Case Report

# Pityriasis lichenoides et varioliformis acuta in pregnancy

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## ABSTRACT

Pityriasis lichenoides et varioliformis acuta is an uncommon disease, especially during pregnancy. In review of the obstetric literature, there was no report of pityriasis lichenoides et varioliformis acuta during pregnancy. A 25-year-old female was seen at 24 weeks of gestation for consultation about a cutaneous disease. She was admitted at 30 weeks of gestation because of threatened premature labor, and some active cutaneous papules presented themselves at that time. After the treatment, cutaneous papules remitted. But at 35 weeks of gestation, she had spontaneous labor. Both the mother and infant were doing well at 5 months postpartum. If pityriasis lichenoides et varioliformis acuta exists in the vagina or cervical bone of the uterus, it is due to infections from lymphatic vasculitis and necrosis. It may cause threatened premature labor and premature rupture of the membrane.

**Keywords:** Pityriasis lichenoides et varioliformis acuta, pregnancy, preterm labor.

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Cutaneous lesions of pityriasis lichenoides et varioliformis acuta (PLEVA), a T cell-mediated cutaneous inflammatory condition, are clinically similar to lymphomatoid papulosis (LyP), leading some authors to hypothesize that they are part of the same spectrum of lymphoproliferative disorders, although reports of the development of cutaneous lymphoma in patients with PLEVA are not as frequent as they are for patients with LyP. Furthermore, unlike in cases of LyP, no systematic search for dominant T-cell clones has been carried out in cases of PLEVA, whereas clones have been detected in a few cases of PLEVA using mainly Southern blot analysis.[1]

Pityriasis lichenoides et varioliformis acuta is a cutaneous disease characterized by multiple small guttate, erythematosquamous nonconfluent lesions. On rare occasions, patients may present with high fever, malaise and ulceronecrotic lesions. The disease was first described by Mucha[2] in 1916, and Habermann[3] used the acronym PLEVA. So it is popularly called Mucha-Habermann disease. Although it may occasionally progress to chronic pityriasis lichenoides, its transformation into a lymphoma is not to be anticipated.

Pityriasis lichenoides et varioliformis acuta has been reported in different races and develops more frequently in young people and males. In a review of the obstetric literature, we found no report of PLEVA during pregnancy. Therefore we report a patient who was diagnosed as a case of PLEVA during the pregnancy and had a preterm delivery.

**Case Report.** The patient is a 28-year-old female, gravida 3 para 2 with a history of one neonatal death with unknown origin and one preterm labor at 34 weeks of gestation after premature rupture of the membranes. She was sent at 24 weeks of gestation for consultation about a cutaneous disease. The patient was in a good health until 24 weeks of gestation when she had symptomatic

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nonconfluent eruption of multiple lesions, evenly distributed on the trunk, face and limbs, and sometimes abrupt onset of an eruption with multiple disseminated, varicelliform cutaneous lesions accompanied by mild fever. At that time she was diagnosed as having PLEVA. Although she was treated with hydrocortisone ointments, lesions repeated relapse and remission.

At 24 weeks of gestation, PLEVA was well controlled with no pregnancy complications and the fetal growth was appropriate, she was admitted at 30 weeks of gestation due to threatened premature labor and was treated by infusion of litodorine chloride and magnesium sulfate, some cutaneous papules presented a purpuric character and were surrounded by an inflammatory halo, the center was vesiculopustular and then necrotic, and covered by dark crusted lesions (Figure 1).

The pathology of those papules showed diffuse extravasation of lymphocyte and erythrocyte. The epidermis showed intracellular edema with vasculitis (Figure 2). Laboratory examination shows the white blood cell count of $12.8X10^3$/UL and C-reactive protein 1.0 mg/dl, she was treated with benzyl penicillin sodium. On the 10th day of hospitalization, cutaneous papules remitted and the white blood cell count and C-reactive protein were within normal range.

But after stopping litodorine chloride and magnesium sulfate, spontaneous labor at 35 weeks of gestation produced a vigorous 1800 gm female infant. Apgar score was 9 at one and 5 minutes.

The pathology of the placenta showed no vasculitis and chorioamnitis. Both the mother and the infant were doing fine at 5 months without remission of maternal cutaneous papules.

**Discussion.** In 1916 Mucha and in 1925 Habermann reported an acute form of pityriasis lichenoides characterized by the abrupt onset of papulovesicular eruptions and gave the name, Pityriasis Lichenoides et Varioliformis Acuta (PLEVA) or Mucha-Habermann disease (MH). In 1966, Degos reported a rare febrile ulceronecrotic variant of MH. MH occurs mainly in young adults, while febrile ulceronecrotic Mucha-Habermann's disease (FUMHD) occurs more frequently in children. The etiology of MH remains obscure, but it may be the result of a hypersensitivity reaction to an infectious agent.

Pityriasis has been reported more frequently inyoung people and males, being a less common form in which there is the abrupt onset of an eruptin characterized by multiple disseminated, varicelliform cutaneous lesions, sometimes accompanied by mild fever, malaise, and headache during the first 2 or 3 days. Cutaneous papules presented a purpuic character and are surrounded by an inflammatory halo, the center is vesiculopustular, then necrotic and covered by a dark adherent curst. Lesions may be accompanied by a burning or itching sensation. They are disseminated on the trunk, limbs, palms, soles, face, scalp, and mucous membranes, particularly on the flexural parts.

Usually PLEVA is a self-limited process that resolves in a few weeks or months. However, the appearance of lesions in successive outbreaks determines the polymorphous aspect characteristic of acute and chronic forms of the disease. In our case, cutaneous lesions were disseminated on the trunk and limbs but not in other portions, and were of the polymorphous type. Acute phase appeared at 30 weeks of gestation in this pregnancy. After 10 days of treatment acute cutaneous lesions remitted and progressed to chronic pityriasis lichenoides.

The etiology is unknown, however the sporadic appearance suggests the possibility of a viral or bacterial infection, allergic and toxic cause. No report of PLEVA during pregnancy was found. It is

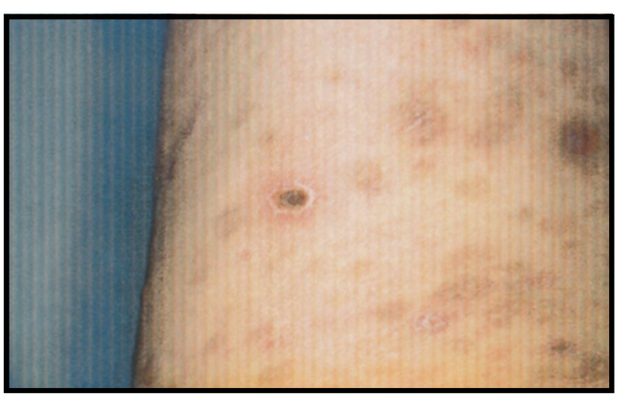

**Figure 1** - Erythematous papules and crusted ulcerative lesions are seen in the femur.

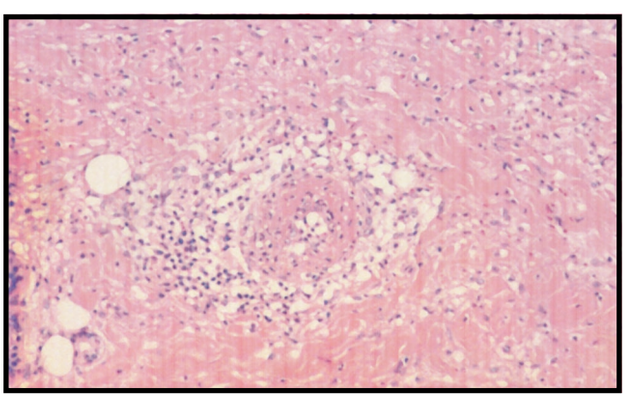

**Figure 2 -** Diffuse extravasion of lymphocyte and erythrocyte, the epidermis shows intracellular edema (Hematoxylin-eosin staining methods).

uncertain whether the pregnancy may affect PLEVA, or PLEVA may affect the pregnancy.

As for the histopathologic findings of PLEVA, there is a dense inflammatory infiltrate, and fibrinoid necrosis may be present in addition to sever lymphocytic perivasculitis and vasculitis of the small dermal blood vessels.[4] The epidermis presents sever alterations with intracellular edema, degeneration and necrosis.

Exocytosis, with the presence of many lymphocytes and erythrocytes in the epidermis, is a constant feature. Dermal capillaries are dilated, and sometimes tumefaction and proliferation of endothelial cells are found. Extra vasation of erythrocytes and penetration of inflammatory cells into the walls of blood vessels are often visible.[5]

Biopsy specimens from lesions of patients with PLEVA were studied by direct immunofluorescence and immunoperosidase technics[6] and showed there is slight vascular deposits of IgM and C3 were present in most lesions. Slight perivascular deposits of fibrin were observed in early lesions, more extensive perivascular interstitial deposits of fibrin were detected in advanced lesions. Most of the infiltrating cells were T lymphocytes; cells with cytotoxic/ suppressor phenotype (T8-positive) were generally more numerous increase in epidermal T8-positive cells over epidermal Leu-3a/T4 positive cells was found in late lesions.

In our case, macrospically PLEVA presented only as cutaneous lesions, not mucosal lesions, for instance in the vagina and the cervical bone. Therefore it is uncertain that PLEVA itself causes the threatened premature labor and premature rupture of the membrane. But after infusion of antibiotics, acute cutaneous lesions remitted, threatened premature labor did not progress, and the white blood cell count and C reactive protein reached the normal range. These facts may suggest that PLEVA latently existed in the vagina or the cervical bone and developed infections from lymphatic vasculitis and necrosis. Our case demonstrated that PLEVA in pregnancy might cause threatened premature labor and premature rupture of membrane.

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