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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.
cally demonstrated, in addition to a larger tuberculoma in the left hemisphere. False localising signs due to multiple tuberculomata are not infrequent. They are reported in 9 cases by Dastur and Desai (loc. cit.) and in 6 per cent of cases reported by Ramamurthi et al. (loc. cit.). A case was described (Dastur and Desai, loc. cit.) where the patient had right hemiparesis, right 3rd, 6th and bilateral 6th cranial nerve paresis. On investigation, the patient was proved to have a large right temporal and right cerebellar tumour. Temporal tumour was found to be tuberculoma and excised while the cerebellar mass regressed with antituberculosis medication. Patient was reported to be alive five years later.

**DISCUSSION**

All the three patients had the constitutional sign of tuberculosis of different degrees. The first two had substantial evidence of systemic tuberculosis. All the three responded well to the drug therapy alone.

As pointed out initially the diagnosis was largely presumptive and not based on direct biopsy report but on the circumstantial and therapeutic evidences which were strong enough to support this etiology. With the advent of effective antituberculosis drugs and improved anaesthetic techniques, the surgical results of excision of tuberculomas have very much improved and yet the morbidity of major craniotomy may not be acceptable to all patients especially those who are ill with toxæmia of systemic tuberculosis. Management with drug therapy alone therefore needs emphasis. While on drug therapy patients are hospitalised and observed. Surgery is indicated when the signs are progressive despite medication or when the mass is large in size. The results of such an expectant method of treatment justify this approach. The masses get smaller and the oedema clears. In genuine fossa lesions even when the masses are small, obstruction to cerebrospinal fluid pathways or pressure by the oedema may be significant. With timely treatment this also improves with drugs (Ramamurthi et al., loc. cit.).

**SUMMARY**

Tuberculoma is one of the most frequent intracranial space-occupying lesions in our country. Properly selected cases can be successfully managed with chemotherapy without reporting to craniotomy and excision. Careful follow-up is necessary. Three illustrative cases are reported.

**REFERENCES**


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**Sodium Laevothyroxine in Pachyonychia Congenita**

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Pachyonychia congenita, a congenital and often familial ectodermal disorder, was first described by Jadassohn and Lewandowsky in 1906. Diasso, while reviewing the literature in 1934, quoted Heller who called attention to the close resemblance between the condition reported by Jadassohn and Lewandowsky (loc. cit.) and the syndrome described as "Congenital dyskeratosis" by Schauer (1926). Schréweis reported in 1935 a similar type of case under the caption "Pachyonychia ichthyosiformis". A few cases of dyskeratosis congenita and Darier's disease, reported in the literature, had some of the features common to all cases while in others they were present or absent. Ormsby in 1943 recognised the title 'pachyonychia congenita' and summarised the clinical features in detail. Palmar and plantar hyperkeratosis, dystrophic changes in the nails, follicular keratosis, dyskeratosis of cornae, lentikulatikia, appearance of verrucous lesions of the knees, elbows, popliteal regions, buttock, legs and ankles and the formation of bullae on the volar aspect of the feet are mentioned as the chief features of the disorder. General ichthyosis of variable degree may also be present. Variations of the clinical features have been reported by different workers. The association of pachyonychia congenita with epidermolysis bullosa in three generations had been reported by May (1962), with mental retardation by Niedelman in 1958, and with steatocystoma multiplex in 8 cases in 4 generations by Vineyard and Scott (1961).

The condition shows a strong sex predilection for males and probably follows more or less a dominant mode of transmission but not of the simple type.

**CASE REPORT**

S. N., Hindu male, aged 14 years, 11th issue of the parents, attended the skin outpatients' clinic in February, 1964, with the complaints of hyperkeratosis of the palms and soles, follicular keratotic lesions all over the body specially on the back, buttock, extremities, joints, face and neck region (Fig. 1, *vide* Plate) along with frequent appearance of blisters on the heels and borders of the soles since the age of 5. He had black thick stony hard nails on his fingers (*vide* Plate) and distorted nails of his toes since birth. The thickness of the nails...
at the free margin varied from 6 mm. to 10 mm. He had an acralial development of small vesicles on the eyebrows and upper lip which gradually became crustous. The hair of the scalp was lustreless, thin and scanty in distribution.

Recurrent deep-seated bullae appeared on the heels and borders of the soles, sometimes exposing the bony structures through the resultant ulcers.

The hyperkeratotic condition of the palms and soles, extended beyond these regions. The skin over the dorsum of the hands and of the feet was thick; this was more marked on the fingers and interphalangeal joints. Follicular keratotic lesions were present all over the body; on the joint areas the skin was thickened, black and warty due to the coalescence of the papular lesions.

The patient had leukoplakia of the tongue. He looked younger than his actual age and his intelligence was much below average.

There was no history of similar illness in his family and no history of consanguinity of his parents.

Investigations — The haematological investigations showed no abnormality. Serological test for syphilis was negative. Microscopical examinations of the material taken from the nail scrapings and the leukoplakic lesions of the tongue with 10 per cent KOH and culture in Sabouraud’s media revealed no fungus. X-ray of the skull showed no abnormality. Stool and urine examinations revealed no abnormal findings. Liver function tests were within normal limits. Blood chemistry: urea, N.P.N., sugar, uric acid was normal. Blood cholesterol was 177 mg. per 100 ml. B.M.R.: -10. Report of thyroid investigation with radio-active iodine (131I): Thyroid/Thigh ratio at 2nd hour: 2.62, percentage uptake of 131I by thyroid at 2nd hour: 13.13, percentage uptake of 131I by thyroid at 24th hour: 39.06 per cent and F.B.I. at 48th hour (per litre of plasma): 0.1356 per cent.

Diagnosis—Normal.

Histopathological examination. — The epidermis showed wrinkling as a whole with slight hyperkeratosis with keratotic plugging of the follicles (Fig. 3, vide Plate). There was some degree of parakeratosis around the plugged follicles; the stratum malpighii was thin at places. A large number of sweat and sebaceous glands was seen in the dermis. PAS resection and silver impregnation did not show any abnormal findings. Historical features were consistent with pachyonychia congenita.

Treatment—The patient was put on Sodium Levothyroxine, 0.2 mg. daily. In 5 weeks, the follicular lesions of the body improved considerably and the roughness of the skin also decreased. The condition of the nails did not show any appreciable change at that time. On increasing the dose to 0.4 mg. daily for further 3 weeks all the nails became loose and fell off leaving thin unren silver coloured nails covering the proximal half of the nail beds. The colour and texture of the hair became black, coarse and thick. Due to some loss of weight and weakness, the drug was gradually reduced to 0.2 mg. The nails began to regrow after their original pattern at a slightly rapid rate and regained the thickness of 3 mm. on the fingers 2 weeks after the stoppage of the dose of 0.4 mg. of sodium laevothyroxine but the improved condition of the skin and face was maintained. At that time, vitamin A, 50,000 i.e., orally, was started along with 0.2 mg. of sodium laevothyroxine daily and continued for 3 months without any appreciable change in the nail condition.

The ulcers of the heels and borders of the soles were treated by antiseptic bath and 2 per cent gentian violet locally, by which the acuteness of the ulcerated conditions was minimised i.e., the discharge was less and pain was relieved. In course of a fortnight the ulcers became dry. But the ulcers recurred after an interval of 2 months approximately during sodium laevothyroxine therapy.

Discussion

A case of pachyonychia congenita, a very rare disease, is reported here. As yet there is no satisfactory treatment of the condition and the therapy advocated so far related to surgical procedures. As the nail condition developed from the matrix, the surgical amputation of phalanges was reported to be useful (Wright and Guequiere, 1947). The removal of nail plate gave no permanent relief. Andrews (1938) and Cosman et al. (1948) have recently advocated surgical excision of nails and soft tissue above the bone and cauterisation of phalanges with full thickness graft, keeping a small portion of the matrix lead to failure with regrowth of nails from the matrix.

Medical treatment with high dose of vitamin A has given no appreciable result. Cystene hydrochloride locally was claimed as of value by Wright and Guequiere (loc. cit.) for blisters of the sole.

In our case, a thyroid preparation brought about an overall improvement. The condition of the skin and hair improved nicely and was being maintained during the period of therapy. With 0.4 mg. of sodium laevothyroxine, thick black nails were shed off but the young nails which grew later remained rudimentary. The drug could not be continued due to intolerance of the patient and he was losing weight. The follicular lesions of the skin have not recurred since the thyroid therapy.

With irregular use of sodium laevothyroxine, 0.4 mg. or 0.2 mg. daily or on alternate days, since the last few months his skin condition was well maintained, i.e., follicular hyperkeratotic lesions did not appear again and the periodic appearance of blisters on the sole has also been minimised to a great extent. Although the nails became thick this was much less compared with the original condition.

The detailed laboratory investigations could not definitely prove hypothyroidism but a latent hypothyroid state may be suggested as the patient improved with small doses of thyroid.

Thyroid helps in epithelisation and keratin formation. The mode of action of the thyroid on nail formation is not yet known. From our case it appears that irregular thickening of the nail plates was caused by thyroid, probably acting on the matrix. It requires further study on a large number of cases to arrive at a definite conclusion. Due to the rarity of the condition, the trial could not be performed in other cases.
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Bilirubin and sodium laevohydroxine was used in the treatment of the case.

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Silicosis in a Mica-mine Worker

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Exposure to silica in the course of mining for gold is a well known occupational hazard. In the mica-mining industry which is confined to a few parts of the World (U.S.A., South Africa and India), exposure to silica and development of silicosis is an inherent risk but the number of cases reported is far less than one would expect, considering the conditions of work in these mines. The micas are a group of complex aluminium silicate compounds of several types. An expositure of several years to ordinary industrial environment containing these substances is necessary to produce the disease. The micas are used in the manufacture of paper products, in the production of some paint products, as lubricants in combination with oils and for making insulating materials in electrical devices. In India, mica mines are located in the states of Bihar, Andhra Pradesh and Orissa. Zones of mica-containing rock are usually found near the walls of the surrounding country rock and also near separations between rocks that are largely quartz and rocks that are composed largely of feldspars. In mining for mica, the procedure is to follow the mica-bearing zones as much as possible. However, it is often necessary to excavate non-micabearing rock in order to reach micabearing zones. Hand drilling is done with no dust control measures in some regions. The case, reported here, is that of a male of 27 years who, ten years earlier, had worked for 15 months in a mica mine in Nellore, South India, as a hand-driller with little or no protective measures. Subsequent to the exposure, he had worked as a pump attendant in the fields for nearly 10 years before presenting himself with symptoms.

Case Report

P., aged 27 years, reported for the first time on 30-10-61 with the complaint of intermittent fever for a year, mostly in the evening and cough with moderate expectoration, occasionally yellowish. He had breathlessness of three months' duration. The illness started with dry cough followed later by cough with expectoration, yellowish at times, of about half an ounce per day. There was no history of haemoptysis. For three months prior to admission, he had breathlessness, more at night, sometimes paroxysmal in nature with wheeze.

He gave his occupation as one of attending an oil pump in the fields. There were no features in the history suggestive of allergic disorder. On further questioning, he said that about ten years earlier he had worked as a hand-driller in a mine without dust control measures in Nellore District of Andhra Pradesh for about 15 years. It was gathered from him that the drilling operations went on under dry conditions, the atmosphere in the mine becoming laden with dust. He left the job as he developed severe cough and subsequently worked as a labourer in an agricultural farm. His cough subsided to a great extent in a month even though he was disturbed occasionally at night by paroxysms of cough.

Physical examination—He was a moderately nourished individual. He was obviously breathless on admission and was orthopneic. Minimal clubbing of the fingers was present. There was no significant lymphadenopathy. The signs in the chest were those of fibrosis in the lower regions associated with generalised bronchoepaem. There was no clinical evidence of pulmonary hypertension, right ventricular hypertrophy or of congestive failure.

Investigations—Total leucocyte count: 12,000/c.mm. with polymorphs 74 per cent, lymphocytes 24 per cent and eosinophils 2 per cent. E.S.R. was 15 mm. in 1 hour and 40 mm. in one hour.