

CASE REPORT

Lyell syndrome: a new therapeutic approach*La sindrome di Lyell: nuovo approccio terapeutico*M. GHIGLIONE¹, R. RIZZO¹, G. GIANNINI², F. MIGLIORI¹¹ Plastic Surgery and Burn Care Unit; ² ImmunoHematology and Transfusional Service, "San Martino" General Hospital, Genoa, Italy**KEY WORDS**

Lyell's syndrome • Intravenous immunoglobulins • Plasmapheresis therapy

PAROLE CHIAVE*Sindrome di Lelly • Immunoglobuline intravenosa • Plasmaferesi***Summary**

Lyell's Syndrome (LS) is described as a toxic epidermal necrolysis. Its main symptoms are bulbous lesions of the skin (flittenna), due to dermo-epidermal detachment interesting at least the 30% of the body surface, particularly concentrated around the mucosal areas. The whole clinical aspect is very similar to a main burn lesion.

Riassunto

La sindrome di Lyell viene descritta come una condizione di necrosi tossica dell'epidermide. I principali segni clinici sono rappresentati da gravi lesioni della cute (flittene), a causa della esfoliazione dermo-epidermica a carico del 30% della superficie corporea, particolarmente localizzata a carico della mucosa orale.

Introduction

Lyell's Syndrome is a very rare condition, its occurrence having been estimated about 1/2 cases for 1 million people/a year.

Etiopathogenesis of LS is not yet today very well known and/or understood: it is usually secondary to one or more drug administration. Therefore it is supposed that more than one cause¹ could be involved, something like a genetically determined drug metabolism combined with a immunological cell-carried reaction.

Mortality is 40-50% in the average. The first clinical signs on skin and mucosa appears between the fourth and sixth week from the beginning of the pharmacological therapy.

Until now the treatment of LS was based on the suspect drug administration suspension, maintenance of an adequate caloric balance, checking and supporting blood circle, protecting and medicating the sore areas, and ev. administration of immunoglobulins at therapeutic dosage (0.4-0.5 gr/kg/die)²⁻⁶.

Material and method

Our Burn Care unit has joined a new therapy protocol of LS projected by Marco Lissia (Department of Plastic Surgery and Burn Care Unit of the University of Sassari).

This protocol assumes the following steps:

- Utilization of fluidized beds type "Monarch VI"
- Positioning of CVC to control the support therapy by the CVP

- Diuresis monitoring by a urethral catheter
- Total parenteral nutrition as well as for any other patient with severe mucosa affections
- Gastro-protector drugs administration
- Limit antibiotics administration only in cases of confirmed infections
- Topical medications of skin lesions
- Use of Intravenous ImmunoGlobulins (IVIG) in association with
- Plasmapheresis

The patients follow two cycles of IVIG: the first three days of treatment 0.8-1 gr/kg/day are administrated; the following three days reduced to 0.5 gr/kg/die. The plasmapheresis therapy is performed, in association with the IVIG, at the 2°, 4° and 6° day from the beginning^{7,8}.

Our department has treated 2 female patients 60 and 74 years old.

After diagnosis and identification of the faulty drugs, performed in other units, the patients were transferred in our burn care unit. On arrival both patients presented severe mucosal involvement, flittenna and detached epidermis (mainly on thorax and trunk) on 30% (1st case) and 50% (2nd case) of BS, and typical petechial lesions (with characteristic "target" shape) diffused over the whole body surface (Fig. 1-2).

In both patients the complete therapy protocol of Lyell's syndrome (as above explained) was followed. For plasmapheresis a continuous flow cellular separator (Dideco Excel) was employed.



Fig. 1. First clinical case (bulbous lesions of the skin (flittena), particularly concentrated around the mucosal areas).



Fig. 2. First clinical case (dermo-epidermal detachment interesting at least the 30% of the body surface).



Fig. 3. Second clinical case.



Fig. 4. Second clinical case

Results

Both patients positively reacted to the treatment, the skin wounds disappeared (within the third and the sixth day of treatment) while the mucosal wound healed within the tenth day. Both patients survived and were dismissed after 15 and 20 days, with complete restore of all the life functions (Fig. 3-4).

Conclusions

It is our concerted opinion that this new therapy protocol for Lyell's syndrome has helped us in the control of a so unclear clinical situation, obtaining a satisfactory healing of the lesion (both skin + mucosa) in a remarkable short time, and quickly re-establishing a satisfactory degree of comfort for the patient. The survival percentage (even if in a small number of cases) is far better compared with standard therapies. Lissia percentages (6 survivals on 7 treated cases) confirm this evidence⁹.

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