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The First Article in The Journal *Actas Dermo-Sifiliográficas* Published With Photographs: Malignant Keratosis Diffusa Fetalis (Fetal Ichthyosis; Congenital Maligna Keratoma, Etc.), by Juan de Azúa

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Hiperkeratosis universal congénita maligna. (Ictiosis fetal; keratoma maligno congénito, etc.).

Por Juan de Azúa.

El caso que presento, pertenece á una especie morbosa, mal conocida y delimitada, que ha recibido múltiples nombres. Su aspecto, semejante á la *Ictiosis*, y la predominancia en ella de alteraciones de cornificación, han servido para denominarla *Ictiosis congénita*, *Ictiosis fetal congénita*, *Ictiosis fetal ó intra-uterina*, *Ictiosis sebácea*, *Keratosis congénita*, *Keratoma difuso congénito*, *Keratoma maligno difuso congénito*, *Keratoma maligno congénito*, *Hiperkeratosis universal difusa congénita*, *Hiperkeratosis fetal*, *Hiperepidermotrofia generalizada y Eritrodermia congénita ictiosiforme con hiperepidermotrofia*, siendo también conocida por el aspecto raramente monstruoso de sus casos más intensos, con el nombre de *feto tarlequín*.

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ACTAS DERMO-SIFILIOGRÁFICAS

SUMARIO.—Oyarzábal: Chancro sifilítico forunculoide del pubis ó inoculación sobre un forúnculo de un chancro sifilítico.—Vacuna en el dorso de las manos.—Alvarez Sáinz de Aja: Dos casos de síndrome de Guillain-Thaon.—J. S. Covisa: Un caso de pénfigo vulgar.—Nonell: Un caso de sifiloma uretral perforante precoz.—J. de Azúa: Hiperkeratosis universal congénita maligna.—Prúrigo en placas simétricas de origen probablemente tuberculoso.—Intoxicaciones mercuriales terapéuticas.—Preparación de los calomelanos, salicilato de mercurio, etcétera, según la fórmula de Lang.—Contagiosidad de la lepra.—Procedimiento del émbolo anestésico para los lavados uretro-vesicales por presión.

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Abstract. Brief comment is made on the first article published in the journal *Actas Dermo-Sifiliográficas* with photographic documentation of the case history and histological findings. It was written by the founder of the journal, Dr. Juan de Azúa, in 1909.

Key words: *Actas,* Azúa, photography, history

EL PRIMER ARTÍCULO CON FOTOGRAFÍAS DE LA REVISTA ACTAS *DERMO-SIFILIOGRÁFICAS*: HIPERKERATOSIS UNIVERSAL CONGÉNITA MALIGNA. (ICTIOSIS FETAL; KERATOMA MALIGNO CONGÉNITO, ETC.), POR JUAN DE AZÚA

Resumen Se realiza un breve comentario del primer artículo publicado en la revista Actas Dermo-Sifiliográficas con documentación gráfica del caso clínico y de los hallazgos histológicos. Fue redactado por nuestro fundador, el Dr. Juan de Azúa, en el año 1909.

Palabras clave: *Actas*, Azúa, fotografía, historia

The first *Actas Dermo-Sifiliográficas* article in which photographs appear was the piece published by Azúa in 1909 entitled "Generalized malignant congenital hyperkeratosis (fetal ichthyosis; malignant congenital keratoma, etc.)."[1]

Azúa describes a case "of a morbid, little known, and infrequent type that has been named in a variety of ways."

Subsequently, he stresses his interest in this patient, since "because severe cases usually result in death after the first hours or days of life, they are observed mainly by midwives, and it is rare for a dermatologist to be able to study them."

Azúa was called in by a famous obstetrician at the Madrid maternity hospital, Dr Enrique de Isla, in May 1909, to "examine a child with a serious skin disorder, born that same day and not likely to survive long." The patient, in fact, died that same afternoon, 10 hours after birth and a half hour before Azúa arrived at the maternity hospital.

Figure 1. Case history

Dr. Azúa takes note of the basic information: the mother was young and strong, not suffering from syphilis, and without skin disorders. The pregnancy and delivery were normal. The fetus was female, of average length and weight, born with respiratory distress and weak sucking reflex.

His description of his examination of the deceased infant is worth transcribing word for word: "The body was still warm. On it I observed, mainly in the folds of the joints, genital organs, neck, scalp, and ears a sebaceous coating, not very thick and dirty white in color, consisting of dead epidermal cells and fat, as subsequently confirmed. The entire skin surface was hard and stiff, resistant, inflexible: the horny layer resembled leather split in several places by long, deep, bloody cracks. These cracks formed radial patterns around the anus, vulva and mouth that recalled the fissures typical in cases of hereditary syphilis. The ears, with rudimentary and malformed auricles barely separate from the head, had a narrow auditory canal clogged with epidermal detritus. The extremely blunt nose had very small nostrils. The mouth was circular, and between its cracked, stiff, and immobile lips a fairly well-formed tongue appeared. The eyes were obscured by eyelid ectropion and could not be seen. Herniated conjunctivae appearing between the everted eyelids formed red protuberances… The vulva was open and flat, with small fissures around the vaginal orifice. There was not the slightest trace of hair, eyelashes or eyebrows.

On the hands and feet, the fingers and toes were cylindrical in shape and thin, the nail plate covered in a horny casing that obscured the nails…" (Figure 1). Figure

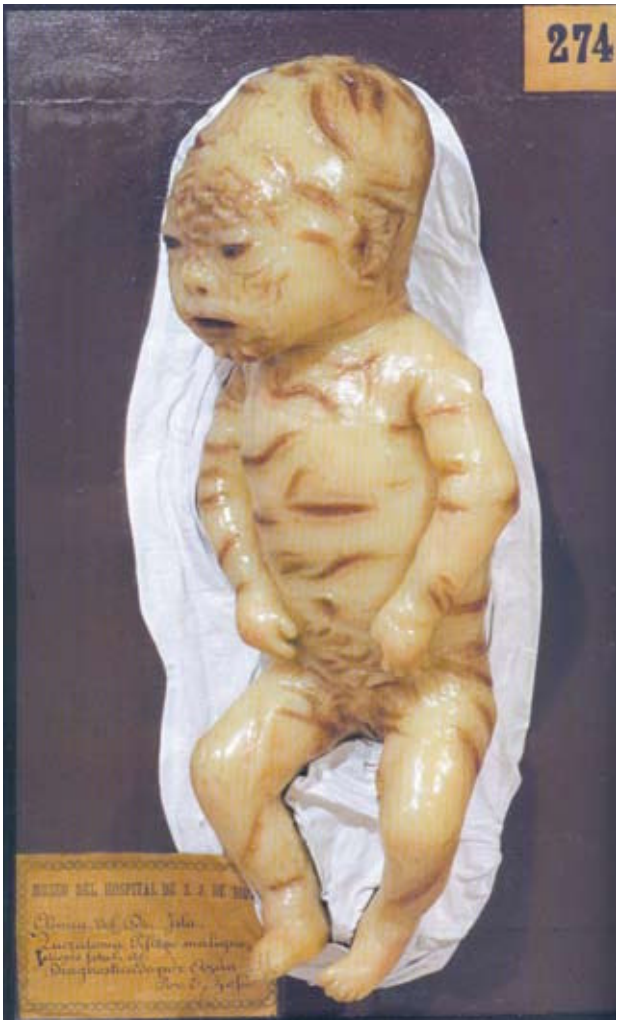

Figure 2. Wax model

274 in the wax model collection of the Hospital San Juan de Dios Museum[2] is a faithful reproduction of this case (Figure 2).

Next, Azúa offers a detailed histological description of the samples taken from the cervical and pectoral regions: "…the horny layer appears enormously thickened… The normal undulation of the stratum corneum is greatly increased and in some samples rather oblique, forming an almost perfect moiré pattern… The inner edge of the stratum corneum is extremely uneven and irregular, penetrating the Malpighian layer to various depths, sometimes so deeply that it forms actual spikes. Summary: severe hyperkeratosis of the horny layer with invasion of hair follicles, and penetration of the Malpighian papillae, atrophy of hair follicles or absence of hair, irregular thickening of the stratum granulosum; Malpighian layer somewhat thickened; small and perhaps scarce sebaceous glands; normal sweat glands; no indication of inflammation of the dermal papillae and dermis."

Figure 3. Histological study

Azúa illustrates the most representative histological findings in this case with two photographs, a first for *Actas Dermo-Sifiliográficas* (Figure 3).

Following a brilliant review of the literature, he concludes his article by drawing 5 conclusions:

1. The cases described with symptoms indicated correspond to 2 different pathologies: lamellar ichthyosis in newborns, and generalized malignant congenital hyperkeratosis.
2. Both diseases are distinct from ichthyosis vulgaris.

3. Attenuated, mild forms of generalized fetal congenital hyperkeratosis exist, including Brocq's congenital ichthyosiform erythroderma with epidermal hypertrophy.
4. The existence of fat droplets in the corpus papillare and papillae, noted by Darier in generalized congenital hyperkeratosis, was not observed in this case.
5. In this disease, the fundamental lesions consist of a massive, dense and generalized hyperkeratosis, with invasion of the pilosebaceous structures and consequent atrophy of the hair follicles and sebaceous glands, and an irregular thickening of the stratum granulosum.

Today we would identify this case as harlequin ichthyosis. This is a rare, serious, and generally fatal ichthyosis found in newborns, in which significant loss of water through the skin and resulting inability to regulate body temperature cause a serious disturbance in electrolyte balance.

Most such cases occur sporadically, although some authors have identified this as an autosomal recessive disorder. Three types have been described, defined by the presence, shrinkage, or absence of keratohyalin granules.[3]

In the future, treatment with oral retinoids and intensive neonatal support may improve the prognosis.

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