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Case Report
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Pseudomonas Sepsis Associated with Skin and Cardiac Involvements in an Infant with Congenital Nephrotic Syndrome

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Abstract

Introduction: Ecthyma gangrenosum and infective endocarditis are two rare complications of Pseudomonas Aeruginosa infections.

Case Presentation: Our presenting case is an infant with congenital nephrotic syndrome who was admitted in the hospital with skin lesions of Ecthyma gangrenosum and manifestations of sepsis. Blood culture revealed Pseudomonas Aeruginosa infection. In spite of no pathologic sound in heart examination and dramatically response to antibiotics, echocardiography showed vegetation in right sided heart associated with mild tricuspid regurgitation

Conclusion: Cases of bacterial sepsis occurs in patients with implanted venous catheter should be evaluated for bacterial endocarditis even if there is a dramatically response and no finding suggestive of cardiac involvement in heart examination.

Keywords: Ecthyma gangrenosum; Infective endocarditis; Pseudomonas sepsis

Introduction

Ecthyma gangrenosum (EG), is a rare invasive cutaneous infection that in majority of cases caused by Pseudomonas. Aeruginosa (PA) and occurs in 30% of Pseudomonas septicemia [1]. This lesion is related to life-threatening septicemic infections and has high mortality. Infections of PA typically seen in immunocompromised patients with severe neutropenia [2], agammaglobulinemic cases or hypogammaglobinemia [3-4]. The EG lesions characterized by red maculae that progress to a hemorrhagic bluish bullae which rupture to form a central area of necrosis surrounded by an erythematous halo. Very rarely this invasive skin lesions may be reported in healthy cases [5-7]. Another rare complication of PA is infective endocarditis (IE) [8]. PA account for <1.8% of bacterial endocarditis [9]. Most cases of Pseudomonas IE seen in intravenous drugs abusers. Isolated right-sided pseudomonas IE generally can be managed with antibiotic therapy, with or without valve surgery [10].

Genetic defects account for the majority of CNS, but especially in developing countries, congenital infections (syphilis, toxoplasmosis, rubella, hepatitis B, human immunodeficiency and cytomegalic viruses) can result to CNS. Mutations in genes Nephrin 1 (NPHS1; Finnish type of CNS), NPHS2, Wilms tumor 1(WT1; Denys-Drash syndrome, Frasier, and WAGR syndromes), laminin-β2 (LamB2; Pierson syndrome), LamB3 (Herlitz junctional epidermolysis bullosa), phospholipase C epsilon 1 (PLCE1), LMX1B (nail-patella syndrome) account for genetic forms of CNS [11].

Case Presentation

Here we present a two-month boy with diagnosis of CNS in first week of life, with negative serologic tests for congenital infections. At first presentation Serum albumin and Creatinine were 1.8 gr /dl and 0.3 mg/dl respectively, and urine protein to Creatinine (mg/mg) ratio was 305/4.4. No genetic study was done since they were not available and kidney biopsy was planned to do after age 3 months to provide better information.

He admitted for fever (axillary temperature = 38.5 ˚C), generalized edema, tachypnea, grunting and poor feeding. A bout one month ago a deep central venous access devices (poly site) was

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Figure 1: Photograph of Ecthyma gangrenosum shows a dark necrotic center with surrounding halo. Necrotic lesion on lower extremities and face (at admission).

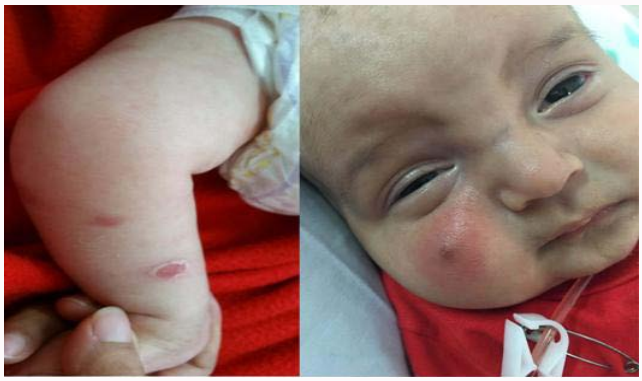

Figure 2: Skin lesions in lower extremities hanged to bullous lesions that ruptured. Erythema surrounding necrotic lesions in lower extremities abruptly decreased, with no prominent change in facial lesion.

implanted for daily Albumin infusion and furosemide. As there is no definite treatment except renal transplantation for patients with CNS, our case never reached remission of proteinuria during treatment. He didn't receive anti-aggregation treatment since parents were unreliable and we concern about risk of hemorrhage. We prefer to recommend them in cases with history of thrombosis.

Control of vital signs showed systolic blood pressure (BP) of 85 mmHg with no measurable diastolic BP, pulse and respiratory rates were 150/minute and 55/minute respectively. At presentation he had nasal flaring without cyanosis and normal heart and lung examinations. Oxygen saturation was 98% on oxygen and urine output was > 1cc/kg/ hour. Different erythematous macula lesions with a central necrotic configuration (target sign) were noted in the lower limbs and face (Figure 1). The lesions were typically EG. After a full laboratory assessment, treatment with Ceftazidime and Vancomycin was started. Vancomycin was started to cover staphylococcal nosocomial sepsis. Laboratory findings are listed in Table 1. Chest X-ray demonstrated prihilar infiltration with no evidence of bacterial lung involvement. In second day of admission, respiratory distress increased (oxygen saturation near to 85% at room), systolic BP decreased to 70 mmH, so Vancomycin changed to Meropenem and combination of Meropenem with Ceftazidime selected since PA sepsis was the most probable diagnosis and combination therapy for PA invasive infections was needed. Also the patient received packed cell infusion due to anemia. In the day 4 of admission patient had no respiratory distress and BP returned to normal values. Edema and induration of skin lesions abruptly decreased and lesions change to bullae that ruptured and a necrotic lesion at central part remained (Figure 2).

Blood culture defined growth of Pseudomonas Aeruginosa. At the day 10, however fever was resolved and there was no abnormal sound or murmur on heart examination, echocardiography was done to check bacterial vegetation on heart leaflets. Small vegetation in septal leaflets of tricuspid valve associated with mild valvar regurgitation were reported (Figure 3). At the day 12 of admission patients discharged and recommended to continue the antibiotics administration through polysite (for 4 weeks) and referee to cardiologist.

Discussion

Congenital nephrotic syndrome (CNS) which characterized by heavy proteinuria, hypoproteinemia, and edema starts soon after birth, and is a rare kidney disorder. The loss of the proteins in the urine leads to hypogammaglobinemia and serum IgG values commonly is below 2% of normal values in these cases. Urinary losses of gamma globulin and complement factors B and D prone patients to infections caused by capsular bacteria such as pneumococci [12]. Prophylactic use of immunoglobulin is not indicated since the infused immunoglobulin rapidly lost into urine [13].

PA is a common environmental gram-negative bacillus which acts as an opportunistic pathogen. Almost always P aeruginosa infections associate with the compromise of host defense mechanisms such as in acquired immune deficiency syndrome (AIDS), and neutropenic patients undergoing chemotherapy [11]. Cutaneous manifestations produced by Pseudomonas infections may be classified into two groups: primary skin lesions and those in which cutaneous lesions occur in the context of Pseudomonas bacteremia [14].

In our case it seems that the source of bacteremia was through central venous catheter. Catheter associated blood stream infections (CABSI) is a condition that has been reported in 7.6% of cases [15]. Infective endocarditis due to PA is a rare condition and less than 20 cases of P aeruginosa IE involving native valves of non intra venous drug users have been reported [16]. All of these cases had a predisposing factor include hemodialysis, cardiac catheterization and surgery, and gastrointestinal or genitourinary procedures. Our case had a central venous catheter a condition like that occurs in intra

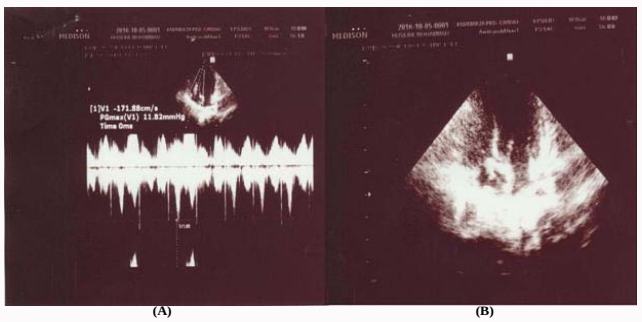

Figure 3: Small vegetation near to septal leaflets of tricuspid valve (M mode echocardiography), and mild tricuspid regurgitation (Doppler echocardiography).

Table 1: Laboratory findings in our patient at admission and during treatment.
1) Day, 2) Hemoglobin, 3) Hematocrit, 4) Erythrocyte sedimentation rate 1st hour, 5) C-reactive protein, 6) Partial thromboplastin time, 7) prothrombin time, 8) red blood cells.

Laboratory tests at admission	Laboratory tests at days 2-10 of admission
Complete blood cell (CB) : White blood cell count (WBC count): 18400 cell/µL Hb[2]: 5.4 gr/dL Hct[3]: 16.4% Platelet count: 163000 cell/µL Differential cell count : Poly morph nuclear cells 74%, Lymphocytes 21%, Band cells 5% ESR 1[4]= 125 mm/h CRP[5]= +++ PTT[6]= 72 (control= 30) seconds PT[7]= 17 (control= 12.5) seconds, INR= 1.8 Total protein= 2.8 mg/dl, Albumin= 1.7 mg/dl	WBC count= 28100 cell/µL (D 12), 16800 cell/µL (D3) 12900 cell/µL (D5) Hb= 11 gr/dL (D2 after packed cell transfusion), 9.3 gr/dL (D3) and 7.1 gr/dL (D5) Platelet count: 50000 cell/µL (D2), 36000 cell/µL (D3), 109000 cell/µL (D5) CRP= negative (at discharge) PTT= 39 (control= 30) seconds (D3)
Blood sugar = 104 mg/dl Urea = 19 mg/dl Creatinine = 0.6 mg/dl Sodium (Na)= 134 meq/L Potassium (K)= 2.9 mg/dl Total Calcium = 8 mg/dl Phosphorus = 5.1 mg/dl Venous blood gases analysis (VBG): PH= 7.34 , PCO_2= 21.8 mmHg, Po_2= 52.2 mmHg O_2 saturation=88.1 % , HCO_3= 14.6 mmol/L BE= -8.3 mmol/L	K= 2.8 mg/dl (D3), K= 4.1 mg/dl (D5), K= 3.1 mg/dl (D10),
Blood culture: Growth of Pseudomonas SPP Sensitive to Amikacin, Ceftazidime, Ceftriaxone, and Meropenem– Piperacillin Tazobactam	
Urine analysis: Specific gravity= 1005 PH= 5 Protein= ++ blood= + WBC= 1-2 cell/HPF RBC[8]= 4-5 cell/HPF urine culture= negative	

venous drug users and predispose them for blood invasion of bacteria [8].

Our case was an infant with a primary kidney diseases (CNS). Massive urinary loss of proteins, gamma globulin and complement factors B and D, and implanting central venous catheter were predisposing factors for invasive bacterial infections. Study by Harris in cases with CNS revealed that the serum IgG levels were <25% in all and most cases had a level <2% of normal infant values [17]. Low serum gamma globulin levels and deficiency of complement factors B and D predispose cases to infection with capsular bacteria such as Streptococcus Pneumonia Other study reported that septic episodes in infants with CNS mainly caused by staphylococci and coliforms [13]. Our case is interesting since Pseudomonas infections are rare in CNS. In addition two rare complications of the infection were found in our patients, including EG and IE, both are rare in clinical practice of Pseudomonas aeruginosa infections.

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