

First hemoadsorption using CytoSorb in a pediatric patient with toxic epidermal necrolysis

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Background/Aims. Toxic epidermal necrolysis (TEN) is an adverse drug reaction associated with the separation of skin and mucous membranes at the dermal-epidermal junction. TEN is characterized by widespread keratinocyte apoptosis and necrosis: the major histocompatibility complex class I leads to clonal expansion of CD8+, which infiltrate the skin while soluble factors (TNF- α , INF- γ and iNOS) induce apoptosis (Figure 1). The disease is extremely rare but particularly serious: mortality is 20 to 25% during acute phase, and from 30-35% at one year. Treatment is essentially supportive, as no effective etiological treatment has been identified.

Methods. We describe a case of a child 8 years-old with an extremely severe clinical picture of TEN (>95% of BSA), SCORTEN score 5 (estimated risk of death \geq 90%) Figure 2.

Fig. 1. Proposed pathogenic mechanism in TEN

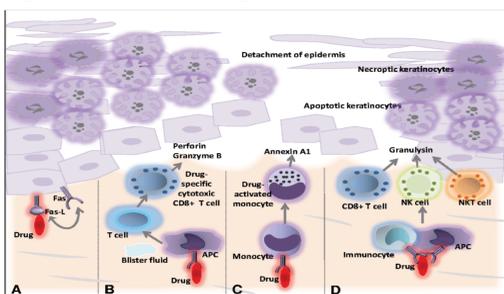


Fig. 2. Scortsen score

Parameters	Scortsen	Probability of death (%)
Age more than 40 years	0-1	3
Presence of malignancy	2	12
Epidermal detachment more than 30%	3	35
Heart rate more than 120/min	4	58
Bicarbonate less than 20 mmol/l	5 or more	90
Urea more than 10 mmol/l		
Glycaemia more than 14 mmol/l		

Results. The child was admitted in emergency department for purpuric macules in the upper trunk and in the face with a rapid spread to a confluent erythema. The family reported oral intake of cephalosporins a week ago. After 24 hours the child developed a progressive respiratory distress and he was transferred in PICU with need of tracheal intubation. The dermatological picture evolved to an extensive detachment of sheets of epidermidis associated to an erosive and hemorrhagic mucositis and he became deeply hypotensive with need of high doses of catecholamines (Adrenaline 0,15 mcg/kg/min, Noradrenaline 0,25 mcg/kg/min, Vasopressin 0,07/Ui/kg/h) associated to iperlactatemia (4.5 mmol/l) and metabolic acidosis.

Fig.3 a,b,c: progressive evolution of the skin lesions from purpuric macules to flaccid bullae and complete detachment of the epidermis from the dermis.

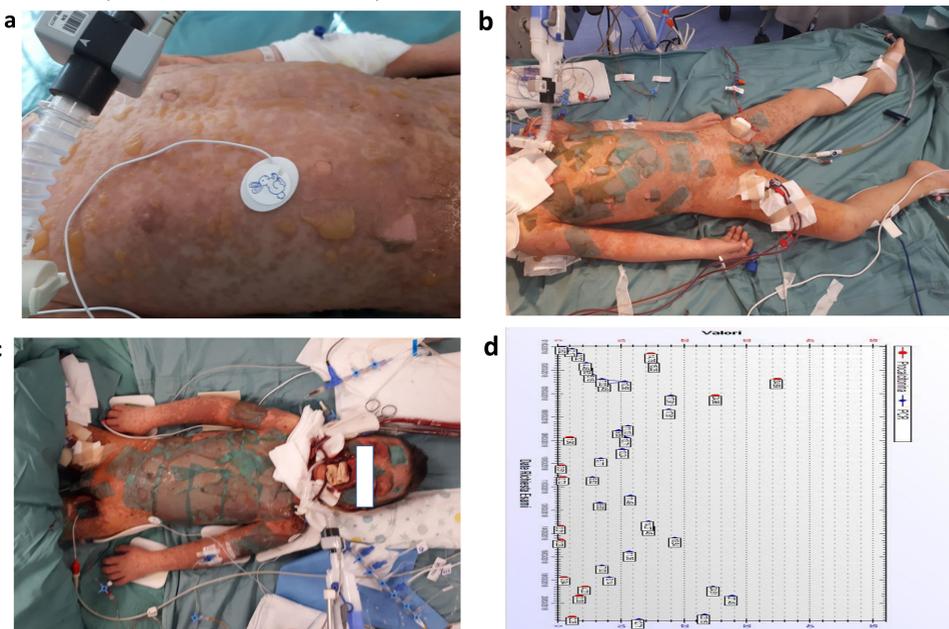
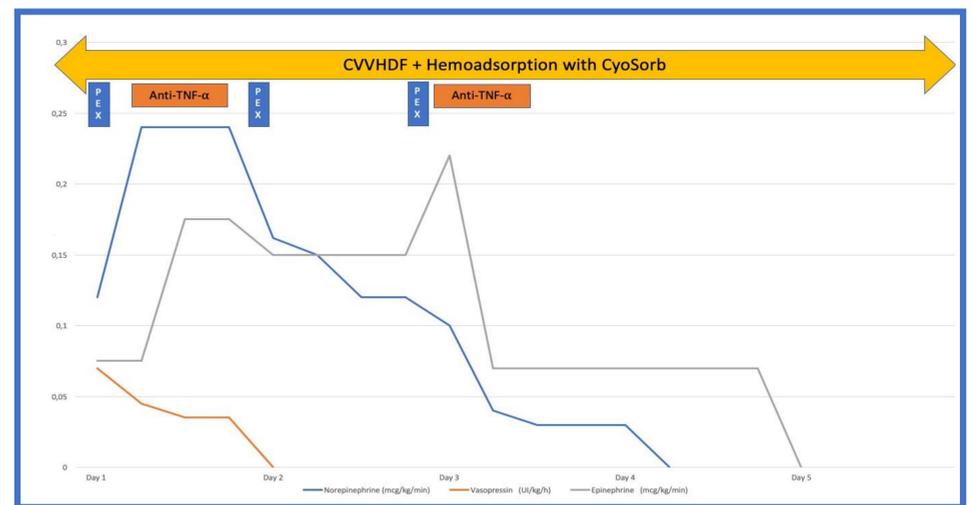


Fig.3 d: trend of the protein C reactive (PCR) and procalcitonin (PCT) during extracorporeal blood purification

Then for massive pulmonary edema and refractory hypoxia a VA ECMO was started. For the rapid evolution of the clinical picture despite the supportive care we decide to start an hemoperfusion with CytoSorb associated to CVVHDF to manage fluid overload. Hemoperfusion continued for 120 hours with three session of plasma-exchange and two administrations of anti-TNF α (1st and 4th days). During these 5 days we assisted to a significative reduction of vasopressors and inotropes (Figure 1). At 18th day he was weaned from ECMO support: unfortunately he died a week later due a Acinetobacter Baumanii septic shock.

Figure 4. Vasopressors and inotrope trend during extracorporeal blood purification



Conclusion. Considering that the pathophysiological process of TEN is by drug-induced cytotoxic T lymphocytes, hemoperfusion can remove soluble-mediators without the potential risks of the transfusion-associated therapies of plasma-exchange. Previously positive experiences of intermittent hemoperfusion in TEN has been already described. We describe the first use of CytoSorb in a pediatric patient with TEN: CytoSorb offered the opportunity to perform a continue hemoperfusion along 24 hours showing beneficial effect on hemodynamic stabilization. Our clinical case was characterized by an extremely severe clinical picture of TEN requiring also an ECMO support, although he died for an infectious complication correlated with the high surface of denuded skin, the patient overcame the acute phase with the combination of supportive and immunomodulatory treatment, based primarily on the hemoperfusion with CytoSorb.

Bibliography

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