The Past, Present, and Future of Pemphigus Syphiliticus

J. Pérez-Pelegay,[a] M.P. Sánchez-Salas,[a] P. Ventura-Faci,[b] M.P. Grasa-Jordán,[a] and F.J. Carapeto[a]

[a]Departamento de Dermatología and [b]Departamento de Pediatría, Hospital Clínico Universitario, Lozano Blesa, Zaragoza, Spain

To the Editor:

Neonatal syphilis is a one of the most serious infections transmitted from mother to fetus.

The World Health Organization estimates that maternal syphilis is responsible worldwide for 460 000 stillbirths or miscarriages, 270 000 cases of congenital syphilis, and 270 000 of underweight or premature newborns.

Transmission occurs after uteroplacental circulation is established; however, the manifestations do not appear until the fourth month and consist of abnormal organogenesis and immunologic abnormalities, often leading to late-term abortion.

We describe an infant born at 30 weeks of gestation, the first common child of a young couple (Spanish father, Romanian mother). A single prenatal visit had been performed, although our hospital had no record of this visit.

The examination revealed an infant with a noteworthy clinical picture at birth, including perceptible abdominal distention on palpation, which also showed considerable liver and spleen enlargement. The most striking observation, however, were large bullous lesions that varied in size between 1 and 5 cm and occupied virtually the entire surface of the palms and soles (Figure 1), as well as other isolated lesions of creamy-white content on the legs and forearms. Further examination showed that the skin was extremely fragile, particularly on the left foot and the third toe of the right foot, and peeled off like the finger of a glove (Figure 2).

The rest of the body was covered with erythematous-desquamative, macular, papular lesions, some of which were actually crusty. The lesions were mainly located on the head between the eyebrows, perinasal, and circumoral region, as well as on the trunk, buttocks, and perianal area (Figure 3). Spoke-like cicatricial atrophic striae could also be seen in the circumoral and perianal area.

Because perinatal infection (congenital syphilis) was suspected, the child was transferred to the neonatal intensive care unit, but died 3 hours later.

Serologic results received 2 days later confirmed the diagnosis. In the case of the newborn, rapid plasma reagin (RPR) was 1/128, *Treponema pallidum* hemagglutination assay (TPHA) was 1/81 920, and anti-*Treponema pallidum* immunoglobulin (Ig) M, 2.9. In the mother, RPR was 1/32, TPHA was 1/81 920, absorption of fluorescent *Treponema* antibodies, 1/200, and anti-*Treponema pallidum* IgM, 1.519. All other serology tests were negative.

Congenital syphilis can occur if a woman with syphilis gets pregnant or if the mother is infected during pregnancy. According to Thomas,[1] the longer the duration of untreated maternal syphilis before pregnancy occurs, the lower the risk will be for the fetus. If the mother acquires the infection in an advanced stage of pregnancy, the newborn may be normal and the clinical manifestations may not appear until weeks or months later. Conversely, if infection occurs during early pregnancy, it can lead to abortion or serious symptoms in the infant.

Pemphigus syphiliticus is a rare condition in Spain, but is on the rise, particularly in developing countries and in Eastern Europe. Industrialized nations are being affected by population changes caused by immigration from underdeveloped countries, particularly from Central Africa, but also from Eastern Europe. These immigrants bring with them certain ideas and habits and some of these groups may find themselves in precarious employment situations.

Prevalences of 3% to 19% have been reported among pregnant women in developing countries; the highest figures are from southeast Africa and southern Sahara. In Zambia, 42% of fetal deaths have been attributed to syphilis and 30% of all perinatal deaths are associated with neonatal syphilis.[2]

Figure 1. Bullous lesions occupying the entire surface of the palm and soles.

Figure 2. Blisters that peeled off like the finger of a glove.

Figure 3. Erythematous and desquamative periorificial lesions.

In Spain, syphilis rates in adulthood increased by about 75% between 1999 and 2004; the total number of cases was 675 per 100 000 inhabitants in 1999 and 1156 in 2004. Cases of congenital syphilis have increased 700%, rising from 2 in 1999 to 16 in 2004, with 9 cases reported in 2000, 8 in 2001, 15 in 2002, and 4 in 2003.

In 2004 the number of reports has continued to climb, with 12 cases described in the province of Malaga alone.[3] In 2005, cases of neurosyphilis in Madrid[4] and malignant syphilis in Galicia[5] and Madrid[6] have been published. The latest study on an epidemiologic outbreak was conducted in Las Palmas de Gran Canaria, in the Canary Islands.[7]

Because of the recent rise of syphilis in Europe, closer control consisting of early screening and treatment of affected pregnant women is necessary, since neonatal syphilis can be prevented through education programs focused on sexually transmitted diseases and good prenatal care (compulsory Venereal Disease Research Laboratory screening in pregnant women).[8]

References

1. Berdasquera D, Fariñas AT, Ramos I. Las enfermedades de transmisión sexual en embarazadas un problema de salud a nivel mundial. Rev Cubana Med Gen Integr. 2001;17:185-90.
2. Fitzpatrick TB, Johnson RA, Wolff K. Color atlas and synopsis of clinical dermatology. 4th ed. Philadelphia: McGraw Hill Co; 2001. p. 889-904.
3. Barrera MV, Bosch RJ, Mendiola M, Frieyro M, Castillo R, Fernández A, et al. Reactivación de sífilis en Málaga. Actas Dermosifiliogr. 2006;97:323-6.
4. Quesada A, Campos L, Rubio C, Martín MA, Herranz P, Arribas JR, et al. Tres casos de neurosífilis precoz en pacientes infectados por VIH. Actas Dermosifiliogr. 2006;97:395-9.
5. Pérez Pérez L, Cabanillas M, Ginarte M, Sánchez Aguilar D, Toribio J. Sífilis maligna en un paciente con infección VIH. Actas Dermosifiligr. 2007; 98:351-4.
6. Fernández Guarino M, Aldanondo Fernández de la Mora I, González García C, Harto Castaño A, Moreno Izquierdo R, Jaén Olasolo P. Sífilis maligna en pacientes con virus de la inmunodeficiencia humana. Actas Dermosifiliogr. 2006;97:400-3.
7. Vilar J, Dehesa L, Gómez Duaso AJ, Bastida J, Rivero P, Domínguez Silva J, et al. Estudio epidemiológico de un brote de sífilis en Las Palmas de Gran Canaria. Actas Dermosifiliogr. 2007; 98:466-9.
8. Grossman KL, Rasmussen JE. Recent advances in pediatric infectious disease and their impact on dermatology. J Am Acad Dermatol. 1991;24:379-89.

Classic Kaposi Sarcoma Associated With Lymphedema Following Arterial Catheterization

D. Barco, M. Alegre, and A. Alomar

Servicio de Dermatología, Hospital de Sant Pau, Barcelona, Spain

To the Editor:

Classic Kaposi sarcoma (CKS) is a vascular neoproliferation typically seen on the lower limbs of elderly patients. The condition is associated with human herpes virus 8 (HHV8); however, its high prevalence in Mediterranean countries suggests that other environmental factors may be relevant in its etiology.

We describe a 59-year-old man with asymptomatic violaceous plaques and nodules present from 1 year earlier on the right leg (Figure 1). The lymphedema observed had been present since a femoral artery catheterization performed 6 years earlier for intestinal bleeding. The histologic study of the lesion showed vascular proliferation of fusiform cells with erythrocytes dissecting the collagen bundles, consistent with Kaposi sarcoma (KS). The immunohistochemical study was positive for HHV8. Contrast-enhanced magnetic resonance angiography showed no vascular abnormalities or arteriovenous fistulas in the lower limbs. Ten cycles of liposomal doxorubicin of 20 mg/m^2/3 wk were administered, and complete clinical remission was achieved (Figure 2).

The classic variant of KS is characterized by violaceous papules or

Figure 1. Onset of violaceous papules and nodules. Note the edema of the affected limb.

Figure 2. Clinical appearance of the lesions after treatment.