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Pemphigus Vegetans: Diffrent Clinical Presentations

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Abstract

Vegetative pemphigus is a rare variety of deep pemphigus. Diagnosis is based on clinical, immunological, and histopathological patterns. Its occurrence during pregnancy is rare. We report 02 cases of vegetative pemphigus including one in pregnancy. It consisted of two women, aged 29 and 40 of old, respectively. One of them was 23 weeks pregnant. Both underwent vegetative lesions located at the large folds and in one of them, there were more than a hundred pustules next to the bullae. Nail damage was present in both patients and non-scarring alopecia. Histopathology was in favor of deep pemphigus. A Hallopeau-like form was retained in one of the patients, contrasting with mucosal involvement. Treatment with corticosteroids was associated with Dapsone in one case due to corticosteroid resistance. Regarding the pregnant woman, the evolution was favorable with a cesarean delivery at 32 weeks + 2 days, of a premature newborn free of any skin lesion. Recurrences were triggered by infections: bacterial dermohypodermitis and vulvovaginitis.

Keywords : Pemphigus, Vegetans, Pregnancy, Dapsone

Introduction: Pemphigus vegetans is a clinical form of pemphigus vulgaris. It is characterized by the vegetative evolution of the lesions and by their arrangement opposite the large folds. It can present in two forms: vegetative pemphigus of the Neumann type and vegetative pemphigus of the Hallopeau type. The diagnosis is set by the combination of several findings: clinical presentation, histological patterns, and immunological examinations. Treatment is based on corticosteroid therapy. The evolution is generally favorable. However, recurrences are possible. We report in this paper 02 cases of vegetative pemphigus, collected in the department of dermatology and venereology of the institute of social hygiene in Dakar, Senegal.

Patients and Observations

Observation 1

This is a 29-year-old patient, with a history of pemphigus vulgaris in 2015 and lost to follow-up for 6 years, who had consulted our department again for treatment of vegetative lesions located in the major folds as well as only at the level of the hands and the feet, concomitant with an attack of the oral mucosa the whole evolving for 3 months. The dermatological examination had objectified the presence of multiple well-defined smelly moist vegetative patches, with irregular edges with a predominant cauliflower appearance at the level of the axillary folds, inguinal folds overflowing on the perineum with nail-like erosions in places (Fig.1: A - B)

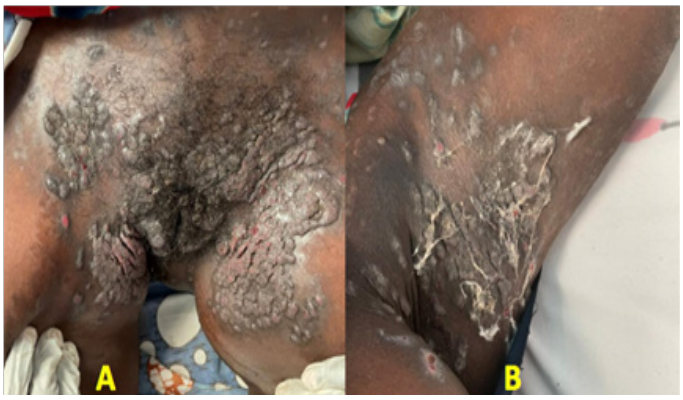
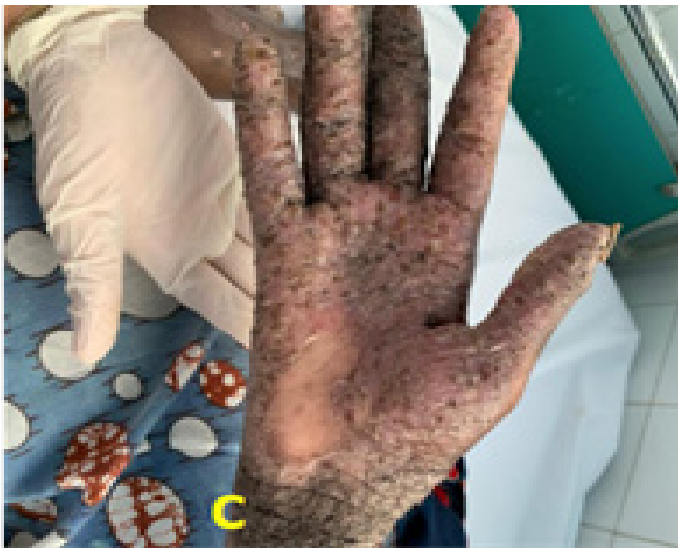


Figure 1.(A-B) : Cauliflower appearance at the axillary and inguinal folds overflowing on the perineum with nail-like erosions in places

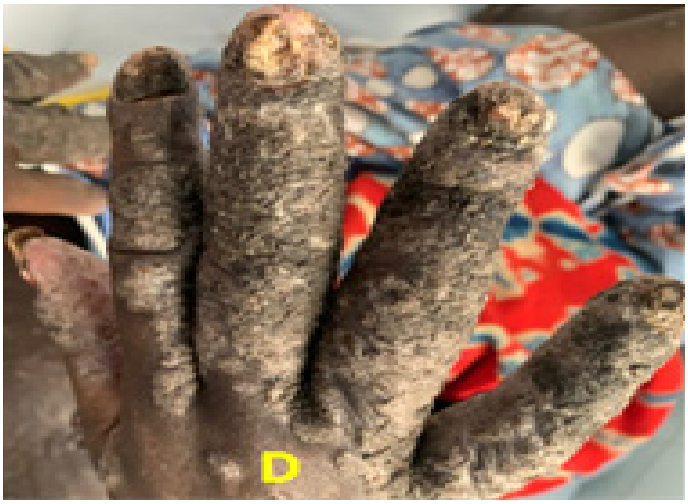
as well as lesions at the palmoplantar level a type of punctate keratosis, crusty and desquamative lesions in places on the palmar level **(Fig.1:C)**

C) Palmoplantar lesions with punctate keratosis, crusty and desquamative lesions in places on the palmar level.



and oozing hyperkeratotic plaques in places on the back hands **(Fig.1: D)**

D) Oozing hyperkeratotic plaques in places on the backhands.



and feet **Fig.2 (A-B)**,

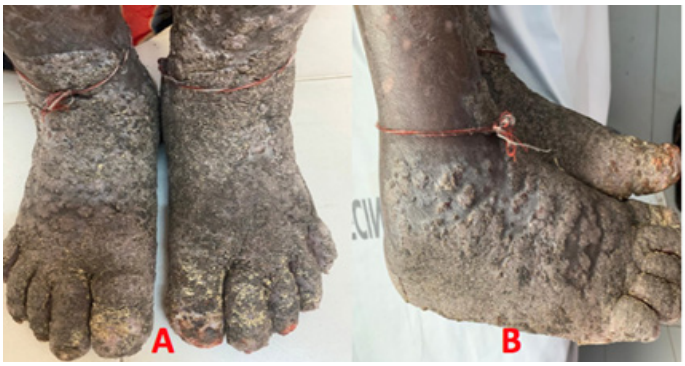


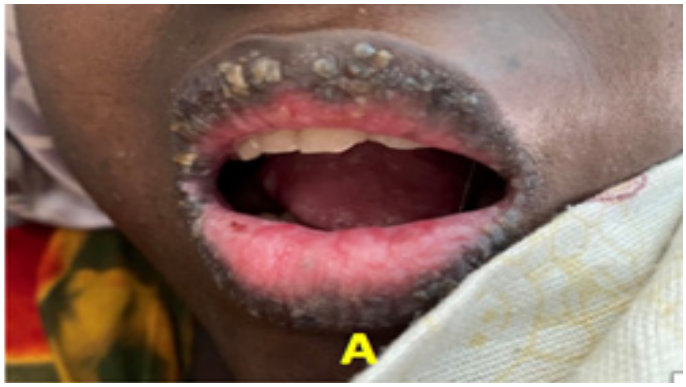
Figure 2.(A-B) : Hyperkeratosic plaques on the feet.

Diffuse nail involvement such as onychoschyzia, nail dystrophy, paronychia and subungual hyperkeratosis (Fig.2 C-D).

Fig.2.(C-D) : onychoschyzia, nail dystrophy, paronychia, and subungual hyperkeratosis

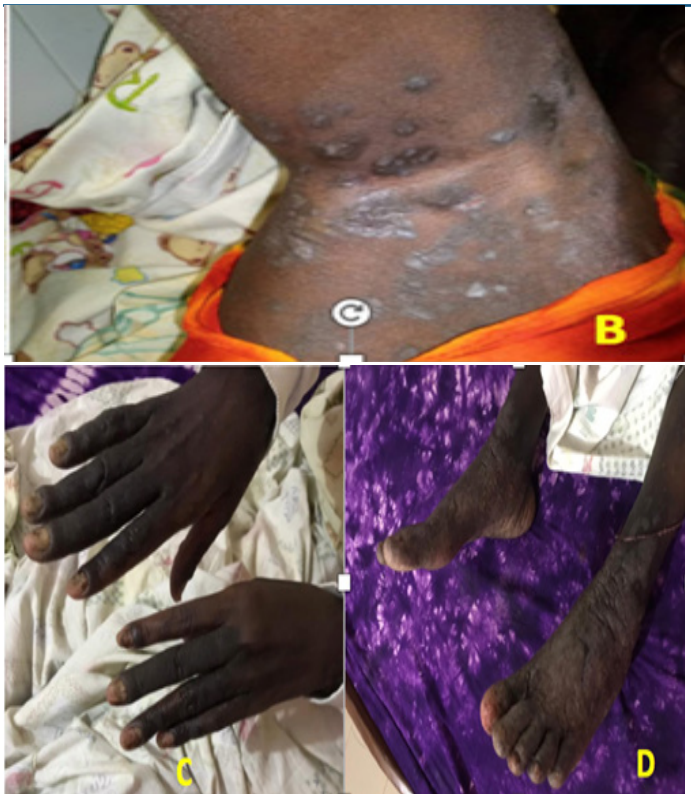
The patient also presented with biangular cheilitis and endobuccal erosions **Fig.3 (A)**.

Figure 3.(A) : Biangular cheilitis and endobuccal erosions



The picture evolved in a context of a 4-month amenorrhea and a deterioration of the general condition stage II of the WHO. A skin biopsy was performed which confirmed the diagnosis of vegetative pemphigus with the presence of herpetic superinfection and an abdominopelvic ultrasound revealed an evolving single pregnancy estimated at 19 weeks. The patient had been put on oral corticosteroid therapy at a dose of 0.75 mg/kg/day, antiviral and local care with obstetric monitoring. Around the 7th month of pregnancy, the patient was on 0.25mg/kg/d of prednisone and she showed regression of the hypertrophic plaques leaving residual post-inflammatory pigmentation **Fig.3(D-B-C)**. The evolution was therefore favorable with almost complete healing, after which the patient was again lost sight of.

(B-C-D) : Regression of the hypertrophic plaques and residual post-inflammatory pigmentatio



She would have delivered by caesarean section at 32 SA +2 days of a premature newborn of 2200 g and which would be free of any skin lesion.

Observation 2

This is a 40-year-old nulligest and nulliparous patient who had consulted in our department with diffuse pruritic bullous eruptions and painful oral and genital lesions. The dermatological examination had objectified flaccid bullae with positive Nikolsky's sign, large post-bullous erosions **(Fig.4 : A-B)**,

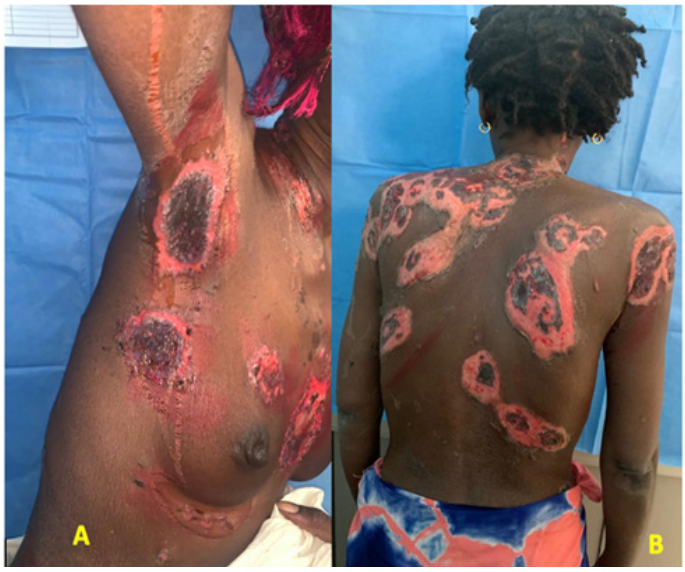
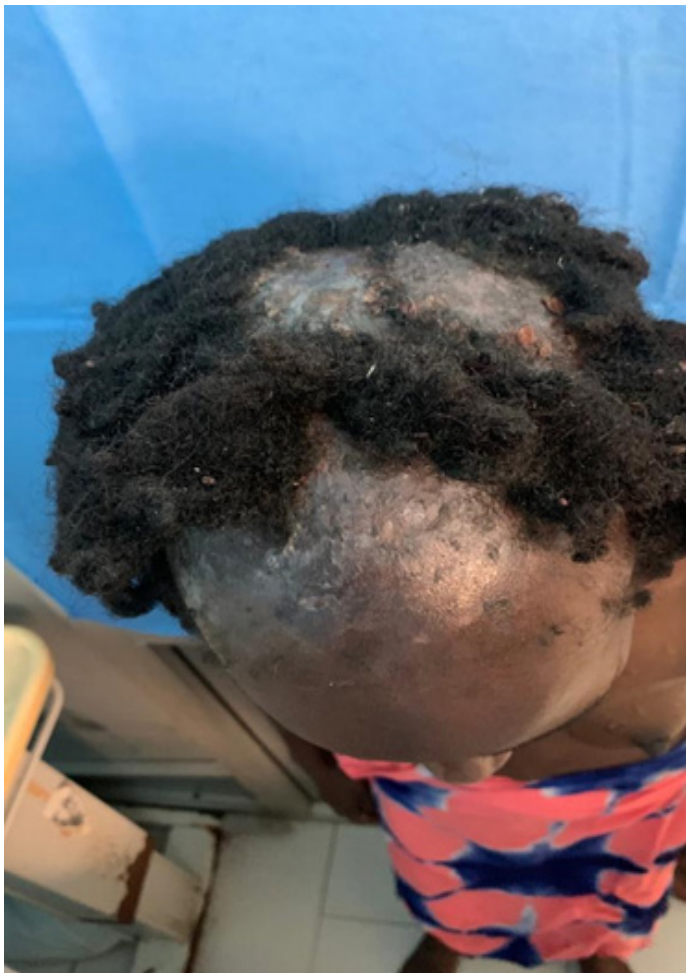


Figure 4.(A-B) : Large post-bullous erosions

Purulent oral and genital erosions, a crusty scaly helmet of the scalp purulent and alopecic in places with a nauseating odor (Fig.5).

Figure 5 : Crusty scaly helmet of the scalp purulent and alopecic in places



The picture evolved in the context of a 3-month amenorrhea with the presence of fetid whitish leucorrhoea. A skin biopsy was performed, which confirmed the diagnosis of vegetative pemphigus with antinuclear antibodies less than 10. An abdominopelvic ultrasound was performed, which returned to normal, and a vaginal swab revealed gardenella vaginalis vaginosis. The patient had been put on oral corticosteroid therapy at a dose of 1mg/kg/day, metronidazole 1g in a single dose, and local care. We have the appearance of new bubbles 1 week later despite well-conducted treatment, hence an association with dapsone at a rate of 100mg/d.

Good evolution after two months of treatment and complete healing of the lesions leaving diffuse post-inflammatory hyperpigmentation (Fig 6 : A-B).

Figure 6.(A-B) : **Post-inflammatory hyperpigmentation**

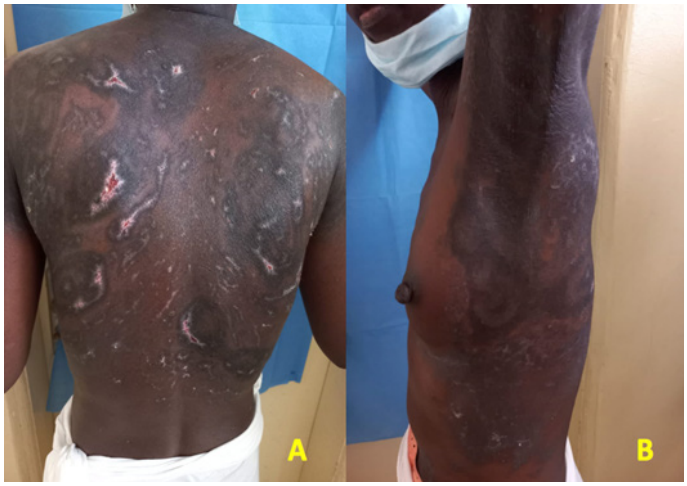


Fig 6.(C-D): **Eruptions pustular on the neck and back**

After 4 months, despite well-conducted treatment, there were recurrences with the appearance of new bullous eruptions with purulent contents in the submammary and axillary folds, trunk and periumbilical region with oozing erosions in places Fig 7: (A-B), eruptions pustular on the neck and back Fig 7: (C-D),

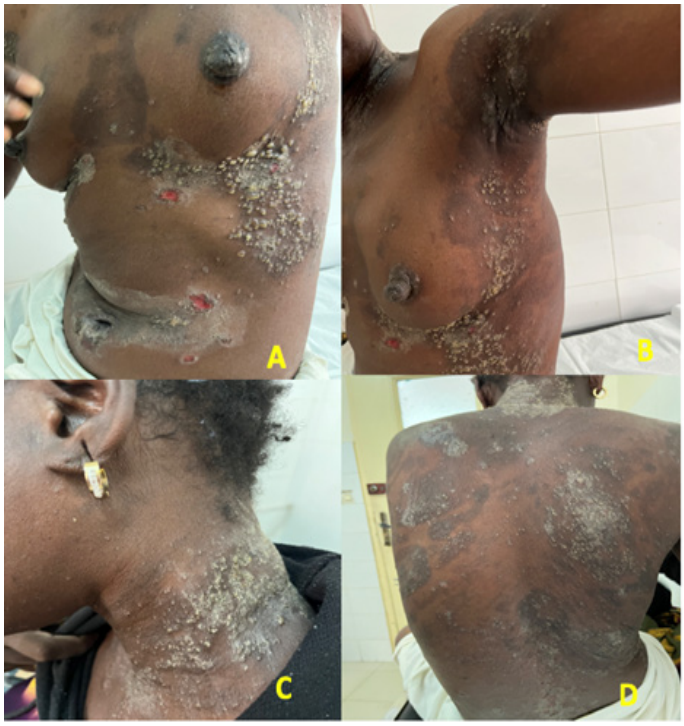


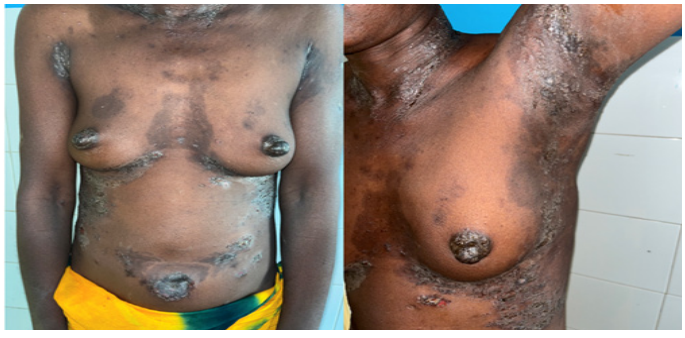
Figure 7.A-B) : Bullous eruptions with purulent content in the submammary and axillary folds, trunk, and periumbilical region with oozing erosions in places

C-D): Eruptions pustular on the neck and back

A crusty erythematous patch with irregular contours on the right leg painful with local heat and a right OMI going up to the ankle taking the pit (Fig. 8: AT). She also had erosions of the oral mucosa (Fig.8 : B), crusty scaly helmet of the scalp with alopecia at the edge (Fig.8 : C) and nail involvement such as onycholysis and paronychia of the left index finger and right middle finger (Fig.8 : D),

fetid whitish leucorrhoea with vaginal pruritus. We retained the diagnosis of a new outbreak triggered by a genital infection confirmed by vaginal swab with search for mycoplasma and positive chlamydia complicated by dermo-hypodermitis of the right lower limb. The clinical appearance of the lesions was in favor of vegetative Hallopeau-type pemphigus.

She received treatment with dapsone 100 mg/d, oral corticosteroid therapy 0.5 mg/kg/d, amoxicillin clavulanic acid 3g/d, doxycycline 200mg/d and daily care. The evolution after three days was marked by scarring of the pustules in the form of vegetative plaques (Fig 9). For patient monitoring, close follow-up with G6PD assay was recommended.



Discussion

Vegetative pemphigus represents only 1 to 2% of pemphigus cases [1]. This was consistent with our study, over a period of 11 years, only two cases had been found. It is a clinical form of pemphigus vulgaris characterized by the vegetative evolution of the lesions and by their location next to the large folds [2,3]. with a female to male ratio of 1.3 to 2.3 to 1. Age at diagnosis is typically 50 to 70 in the United States and European countries, and 30 to 50 in other countries [4,5].

There are two clinically recognized forms of vegetative pemphigus, the Hallopeau type and the Neumann type. The Hallopeau type has an indolent course and characteristically presents pustules which heal in the form of vegetative plaques. The oral mucosa is often not affected contrary to what was found in our patient. The Neumann type is more severe and refractory to treatment, with vegetations developing during an eruption of vesiculobullous lesions [6].

Skin lesions rupture and ulcerate, and warty, crusty vegetative patches form over the erosions. These hyperkeratotic lesions characteristically present in the intertriginous areas, including the inguinal/inguinal folds, armpits, thighs, and flexor surfaces [7].

Two German studies had questioned the subclassification of vegetative pemphigus into Hallopeau and Nemann subtypes, finding characteristics of the two subvariants observed during the disease [8,9].

Its diagnosis is confirmed by the histological examination which shows a supra-basal bullous intraepidermal detachment, acantholysis without keratinocyte necrosis, edematous dermis and site of a lymphocyte infiltrate with eosinophils and the presence of pustules. Direct immunofluorescence shows deposits of IgG and C3 in “net mesh” in the epidermis. The pathophysiology of vegetative pemphigus is poorly understood [3].

A triggering factor is identified in several cases, such as drugs, infections or malignancies [10]. A relapse of pemphigus can most often be observed following an untimely treatment break, herpetic superinfection, herbal medicine, hormones or the tapering off of corticosteroid therapy [11]. In our observation, Chlamydia and Mycoplasma infection were found as triggering factors for relapses, while in other studies the most common infections were herpes infection and bacterial infection due to staphylococcus aureus [12,13].

The occurrence of pemphigus during pregnancy is rarely reported, about fifty cases are described in the medical literature [14].

The disease can occur at any stage of pregnancy or postpartum. Most reported cases of pemphigus during pregnancy correspond to pemphigus in its vulgar or foliaceous form [15,16].

Regarding treatment, topical and/or oral corticosteroids are recommended as first-line treatment in current consensus documents especially in pemphigus vulgaris, which can be extended to the vegetative pemphigus subgroup [17,18]. It is widely accepted that their use has significantly reduced morbidity and mortality in these patients [19].

Patients with Hallopeau-type pemphigus often have relatively mild disease, requiring lower doses of systemic corticosteroids to control mucocutaneous manifestations, and usually have prolonged remission [20].

As in our case, several retrospective studies that included a small number of patients with pemphigus vulgaris suggested the efficacy of dapsone as an adjuvant treatment associated with corticosteroid therapy and, in some cases, immunosuppressive therapy.

The cortisone-sparing effect of dapsone is therefore often suggested but not demonstrated in studies. Moreover, the real effectiveness of this treatment is difficult to assess because dapsone is often combined with other treatments (dermocorticoids, corticosteroids, immunosuppressants, etc.). The doses of dapsone used vary from one study to another (25 to 200 mg/d) and side effects are frequent at high doses (hepatitis, haemolysis) [20].

Concerning our patient in pregnancy, only prematurity was found as complications, in other studies we found worsening of symptoms and exacerbation of lesions in women with known pemphigus during pregnancy [21]. Fetal death can occur in 5 to 12% of cases, thus justifying increased fetal surveillance [15-21] and close collaboration between dermatologists and obstetricians [22].

Other complications such as prematurity and abortion can be observed [23]. The evolution of the lesions is spontaneously favorable after 3 to 4 weeks with local care. No case of recurrence or evolution towards pemphigus in adulthood has been reported to date [24]. Nevertheless, the incidence of neonatal pemphigus during the pregnancy of a mother with pemphigus vulgaris is 30 to 45% [14]. There is no established correlation between the maternal antibody titer of pemphigus vulgaris and the presentation

of the disease in the neonate [25,26]. This is supported by cases of neonatal pemphigus from mothers without active disease and cases of healthy newborns born to mothers with highly active disease [25,27].

During pemphigus vegetans infections can be found secondary to the lesions: Staphylococcus aureus, herpes and fungal organisms are the most common. There may be an increased susceptibility to systemic infections due to chronic use of immunosuppressive treatments. But also malnutrition due to damage to the oral mucosa causing pain, reduced oral intake and weight loss [28].

Autoimmunity is not curable, but rather only treated and controlled. If left untreated, pemphigus, including vegetative pemphigus, can be fatal within 5 years due to severe blistering, secondary infection, and malnutrition. Mortality is about 5 to 15% per year [29].

Conclusion

Pemphigus vegetans is a clinical form of pemphigus vulgaris, there would be a coexistence between the two subtype hallopeau and Neumann. Its occurrence in pregnancy is rare with a risk of prematurity. The treatment is generally based on corticosteroid therapy and an adjuvant treatment based on dapsone is effective in the event of corticosteroid resistance. The evolution is generally favorable but there is the possibility of recurrences triggered by infections

References

1. A Abdou, N E l Moussaoui, F Z Lamchahab, K Senouci, B Hassam (2013) Étude épidémioclinique d'une forme rare de pemphigus : pemphigus végétant au CHU Ibn Sina de Rabat Service de dermatologie et vénéréologie, CHU Ibn Sina, Rabat, Maroc : P263.
2. Zaraa I, Sellami A, Bouguerra C, Sellami MK, Chelly I, et al (2011) Pemphigus vegetans: à clinical, histological, immunopathological and prognostic study: pemphigus vegetans. J Eur Acad Dermatol Venereol 25 : 1160-1167.
3. Dhamija A, Kothiwala R, Meherda A, D'souza P (2012) Pemphigus vegetans: an unusual presentation. Indian Dermatol Online J 3 : 193-195.
4. Langan SM, Smeeth L, Hubbard R, Fleming KM, Smith CJ, et al (2008) Pemphigoïde bulleuse et pemphigus vulgaire - incidence et mortalité au Royaume-Uni : étude de cohorte basée sur la population. BMJ. 09 juillet 337 : a180.
5. Min MS, Damstetter E, Chen AYY (2018) Troubles bulleux auto-immuns dans le cadre d'une infection par le virus de l'immunodéficience humaine. Int J Dermatol pour femmes. 2018 septembre 4 :159-165.
6. Jain V, Jindal N, Imchen S (2014) Vegetans pemphigus localisés sans implication des muqueuses. Indien J Dermatol. 2014 mars 59 : 210.
7. Cozzani E, Christana K, Mastrogiacomo A, Rampini P, Drosera M,et al (2007) Pemphigus vegetans Type Neumann avec auto-anticorps anti-desmogléine et anti-périplakine. Eur J Dermatol. 2007 novembre-décembre 17 : 530-533.
8. Jansen T, Messer G, Meurer M, Plewig G (2001) Pemphigus végétaliens. Une perspective historique. Hautarzt 2001 juin 52 : 504-509.
9. B Monshi, M Marker, H Feichtinger (2010) JDDG : Journal de 2010 - Wiley Online Library
10. Ragragui Ouasmin H, Saddouk H, Sof K, Dikhaye S, Zizi N.Pemphigus végétant de type HALLOPEAU : Une présentation clinique inhabituelle d'une dermatose rare Service de Dermatologie, Université Mohammed V de Rabat, Faculté de Médecine et de Pharmacie, Rabat, Maroc.
11. Diallo M, Diatta B A, Diop A, Ndiaye M, Ndiaye M T, et al (2017) Epidemiological and clinical aspects of pemphigus epidemiological and clinical aspects of pemphigus in Senegal.Department of Dermatology, Aristide Ledantec University Hospital of Dakar, Senegal Corresponding author Prof. Moussa Di 8: 5-9.
12. MA Tufano , A Baroni , E. Buommino , E Ruocco, ML Lombardi, et al (1999) Détection de l'ADN du virus de l'herpès dans les cellules mononucléaires du sang périphérique et les lésions cutanées des patients atteints de pemphigus par réaction en chaîne par polymérase .British Journal of Dermatology 141 : 1033-103.
13. Amagai M, Yamaguchi T, Hanakawa Y, Nishifuji K, Sugai M, et al (2002) Staphylococcal exfoliative toxin B specifically cleaves desmoglein 1. J. Invest Dermatol 118 : 845-50.
14. Carvalho AA, Santos Neto DAD, Carvalho MADR, Eleutério SJP, Xavier AREO, et al (2019) Neonatal pemphigus in an infant born to a mother with pemphigus vulgaris : a case report. Rev Paul Pediatr 37 :130-134.
15. Kardos M, Levine D, Gürcan HM, Ahmed RA (2009) Pemphigus vulgaris in pregnancy: analysis of current data on the management and outcomes. Obstet Gynecol Surv 64 :739-749.
16. Galarza C, Gutiérrez EL, Ramos W, Tello M, Ronceros G, et al (2009) Endemic pemphigus foliaceus in a pregnant woman. Report of one case. Rev Med Chil 137 :1205-1208.
17. Harman KE, Albert S, Black MM (2003) Guidelines for the management of pemphigus vulgaris. Br J Dermatol. 149 :

926- 937.

18. Joly P, Bernard P, Bedane C, Prost C, Ingen-Housz-Oro S (2011) Recommandations des centres de référence des maladies bulleuses auto-immunes pour le diagnostic et la prise en charge du pemphigus. Ann Dermatol Vénéréologie 138 :252- 258.
19. Almeida HL Jr, Neugebauer MG, Guarenti IM, Aoki V (2006) Pemphigus vegetans associated with verrucous lesions: Expanding a phenotype. Clinics (Sao Paulo) 61 : 279-82.
20. Pascal Joly, Noémie Litrowski (2011) Carole Sin.Pemphigus: revue de la litterature 29: 432-436.
21. Clinique Dermatologique, Hopital Charles Nicolle, Université de Rouen, Rouen, France.
22. Prendiville JS, Israel DM, Wood WS, Dimmick JE (1994) Oral pemphigus vulgaris associated with inflammatory bowel disease and herpetic gingivostomatitis in an 11-year-old girl. Pediatric dermatology 11 :145-150.
23. Daneshpazhooh M, Chams Davatchi C, Valikhani M, Aghabagheri A, Mortazavizadeh SM, et al (2011) Pemphigus and pregnancy: à 23-year experience. Indian J Dermatol Venereol Leprol. 77 : 534.
24. Gushi M, Yamamoto Y, Mines Y, Awazawa R, Nonaka K, et al (2008) Neonatal pemphigus vulgaris. J Dermatol 35 :529-535.
25. Fainaru O, Mashiach R, Kupferminc M, Shenhav M, Pauzner D, et al (2000) Pemphigus vulgaris in pregnancy: a case report and review of lithe literature. Hum Reprod 15 :1195-1197.
26. Panko J, Florell SR, Hadley J, Zone J, Leiferman K, et al (2009) Neonatal pemphigus in an infant born to a mother with serologic evidence of both pemphigus vulgaris and gestational pemphigoid. J Am Acad Dermatol 60 : 1057-1062.
27. Amer YB, Al Ajroush W (2007) Pemphigus vulgaris in a neonate. Ann Saudi Med. 27 :453-455.
28. Kalayciyan A, Engin B, Serdaroglu S, Mat C, Aydemir EH, et al (2002) A retrospective analysis of patients with pemphigus vulgaris associated with pregnancy. Br J Dermatol 147:396-397.
29. Kavala M, Topaloğlu Demir F, Zindanci I, Can B (2015) Atteinte génitale dans le pemphigus vulgaire (PV) : corrélation avec les résultats cliniques et cervicovaginaux des frottis vaginaux. J Am Acad Dermatol 73 : 655-659.
30. Ellebrecht CT, Payne AS (2017) Fixer la cible pour le traitement du pemphigus vulgaire. Aperçu JCI. 09 mars 2 : e92021.