Case Report / Olgu Sunumu

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Auto-amputation of the Limbs and Early Colostomy's Effect on Life Quality and Survival on the Patients with Severe Meningococcemia Related Purpura Fulminans: Two New and Different Approaches

Meningokoksemiye Bağlı Şiddetli Purpura Fulminans Hastalarında Ekstremitelerin Oto-ampütasyonu ve Erken Kolostominin Yaşam Kalitesi ve Sağkalıma Etkisi: İki Yeni ve Farklı Yaklaşım

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Abstract

Purpura fulminans (PF) is a rare but fatal thrombotic disease caused by microvascular thrombosis due to coagulation disorder. Necrosis can progress to muscle and bone tissue contributing to late mortality and morbidity. A healthy 4-month-old girl and a 7-month-old girl had previously admitted to our intensive care unit with severe PF due to meningococcemia. Both patients had a severe and critical period in the pediatric intensive care unit and were supported with extracorporeal treatments. We opened colostomy for perineal infection because of large and deep skin lesions. In addition, we waited for a full auto-amputation involving all extremities instead of early surgical amputation. Both survived and acceptable limb function and some mobilization capacity were preserved. In conclusion, we think that early colostomy in severe perineal infections and auto-amputation options in severe extremity involvement should be preferred in order to get better results.

Keywords: Purpura fulminans, pediatric intensive care, auto-amputation, colostomy, children

Öz

Purpura fulminans (PF), pıhtılaşma bozukluğuna bağlı mikrovasküler trombozun neden olduğu nadir fakat ölümcül bir trombotik hastalıktır. Nekroz, kas ve kemik dokusuna ilerleyerek geç mortalite ve morbiditeye katkıda bulunabilir. Dört aylık sağlıklı bir kız ve 7 aylık bir kız çocuğu daha önce yoğun bakım ünitemize meningokoksemiye bağlı şiddetli PF ile başvurmuştu. Her iki hasta da çocuk yoğun bakım ünitesinde ağır ve kritik bir dönem geçirdi ve ekstrakorporeal tedavilerle desteklendi. Geniş ve derin deri lezyonları nedeniyle perineal enfeksiyon için kolostomi açtık. Ayrıca erken cerrahi ampütasyon yerine tüm ekstremiteleri kapsayan tam bir otoampütasyon beklendi. Hem hayatta kalmış hem de kabul edilebilir uzuv işlevi ve bazı mobilizasyon kapasitesi korunmuştur. Sonuç olarak, şiddetli perineal enfeksiyonlarda erken kolostomi, ağır ekstremite tutulumlarında otoampütasyon seçeneklerinin daha iyi sonuç almak için tercih edilmesi gerektiğini düşünmekteyiz.

Anahtar Kelimeler: Purpura fulminans, pediyatrik yoğun bakım, otoampütasyon, kolostomi, çocuk

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Introduction

Purpura fulminans (PF) is a mortal thrombotic disease caused by coagulation disorder related microvascular thromboses which leads to rapid skin necrosis and disseminated intravascular coagulation.[1] PF related morbidity consists of scarring and amputation of fingers and extremities.[2]

Patients should be evaluated rapidly with support of airway, breathing and circulation in case of need, high concentrate oxygen administration, intravenous (IV) or intraosseous line access with rapid fluid resuscitation followed by needed culture studies and IV antibiotic administration. Patients non-responsive to IV fluid resuscitation should be given inotropic or vasopressor agents.[3] We would like to share our experience and management of two pediatric PF cases.

Case Reports

Case 1

Previously healthy 4-month-old girl was admitted to another hospital with 38.6 °C axillary temperature and petechial rash on legs. Patient's clinical status deteriorated rapidly in following few hours with rash spreading to arms and trunk.

Glasgow Coma scale was 7 and therefore patient was intubated (Figure 1A). Patient was given 20 cc/kg 0.9% normal saline 4 times for severe hypotension afterwards adrenalin infusion (0.2 mcg/kg/min), noradrenaline infusion (0.1 mcg/kg/min) and dopamine infusion (10 mcg/kg/min) had started for refractory septic shock. Despite being given catecholamine infusions, patient's shock findings continued, so hydrocortisone treatment was added (50 mg/m^2), adrenaline and noradrenaline doses were titrated for refractory hypotension. Blood culture was positive for *Neisseria meningitidis* whereas cerebrospinal fluid (CSF) culture did not reveal any organisms. The 6th day of her hospitalization, she had oliguria unresponsive to furosemide treatment which progressed to pulmonary edema, so patient received venovenous hemodialysis for 4 days. Patient was consulted to pediatric immunology and allergy department for severe disease, but work-up and patient's history showed no sign of immune deficiency. On the 25th day she was extubated and proceeded to high flow nasal cannula oxygenation. On the 23rd day, colostomy was opened by pediatric surgery department for severe tissue loss of anogenital region (Figure 1B). On the 87th day; after epithelization of perianal and gluteal lesions; colostomy was closed. Patient was consulted to plastic surgery department for distal extremity necrosis hence debridement of lesions was made (Figure 1C). The patient was discharged on the 127th day. After being discharged, the patient received 2 operations for tissue loss of extremities. The first operation was made by plastic surgery department at 21 months of age for tissue loss of lower extremities with skin graft taken from patient's back. In operation, elongated bleeding led to evaluation of coagulation factors hence her factor VII level was found to be low (21.1%). She was operated for the second time at 26 months of age for correction of upper extremity contractures. She is now 28 months old; her cognitive functions are well. She can sit and walk on her own. She can use her right hand and fingers despite losing her distal fingers of right hand. She also has upper and lower extremity contractures (Figure 1D).

Case 2

Previously healthy 7-month-old girl was admitted from another hospital with 2-day history of fever and purpuric rash on her body (Figure 2A). First physical examination at pediatric intensive care unit (PICU) showed that the patient as in generally bad condition with bilateral poor light reflexes,

She was given 20 cc/kg serum physiologic 3 times for her tachycardia, hypotension and prolonged capillary refill time. In the following hours, patient remained hypotensive and her lactate levels increased, therefore adrenaline (0.1 mcg/

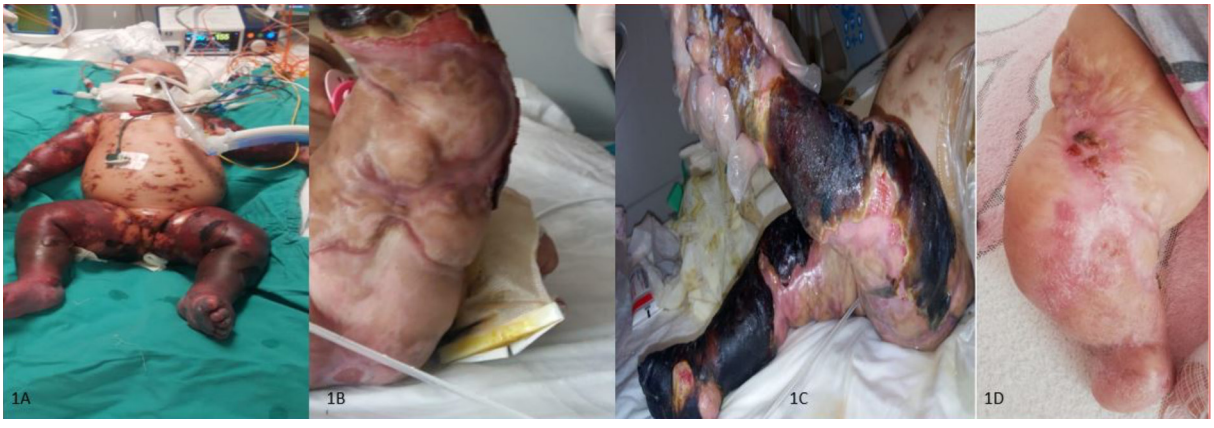

Figure 1. 1A) Diffuse purpuric rash, **1B)** recovery after colostomy, **1C)** ischemia and necrosis of PF, **1D)** Auto-amputated right foot and right leg contracture
PF: Purpura fulminans

kg/min), noradrenaline (0.1 mcg/kg/min) and dopamine (10 mcg/kg/min) infusions were started and increased as needed. Patient's both legs appeared ischemic on the end of the second day. Doppler ultrasonography showed no signs of blood flow on the distal extremities, so IV nitroglycerine infusion was started for vasodilator effect. Bacterial panel of CSF was positive for *N. meningitidis*. Patient was consulted to pediatric immunology and allergy department for severe disease, but work-up and patient's history showed no sign of immune deficiency. In the following days, the lesions on the trunk, face and arms fell off but remained ischemic in both legs (Figure 2B). There were also lesions in the epiglottis. Doppler ultrasonography of the lower extremities revealed blood flow in the femoral, popliteal and posterior tibial arteries. On the 25th day, a colostomy was opened for deep tissue loss and delayed healing of perineal lesions. After the colostomy was opened, patient's perianal legions rapidly healed. Necrosis in the lower extremities and fingers of the patient was left to auto-amputation. The patient was extubated on the 26th day. Currently 17 months old, his right foot was lost due to auto-amputation. He can sit on his own but cannot walk. The patient's colostomy was closed by the pediatric surgery department after 2 months (Figure 2C).

Discussion

PF is a mortal syndrome with acute onset, progressive necrosis and cutaneous bleeding and usually associated with meningococcemia, sepsis, varicella zoster, pneumococcal and meningitis infections.[4] In a study of Gürgey at al.[5] on showed that, 16 patients whose ages were between 3.5 months and 12 years with PF were caused by infections on 7. Our first patient's blood culture and second patient's CSF culture was showed to be positive for *N. meningitidis*. First patient was diagnosed with factor VII deficiency. PF can be observed in neonatal period because of congenital or acquired protein C deficiency or other coagulation disorders.[6] Prothrombotic risk factors such as factor V Leiden mutation, prothrombin G20210A mutation and antiphospholipid antibody were all negative in our cases.

PF related lesions are mostly seen on trauma and pressure exposed regions such as hip, extremities, trunk and scalp. Treatment usually consists of debridement and skin graft surgeries.[7] Our two case showed lesions mostly on upper and lower extremity, additionally our first case had lesions on her left lower eyelid whereas our second case had lesions on her epiglottis. Debridement of necrotic tissues in the lower and upper extremities was performed by the plastic surgery department.

PF treatment consists of treating the underlying cause, recovery of normal coagulation functions, preventing thrombosis related tissue damage and decreasing mortality and morbidity. There isn't a globally accepted treatment strategy for treating PF but antimicrobial therapy and TDP administration is suggested.[8] A 10-year study of Warner et al.[6] showed that, among 70 patients (mean age: 13) with PF, proven etiology was *N. meningitides* among children whereas *Streptococcus pneumonia* was the main etiologic agent among adults. Patients were given fast antibiotic administration, fluid resuscitation, respiratory and inotropic support with corticosteroid therapy and protein c replacement if needed. 90% of the patient's lesions led to complete layer skin necrosis with a need for debridement and 25% of the patients received surgical limb amputation.[6] Ahmad et al.[9] reported on their 2 case that, meningococcemia related PF should be managed by PICU for vital functions, plastic surgery consultation should be made if wide purpuric-necrotic lesions are present and should be given multidisciplinary approach. Both of our cases were consulted to plastic surgery department for skin necrosis and debridement and skin grafting were made by them. Additionally, we experienced that partial exchange had a positive effect on recovery.

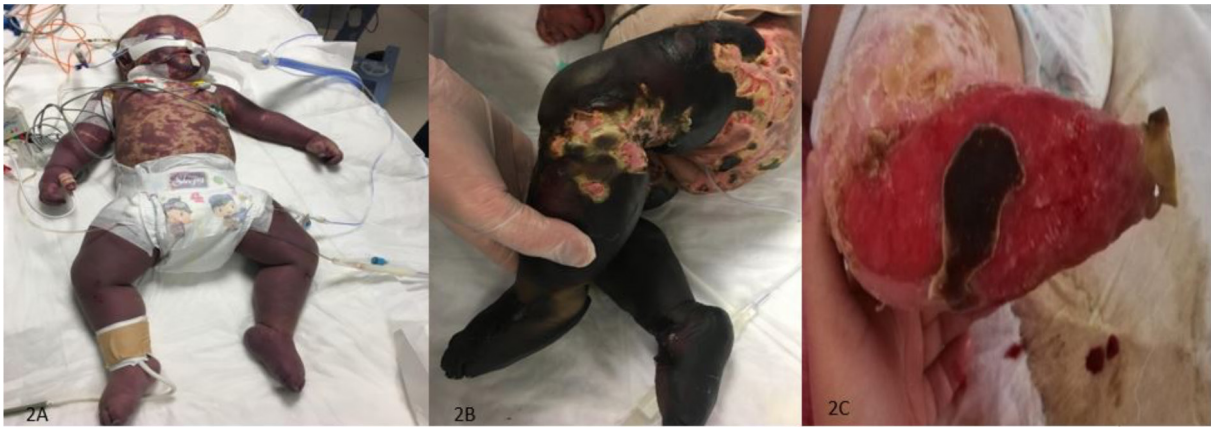

Figure 2. 2A) Whole body was covered by PF, **2B)** necrotic tissue of the limbs, **2C)** auto-amputation of the right foot
PF: Purpura fulminans

PF's mortality rate is approximately 40%.[6] Despite the fact that our two cases had severe illness with perineal scar which resulted with colostomy, No literature unnecessary need of long term mechanic ventilation and continuous renal replacement therapy; we managed to give survival and good prognosis. Perianal lesions may cause gastrointestinal flora associated severe sepsis which was reported to have high mortality rates of 78%.[10] In immunocompromised patients, pseudomonas aeruginosa infections remain to be one of the main reasons for disseminated infections which can progress to deep tissues.[11] Necrosis of perianal region may cause sphincter dysfunction and therefore secondary incontinence. Treatment strategy consists of antimicrobial therapy, debridement of necrotic tissue and skin transplantation.[12] Colostomy may be needed for speeding the recovery of affected colon and skin.[12] irrelevant We decided to open colostomy for preventing the necrotic tissue from contamination with gastrointestinal flora via stool, therefore preventing secondary infections and fastening recovery of the skin.

Perianal lesions may cause severe sepsis due to gastrointestinal flora, which has been reported to have a high mortality rate of 78%.[10] We decided to open a colostomy to prevent contamination of necrotic tissue with the gastrointestinal flora through feces, thus preventing secondary infections and accelerating the healing of the skin. It resulted in rapid recovery of perianal lesions.[13]

Meningococcemia, wide purpuric necrosis of the extremities may result with amputations. If necrosis induces sepsis itself, amputation is indicated.[14] Gürgey et al.[5] reported that, out of the 16 patients of their PF study, 3 of them had bilaterally hand, 2 of them had finger, 3 of them had foot finger, 1 of them right hand and 1 of them bilaterally lower extremity amputations. Ghosh et al.[15] reported (similar results) on 28 pediatric patients with PF. One patient had to undergo a below knee surgical amputation and one patient had auto-amputation of the digits. Powars et al.[16] they reported that 10 of 28 patients with PF developed deforming auto-amputation secondary to dermal microvascular thrombosis and hemorrhagic necrosis. Both of our cases were left to auto-amputation. Although only partially compatible with literature, we observed auto-amputation to be related with better life quality, posture and walk. Therefore, we think that auto-amputation is a better extremity protected approach rather than surgical amputation.

In conclusion, meningococcemia related PF is a rare and highly mortal disease. Early term colostomy prevents secondary sepsis and quicken perianal open scars. Lastly, we think that meningococcemia and PF related ischemic tissue loss should be managed with auto-amputation rather than surgical amputation for we think that auto-amputation results with more extremity tissue and life quality. Therefore, in the presence of meningococcemia related extremity ischemia, surgical amputation should not be preferred. Further studies and case reports are needed in this subject.

Ethics

Informed Consent: Approval was obtained from the family of the participants.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Concept: E.B., T.K., E.İ., E.Ç.T., H.Ö., Design: E.B., T.K., E.İ., E.Ç.T., H.Ö., Data Collection or Processing: C.Y., E.B., E.G., A.G., Writing: E.B., T.K., E.G., C.Y., A.G., B.B., F.K., S.S., E.Ç.T., H.Ö., E.Ç., E.İ.

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References

1. Hazelzet JA. FCCM. Diagnosing meningococcemia as a cause of sepsis. Pediatr Crit Care Med. 2005;6(Suppl 3):S50-4.
2. James WD, Elston DM. Cutaneous vascular diseases, in Andrew's diseases of the skin: clinical dermatology. 10th ed. Philadelphia;2006.
3. Dinh TA, Friedman J, Higuera S. Plastic surgery management in paediatric meningococcal-induced purpura fulminans. Clin Plast Surg. 2005;32:117-21, ix.
4. Smith OP, White B. Infectious purpura fulminans: diagnosis and treatment. Br J Haematol. 1999;104:202-7.
5. Gürgey A, Aytac S, Kanra G, Secmeer G, Ceyhan M, et al. Outcome in children with purpura fulminans: report on 16 patients. Am J Hematol. 2005;80:20-5.
6. Warner PM, Kagan RJ, Yakuboff KP, Kemalyan N, Palmieri TL, et al. Current management of purpura fulminans: a multicenter study. J Burn Care Rehabil. 2003;24:119-26.
7. Alyamac Dizdar E, Oguz S, Nur Sari F, Erdeve O, Ozkan Ulu H, et al. Enterocutaneous fistula secondary to purpura fulminans in a preterm infant. J Pediatr Hematol Oncol. 2009;31:753-5.
8. Darmstadt GL. Acute infectious purpura fulminans: pathogenesis and medical management. Pediatr Dermatol. 1998;15:169-83.
9. Ahmad Z, Jordan R, Das-Gupta R, Srivastava S. Two cases of meningococcal purpura fulminans: the 'less is more' approach. Int Wound J. 2014;11:636-40.
10. Morcos B, Amarin R, Abu Sba A, Al-Ramahi R, Abu Alrub Z, et al. Contemporary management of perianal conditions in febrile neutropenic patients. Eur J Surg Oncol. 2013;39:404-7.
11. Chan YH, Chong CY, Puthucheary J, Loh TF. Ecthyma gangrenosum: a manifestation of Pseudomonas sepsis in three paediatric patients. Singapore Med J. 2006;47:1080-3.
12. Khalil BA, Baillie CT, Kenny SE, Lamont GL, Turnock RR, et al. Surgical strategies in the management of ecthyma gangrenosum in paediatric oncology patients. Pediatr Surg Int. 2008;24:793-7.

13. Quarmby CJ, Millar AJW, Rode H. The use of diverting colostomies in paediatric peri-anal burns. Burns. 1999;25:645-50.

14. Ichimiya M, Takita Y, Yamaguchi M, Muto M. Case of purpura fulminans due to septicemia after artificial abortion. J Dermatol. 2007;34:786-9.

15. Ghosh SK, Bandyopadhyay D, Dutta A, Jane EP, Biswas SK. A Profile of 23 Indian Patients with Purpura Fulminans: A Retrospective, Descriptive Study. Indian J Dermatol. 2020;65:381-7.

16. Powars D, Larsen R, Johnson J, Hulbert T, Sun T, et al. Epidemic meningococcemia and purpura fulminans with induced protein C deficiency. Clin Infect Dis. 1993;17:254-61.