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Case Report

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Ecthyma gangrenosum in a previously healthy pediatric patient and associated facial paralysis and persistent hyperplastic primary vitreous

Ibrahim Hakan Bucak[1], Gökhan Tümgör[2], Eda Mengen[2], Fatih Temiz[3], Mehmet Turgut[1]

[1] Department of Pediatrics, Faculty of Medicine, Adýyaman University, Adýyaman, Turkey
[2] Department of Pediatrics, Faculty of Medicine, Cukurova University, Adana, Turkey
[3] Department of Pediatrics, Adana Numune Training and Research Hospital, Adana, Turkey

Summary

Background: Ecthyma gangrenosum is an infective lesion of the skin and mucosal membranes. It is most commonly caused by *Pseudomonas aeruginosa,* and the most important risk factors are malignancy and neutropenia. However, it has rarely been reported in children who were previously healthy. Persistent hyperplastic primary vitreous has been described as the persistence of the fetal hyaloid vascular system. Acute otitis media with facial paralysis is an infrequent association.

Case Report: We report the case of a 5-month-old boy hospitalized because of fever, otorrhea and necrosis on his body. He had peripheral facial paralysis on the same side as otorrhea. Leukocoria was determined in the right eye. He had many gangrenous ulcers on the extremities and body.

Conclusions: We present a previously healthy pediatric patient diagnosed with persistent hyperplastic primary vitreous, ecthyma gangrenosum (by the septicemia of *P. aeruginosa),* and peripheric facial paralysis (a complication of acute otitis media), admitted to hospital.

key words: **children • ecthyma gangrenosum • facial paralysis • pseudomonas septicemia • persistent hyperplastic primary vitreous**

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Author's address: Ibrahim Hakan Bucak, Department of Pediatrics, Faculty of Medicine, Adıyaman University, Adıyaman, Turkey, e-mail: ihbucak@hotmail.com

Background

Pseudomonas aeruginosa is an opportunistic pathogen. It has been reported that *P. aeruginosa* affects the infection frequency in patients who have a predisposing factor. *P. aeruginosa* rarely leads to systemic disease in a previously healthy child [1]. Ecthyma gangrenosum (EG) is the characteristic skin lesion of *P. aeruginosa* [2]. Infection by *Staphylococcus aureus, Serratia marcescens, Aspergillus* spp and *Mucor* spp also lead to EG [3,4]. Persistent hyper-plastic primary vitreous (PHPV) has been described as a wide-spectrum disorder caused by the persistence of the fetal hyaloid vascular system during the development of the eye [5]. PHPV must be considered in differential diagnosis of leukocoria. Acute otitis media (AOM) is one of the most common infectious diseases of childhood; however, peripheral facial paralysis (PFP) is a very rare complication of AOM [6]. In this case report, we present a previously healthy pediatric patient who has been diagnosed with PHPV, EG (by the septicemia of *P. aeruginosa),* and peripheral facial paralysis (a complication of AOM). This patient came to our clinic with extensive ecchymosis and ulcers on his body.

Case Report

A previously healthy 5-month-old boy was hospitalized because of an ongoing 6-day fever, yellow purulent otorrhea, extensive ecchymosis, and necrosis on his body. In his medical history check, we learned that the patient had been diagnosed for AOM by another hospital and had been hospitalized in that institution. We were also informed that a skin eruption began on his body during the treatment received at that hospital. On physical examination, his temperature was 38.7°C, pulse 120/min, respiratory rate 45/min, blood pressure 85/55 mmHg, weight 9 kg (%90–97), height 68 cm (%75–90), and capillary refill time of 5 seconds. During clinical examination, his general appearance was not good. He had moderate dehydration and respiratory distress. He had yellow purulent otorrhea in his left ear. PFP was determined on the left side of his face (Figure 1).

His right eye had leukocoria. There were many ulcers with gangrenous centers surrounded by ecchymosis. From those, 4–5 patches were located on the front and back of the body, and 5–6 were located at the extremities (Figure 2).

His general chest examination revealed bilateral crepitant rales. Cardiovascular and abdominal examinations were normal. Complete blood count (CBC) showed pancytopenia and a band was seen >%10 at the peripheral blood smear. Fibrinogen was 369 mg/dl (220–496 mg/dl). Bleeding time was normal, but other coagulation parameters were abnormal. Erythrocyte sedimentation rate (ESR) was 80 mm/h. Biochemical results were normal. Laboratory results are shown in Table 1. Amphoric, amikacin, and vancomycin treatment were started after the blood culture and otorrhea swab cultures were sampled. The patient was started with red blood cell and fresh frozen plasma supplement. Every day, the wounds were cleaned and treated with topical antibiotics and saline.

Cranial CT, orbital CT and orbital ultrasonography were performed to explain the leukocoria and PFP. Orbital CT and orbital ultrasonography images were evaluated by the radiologist and ophthalmologist; retinal detachment and

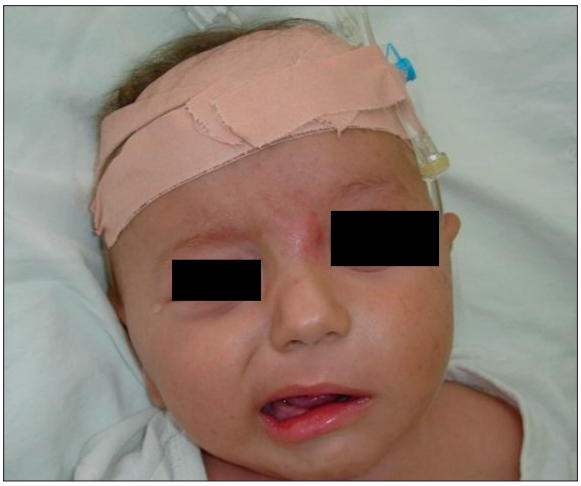

Figure 1. Photograpy of patient peripheral facial paralysis (PFP).

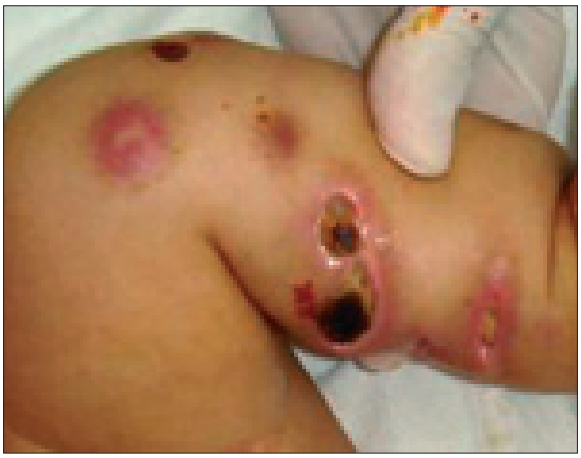

Figure 2. Photograpy of ulcers with gangrenous centers surrounded by ecchymosis on extremites.

PHPV were diagnosed. Cranial CT was reported as normal. PFP was thought to be a complication of the AOM. Systemic steroid and synthetic teardrops were used in the treatment. PFP was cured at the 21st day.

Blood culture and otorrhea swab culture were positive for *P. aeruginosa.* Immunoglobulins and lymphocyte subgroups of the case were normal compared with same-age children (Table 1). The nitroblue tetrazolium and sweat chloride test results were normal. Anti-HCV antibody and anti-HIV antibody were negative. Amikacin and vancomycin treatments were completed in 14 and 21 days, respectively. Control CBC, CRP, ESR and coagulation parameters of the patient were all normal.

Discussion

P. aeruginosa is a gram-negative rod, a strict aerobe, and a classic opportunist pathogen. It has been found to form colonies on skin, throat, stool or nasal mucosa. *P. aeruginosa* bacteremia in children was seen in 3.8 out of 1000 patients over the past 10 years. The average mortality rate is 20%. Infection frequency and mortality rate increase in patients with burns, trauma, malnutrition, cystic fibrosis, malignancies or immunocompromised situations. *P. aeruginosa* infection is rare in individuals without predisposing risk factors [2]. *P. aeruginosa*

Table 1. Hematological and chemical values in case.

	1st day	21st day
White cell count (/mm^3)	3600	12000
Haemoglobin (gr/dl)	6.6	14
Haematocrite (%)	20	41
Platelet (/mm^3)	54000	324000
CRP (<5 mg/L)	188	0.1
ESR (mm/hr)	80	7
PT (10–14 sec)	17.5	13
PTT (23–35 sec)	37	31
inr	1.2	1.1
IgA (4.4–84 mg/dl)	20	
IgM (33–126 mg/dl)	130	
IgG (172–1069 mg/dl)	250	

CRP – c-reactive protein; PT – prothrombin time; PTT – partial thromboplastin time; ESR – erythrocyte sedimentation rate.

has been reported as the cause of osteomyelitis, septicemia, meningitis, pneumonia, urinary tract infection, keratitis, endophthalmitis, external otitis and catheter-related infection [2,7]. EG, the characteristic skin lesion of *Pseudomonas*, is caused by direct inoculation or metastasis secondary to septicemia. EG begins as pink macules that progress to hemorrhagic nodules, which then turn into ulcers. This process generally takes about 12 hours [2–4]. *P. aeruginosa* produces lecithinase, collagenase, lipase, and hemolysins, which may cause skin ulcers [7]. EG has been reported to be caused by infection with *S. aureus, S. marcescens, Aspergillus* spp and *Mucor* spp; therefore, cultures must be taken to determine the agent [3,4,8]. *P. aeruginosa* was positive in the blood culture and otorrhea swab culture. Although lesions are generally found to be located in gluteal and perineal areas, the lesions of this specific case were located on the extremities and the body [4]. Ecthyma gangrenosum is very rare condition in a previously healthy child. In 2006 Viola et al reported *P. aeruginosa* sepsis in 73 previously healthy children without underlying medical problems [9]. In a clinical study by Huang et al., 121 previously healthy patients who were younger than 15 years and who were found to have *P. aeruginosa* septicemia, had complaints of high fever (91%) and diarrhea (72%). The characteristic skin lesion was 50–64% positive in some reports [1]. Fever and skin lesions were general complaints of our case. Furthermore, Huang et al reported that neutropenia (<5000/mm^3) (57%) and thrombocytopenia (<100000/mm^3) (34%) were the most common laboratory results. In our case, pancytopenia was determined in the CBC, and at the end of the treatment CBC was normal. In the literature, during the course of the disease or after the treatment, some complications can be determined, such as cyclic neutropenia, malnutrition, hypogammaglobulinemia, and abnormal neutrophil function [4,7]. Underlying disease must be investigated in cases such as this, but no abnormal results were determined.

AOM is the most common infection of childhood [6]. Facial paralysis is a very rare complication of AOM (seen in about 1–4% of cases), and antibiotic application decreases the appearance of facial paralysis [10]. Duman et al., Viola et al., and Patigaroo et al. reported ecthyma gangrenosum and AOM or externa with facial nerve palsy in healthy infants [9,11,12]. Our case had an AOM and PFP. Systemic antibiotic therapy and systemic steroids were used, and the treatment was a success. In some cases, facial nerve decompression treatment is necessary due to unsuccessful antibiotic and steroid treatment [6,10]. In our case no facial nerve decompression treatment was needed.

Differential diagnoses of leukocoria, congenital cataracts, PHPV, retinopathy of prematurity (ROP), and retinoblastoma must be considered [5]. There was no cataract at the eye examination in this case. ROP was not considered because of unilateral leukocoria and full-term birth history. Since orbital mass and calcification were not shown by the orbital CT and orbital ultrasonography, retinoblastoma diagnosis was excluded. Retinal detachment was shown by orbital ultrasonography, and hyaloid artery residue was seen by the orbital CT. The presence of unilateral leukocoria, retinal detachment and hyaloid artery residue are compatible with PHPV. PHPV is a clinicopathologic status that occurs by the persistence of fetal hyaloid fibrovascular tissue [5]. PHPV presented with unilateral leukocoria and/or microphthalmia, cataracts, retrolental opacity, persistent hyaloid artery, retinal dysplasia, and retinal detachment was seen at the orbital ultrasonography, orbital CT and orbital magnetic resonance with term infants [5,13]. We could find no report in the literature of cases with the simultaneous existence of both PPHV and ecthyma gangrenosum. Hence, we think that the occurrence of both PPHV and ecthyma gangrenosum in our case was a coincidence. PHPV treatment can be surgical or conservative. In our case surgical treatment aimed to prevent complications and to salvage the vision. After treatment of the EG and PFP, the patient was transferred to ophthalmology service for treatment of PHPV.

Conclusions

The association of PHPV, AOM, and facial paralysis with Pseudomonas sepsis and ecthyma gangrenosum has not been reported previously. This case is special because of its interesting association of a previously healthy infant with pseudomonas sepsis, ecthyma gangrenosum, facial paralysis and PHPV, as determined on the visual test.

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