Use of Articles in the Pachyonychia Congenita Bibliography

The articles in the PC Bibliography may be restricted by copyright laws. These have been made available to you by PC Project for the exclusive use in teaching, scholarship or research regarding Pachyonychia Congenita.

To the best of our understanding, in supplying this material to you we have followed the guidelines of Sec 107 regarding fair use of copyright materials. That section reads as follows:

Sec. 107. - Limitations on exclusive rights: Fair use
Notwithstanding the provisions of sections 106 and 106A, the fair use of a copyrighted work, including such use by reproduction in copies or phonorecords or by any other means specified by that section, for purposes such as criticism, comment, news reporting, teaching (including multiple copies for classroom use), scholarship, or research, is not an infringement of copyright. In determining whether the use made of a work in any particular case is a fair use the factors to be considered shall include - (1) the purpose and character of the use, including whether such use is of a commercial nature or is for nonprofit educational purposes; (2) the nature of the copyrighted work; (3) the amount and substantiality of the portion used in relation to the copyrighted work as a whole; and (4) the effect of the use upon the potential market for or value of the copyrighted work. The fact that a work is unpublished shall not itself bar a finding of fair use if such finding is made upon consideration of all the above factors.

We hope that making available the relevant information on Pachyonychia Congenita will be a means of furthering research to find effective therapies and a cure for PC.
SOCIETY TRANSACTIONS

PHILOSOPHY


LUPUS ERYTHEMATOSUS. Presented by Dr. Ludwig Oulmann.

R. L., a woman aged 29, of German descent, was presented before the Section of Dermatology and Syphilis of the New York Academy of Medicine in February 1935 by Dr. Spiegel with the diagnosis of erythema elevatum diutinum. As there had always been scaliness and follicular plugs I could not agree with the diagnosis but regarded the case as one of lupus erythematosus. The patient has received two more injections of a gold preparation, and considerable improvement has resulted. There was a superficial patch with adherent scales on the left cheek. A new lesion has developed on the right ear since the first presentation. Tests with tuberculin were positive.

PACHYONYCHIA CONGENITA. Presented by Dr. George C. Andrews.

I. L., a boy aged 11 years, has the following history: Two weeks after birth the nails grew out like talons. When the child was 6 months old the nails of the hands and feet began to fall off at intervals of about two months. The formation of an area of redness and boginess surrounding the nail bed preceded the shedding of the nail. When the patient was 18 months old warty lesions developed on the knees; later similar lesions appeared on the elbows, popliteal areas, buttocks, legs and ankles. At irregular intervals a series of bullae developed.
These occurred on the toes and heels and along the borders of the feet. The bullae were painful, and when they were opened there remained a raw denuded surface which was slow in healing.

The striking feature of the specimen was a thickening of the epidermis due to acanthosis and parakeratosis, especially pronounced about the pilosebaceous follicles. The rete pegs were lengthened, and about the follicles the epidermal thickened caused funnel-shaped prolongations extending into the corium. The openings of the follicles were dilated and plugged with imperfectly cornified and somewhat degenerated horny material, and horny plugs were also present in the sweat pores.

The basal cell layer was irregular, and the cells were swollen. There was granular degeneration in the prickle cells; the nuclei stained deeply, and some were crescents, being pushed to one side of the cell by hydrops. They resembled the *corps ronds* found in cases of Darier's disease. The papillary bodies between the rete pegs were elongated, and in some places the apexes came near the surface of the skin.

The blood vessels of the corium were dilated and surrounded by lymphocytes, mast cells, connective tissue cells and an occasional plasma cell; the appearance suggested a mild inflammation from the pressure or irritation of the overlying thickened epidermis. The connective tissue seemed normal.

The nails of all the toes and fingers have been surgically removed by another physician. Some nails have regrown in distorted fashion. The bullae and denuded areas have responded slowly to application of soothing and antiseptic ointments. However, new bullae continue to develop at intervals. Ultraviolet therapy seems beneficial. Application of keratolytic pastes has relieved the nutmeg grater-like scales on the legs and elbows.

**DISCUSSION**

Dr. Howard Fox: I agree with the diagnosis of this rare condition. The appearance of the nails was formerly precisely similar to that in the case originally described by Jadassohn and Lewandowsky. At present the nails have changed in appearance as a result of their removal several months ago. The numerous warty lesions and the vesicles are all part of the syndrome described by Jadassohn. Leukoplaikia has not been present in this case. In the case which I reported the condition closely resembled the one in this patient.

**ACTINOMYCOSIS. Presented by Dr. George C. Andrews.**

M. H., a Negress aged 35, entered the Vanderbilt Clinic on Oct. 17, 1934, complaining of pain and swelling in the lower portion of the face on the left side and of inability to open her mouth.

The left lower third molar had served as a bridge abutment for years. Because the patient suffered from pain beneath this molar a dentist gave the injections to cure what he diagnosed as “pus” in the tooth. Pain followed and was present for two months; then the face became swollen. The tooth was extracted on Sept. 26, 1934. The swelling did not subside, and the jaw felt stiff. The patient could open the mouth about 2 cm. The swelling involved the angle of the jaw anterior to the ear. No submaxillary involvement was noted. Anterior to the swelling there was edema. On Nov. 8, 1934, an incision in three areas over the mass released thick, foul-smelling pus and necrotic cellular tissue. All incisions communicated. A smear of pus was found to contain sulfur granules caused by actinomycosis.

Treatment has consisted of administration of a high vitamin diet, local applications of potassium iodide and roentgen treatment, a dose of 160 kilovolts, filtered with 1 mm. of aluminum and 0.25 mm. of copper, 150 roentgens being administered at each treatment. The patient received a total of seven treatments; the last one was given on March 11, 1935.

**ACTINOMYCOSIS. Presented by Dr. George C. Andrews.**

I. R., a boy aged 13, entered the Vanderbilt Clinic complaining of a gradually increasing red swelling on the left side of the jaw, which began in July 1934. The mass was incised to open his mouth. A diagnosis of actinomycosis was instituted on a purely clinical basis, and potassium iodide and roentgen therapy were administered weekly; 1935.

Dr. Howard Fox: I agree with the diagnosis of actinomycosis. The tissues have been treated for straws and mucus membranes (mucous membrane when the tissue was cut from the skin before the plaque).

**LICHEN PLANUS.**

This woman was seen in 1935. The typical picture is of a reddish-brown, bluish color of papules and typical patches arranged in straight lines and other parallel lines. Examination showed 10,000 white blood cells with normal nitrogen and uric acid. Phthalein test (150 mg. per cent.) Flushing was noted.

Treatment has consisted of nitrogen and antibiotics with the addition of a drug. The case has been followed for three years.

Dr. Howard Fox: I agree with the diagnosis of lichen planus. The lesions are not caused by a drug. The treatment has consisted of potassium iodide and antihistamines.