for dengue and for JE that came out to be negative. C-ANCA was done for 1 case but that also came to be negative; acid-fast bacteria (AFB) was also negative; one patient on computed tomography (CT) showed mild ventriculomegaly, this patient succumbed to death on 4th day of admission. The rest patient on CT was within normal limits. The patients had extensive dry black gangrene at buttocks of 4–5 cm which on debridement involved muscle and after debridement bone was exposed (Fig. 4).

Histopathological evaluation of nasal necrotic debris was done for only one patient and that was reported as acute on chronic nonspecific inflammation with necrosed stratified lesion with subdermal edema with mononuclear cell infiltrates and dense inflammation in subepithelial tissue.

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### Table 1: Overview of all the cases

<table>
<thead>
<tr>
<th>S. No.</th>
<th>Age/sex</th>
<th>Laterality</th>
<th>Symptom</th>
<th>Signs</th>
<th>Investigations</th>
<th>Diagnosis</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>5 m/M</td>
<td>Bilateral</td>
<td>Fever, cough, labored breathing for 4 days</td>
<td>Black patches over buttocks and thigh, necrotic debris in the nasal cavity with the destruction of cartilaginous nasal septum, gangrenous patch over bilateral pinna</td>
<td>Hb: 6 gm/dL&lt;br&gt;ESR: 35 mm&lt;br&gt;TLC: 14,000&lt;br&gt;P72L24E2M2 Culture: <em>Escherichia coli</em> (pus from nose)&lt;br&gt;<em>P. aeruginosa</em> (blood)&lt;br&gt;Urine: Traces of albumin&lt;br&gt;Budding test + Biopsy: Acute on chronic nonspecific inflammation with necrosis</td>
<td>EG with purpura fulminans with cutaneous vasculitis</td>
<td>Antibiotics with wound debridement</td>
</tr>
<tr>
<td>2</td>
<td>5 m/M</td>
<td>Unilateral</td>
<td>Cough × 4 days; Fever and rash × 2 days; Single episode of vacant stare</td>
<td>Ulcer over elbow and anterior aspect of right thigh, ulcer over the right cheek</td>
<td>Hb: 6 gm&lt;br&gt;TLC: 18,000&lt;br&gt;P64L29M7 Urine: Microscopy, 5–8 cells/mm³&lt;br&gt;Pus culture: Sterile&lt;br&gt;Gram stain/AFB: C-ANCA&lt;br&gt;Blood group: O+ Blood culture: <em>P. aeruginosa</em> Chest X-ray: Infiltration with prominent bronchogram</td>
<td>EG with aspiration pneumonitis</td>
<td>Died</td>
</tr>
<tr>
<td>3</td>
<td>10 m/F</td>
<td>Bilateral</td>
<td>Fever with active cold × 6 days; Rashes over body × 3 days; Decreased appetite × 4 days</td>
<td>A rash over bilateral pinna; Healing ulcer over buttock and anterosuperior thigh; A maculopapular rash over lower limbs</td>
<td>Hb: 7.2&lt;br&gt;TLC: 14,300&lt;br&gt;P74L24M2 Urine: Pus cell, 6–8 cells/mm³&lt;br&gt;Epithelial cells: 4–6&lt;br&gt;Traces of albumin&lt;br&gt;Blood group: A+ Blood culture: <em>E. Coli</em>, enterococci Pus culture sterile, Dengue/JE IgM Elisa negative</td>
<td>Meningococcemia with septicemia with cutaneous vasculitis</td>
<td>Antibiotics with wound debridement</td>
</tr>
<tr>
<td>4</td>
<td>5 m/M</td>
<td>Bilateral</td>
<td>Fever and cold cough for 8 days; Swelling over body × 3 days</td>
<td>Discoloration skin over nasal tip and philtrum; Ulcer over forearm, bilateral buttocks, anteromedial thigh; Bilateral toes gangrenous</td>
<td>Hb: 7 gm/dL&lt;br&gt;TLC: 20,000&lt;br&gt;ESR: 65 mm&lt;br&gt;Blood culture: <em>P. aeruginosa</em> CSF: Sterile</td>
<td>Viral hemorrhagic fever with septicemia with pyoderma gangrenosum</td>
<td>Died</td>
</tr>
<tr>
<td>5</td>
<td>1 m/M</td>
<td>Bilateral</td>
<td>Fever for 4 days; Vomiting: two episodes; Rashes all over body × 3 days</td>
<td>Multiple dark-colored patches 4–5 cm over both cheeks, buttocks along with healing ulcer over the nose and upper lips</td>
<td>Hb: 10 gm&lt;br&gt;TLC: 12,500&lt;br&gt;N60L36MO2&lt;br&gt;RBS: 60 Pus culture: CSF: Sterile Blood culture: <em>P. aeruginosa</em></td>
<td>EG</td>
<td>Antibiotics with wound debridement</td>
</tr>
</tbody>
</table>

EG, ecthyma gangrenosum; Hb, hemoglobin; JE, Japanese encephalitis; IgM, immunoglobulin M
The treatment protocol followed was primarily conservative. Patients were kept in the intensive care unit with close monitoring of vitals. β-Lactam antibiotics combined with antipseudomonal antibiotics and systemic corticosteroids (dexamethasone) were started with proper hydration and nebulization. Systemic steroids have been found to be effective in the acute form of the disease. Wound debridement with proper dressing was carried out for the ulcerated site. Three patients improved within 7–10 days but two patients had an aggressive course of the disease and did not survive. At the time of discharge, patients were strictly advised for regular follow-up for plastic reconstruction of the necrosed ulcer site for future skin grafting. Only one patient came for follow-up after 1 month and was referred to plastic surgery for skin grafting.

**CONCLUSION**

This case series highlights the rare presentation of infantile febrile cutaneous infections with otorhinolaryngologic involvement presenting in suburban populations that presented at our center. These sporadic cases were not reported prior to or later to this period of study. Ecthyma gangrenosum is a necrotic infection that presents in infancy with and even without bacteremia. Preceding upper respiratory infection (URI) and fever with rash are dangerous signs that should be aware of parents to seek earlier medical attention. These necrolytic infections can be managed conservatively but close monitoring and earlier intervention is quite important.

**Clinical Significance**

This series is an attempt to report rare cases of necrotic infectious diseases in infants that have otorhinolaryngologic manifestations. Infants are very prone to viral febrile illness complicated by bacterial infections. Timely management can prevent any grave complications.

**Ethical Approval**

The authors hereby declare that this study has been conducted under all ethical standards and that no humans or animals have been experimented with or tortured during the study period.

**REFERENCES**