

Purpura Fulminans in a 9-Year-Old Boy with Auto-Amputation of Digits: A Case Report

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Abstract

Purpura fulminans may have infectious, hereditary and idiopathic etiology. We came across a case of purpura fulminans in a 9-year-old boy who had been healthy for first 4 years of his life and suddenly developed gangrenous lesions on extremities which proceeded to the detachment of some digits. Laboratory evaluation revealed mild anemia, hypoalbuminemia, high platelet count, elevated aPTT, PT and INR, mild proteinuria and high urinary protein to creatinine ratio. Child was managed by warfarin therapy.

Keywords: Hypoalbuminemia • Purpura fulminans • Hypotension

Abbreviations: DIC: Disseminated Intravascular Coagulation • aPTT: Activated Partial Thromboplastin Time • PT: Prothrombin Time • INR: International Normalized Ratio • RBCs: Red Blood Cells • tPA: Tissue Plasminogen Activator • PROC: PROtein C • IV: Intravenous

Introduction

Purpura fulminans is a syndrome characterized by progressive hemorrhagic infarction of skin and intravascular thrombosis [1]. It is often classified into three etiological types, viz. idiopathic, neonatal and infectious with infectious type being commonest of all [2]. Neonatal form result from inherited deficiency of vitamin K dependent pro-fibrinolytic proteins, protein C and S; infectious form is a consumptive coagulopathy [1] due to acquired deficiency of above mentioned proteins by endotoxins of gram negative bacteria especially *Neisseria meningitidis* and *Streptococcus pneumoniae* while idiopathic is rarest and least understood of all, however some autoimmune mechanism has been proposed [2]. Clinically it can present both in acute and chronic forms where acute form presents with some acute infection or severe septicemia whereas chronic form manifests some days after a febrile illness in pediatric patients usually [3]. The disease begins with appearance of cutaneous erythema which progresses to central irregular blue-black necrotic areas being painful at first but later become painless due to sensory loss over the skin owing to extensive necrosis [2]. Many patients present with classical symptoms of acute attack of Disseminated Intravascular Coagulation (DIC) [4] like fever, dyspnea, bleeding, bruising and hypotension [5]. Besides meningococemia and pneumococcal infections, some common illnesses have been reported to have purpura fulminans as their uncommon sequelae including *Hemophilus influenzae* type b infection [6], varicella [7], malaria [8-15], rickettsial infection [9], staphylococcal infection [10], candidiasis [11], Mowat Wilson syndrome with asplenia [12], *Capnocytophaga canimorsus* infection after dog bite [13], dengue hemorrhagic fever [14], *Lactobacillus paracasei* liver abscess [16], *Vibrio vulnificus* Septicemia [17], *Klebsiella rhinoscleromatis* septicemia [18], *Salmonella meningitis* [19], *Xanthomonas maltophilia* infection with aplastic anemia [20], and *Serratia marcescens septicemia* [21].

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Case Report

A 9-year-old boy, normal by birth, spiked fever at the age of 4 years due to a bacterial infection which led to dark necrotic lesions on limbs finally causing spontaneous detachment of distal and middle phalanges of 4 fingers of right hand, 2 fingers of left hand, all toes of left foot and 2 toes of right foot for which some laboratory investigations were done at that time but they could not be retrieved, hence, the precipitating agent left unexplored. Later, at the age of 7 years, his limbs began blackening and his blood workup (Table 1) revealed low aPTT and PT with elevated platelet count but normal INR while urinalysis showed mild proteinuria, 3+ hematuria, some pus cells and RBCs. At 9-year-age, he now presented with fever of 103°F for 4 days which was sudden in onset and continuous in character. On examination, some already darkened and some darkening necrotic tissue on left foot stump, knees, forearms and extensor surfaces of legs was observed. Laboratory evaluation of the blood of child showed mild anemia, hypoalbuminemia, high platelet count, elevated aPTT, PT and INR and on urinalysis, mild proteinuria, increased urinary protein to creatinine ratio were observed. On Doppler studies, perfusion of all limbs was found to be normal. His blood group was B+ and had no previous history of blood transfusion or family history of purpura fulminans. After warfarin therapy, coagulation profile as well as symptomatology of the patient improved. His laboratory reports for protein S and C showed no deficiency state after which he was discharged from the hospital.

Table 1. Laboratory investigations.

Variables	Sep 2017	Feb 2020
Blood		
aPTT (s)	25.9	50.5
PT (s)	10.6	48.6
INR	1.0	4.4
Platelet count/ μ L	587,000	629,000
Hb (g/dL)	10.1	10.3
Albumin	Normal	Low
Urine		
Proteinuria	+	++
Hematuria	+++	-
Pus cells	Present	Absent
RBCs	Increased	Normal
Urinary protein to creatinine ratio	Normal	Raised

Discussion

Purpura fulminans was first observed and mentioned as a separate disease in 1884 by Guelliot. This is a rare condition with prevalence of neonatal/hereditary form being 1 in 500,000-1,000,000 [22], high morbidity, and case fatality rate of 50% [23]. Its neonatal form is caused by genetic mutation of PROC (PROtein C) gene and can be inherited both in autosomal dominant (monoallelic mutation) and autosomal recessive (biallelic mutation) with latter manifesting clinically as purpura fulminans and being more severe [24]. In case of infectious etiology, endotoxins from infectious agents cause marked quantitative decrease in the protein C and antithrombin levels [25] pointing towards consumptive coagulopathy. Diagnosis of purpura fulminans requires both clinical and laboratory assessment. Main laboratory investigations significant for its diagnosis include, full blood count for sepsis, blood coagulation parameters (aPTT, PT, INR) for evaluation of functional integrity of clotting and anti-clotting mechanisms, blood levels of Protein C and Protein S, and genetic testing for PROC mutations [26]. Specific testing for pathogens can be done if there is high index of suspicion e.g., serology for varicella. In case of acute attack, both labs and clinical picture show signs indicating DIC while in disease developing over time, there may be variable presentations ranging from mild thromboembolic phenomenon with little changes in laboratory parameters to severe ischemic necrosis of the extremities with highly deranged coagulation profile.

Heparin, Protein C extract, fresh frozen plasma, antithrombin-III infusion and currently recombinant t-PA (tissue plasminogen activator) are main resorts of medical management of the coagulopathy seen in the disease. Broad spectrum intravenous antibiotics can be given in patients with sepsis. Symptomatic management can be done by epoprostenol, topical nitroglycerin, IV dextran and plasmapheresis while role of epsilon aminocaproic acid, vitamin K, ketanserin, warfarin, hirudin and glucocorticoids remains controversial in this regard. In the case presented in our study, patient was normal by birth for 4 years after which he developed post-infectious purpuric lesions and he did not have any family history of the disease. It rules out neonatal form, so the probable etiological basis of patient's disease was infectious. Literature has reported few cases of purpura fulminans with auto-amputation of the digits, but no such previous case has been reported from Rawalpindi Medical University Allied hospitals and even from the whole country.

Conclusion

Purpura fulminans is a rare disease usually affecting pediatric group with varying severity but very often associated with disability and death as it causes irreversible damage. Determination of its cause is mandatory for directing treatment strategies and ultimate recovery of patient depends on the cause, severity and extent of the disease.

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