Tylosis palmaris et plantaris familiaris in association with Dupuytren’s contracture: A case of ectodermal anomaly

A 92-year-old man was admitted to hospital for confusion. Clinical examination revealed hyperkeratotic lesions on the palmar and plantar surfaces, diagnosed as tylosis palmaris et plantaris familiaris (TPEP), and Dupuytren’s contracture on his left hand. No other clinical, radiological or paraclinical findings were identified. His father and two brothers had the same hyperkeratotic lesions.

Tylosis palmaris et plantaris familiaris (TPEP) is a rare familial ectodermal anomaly of the palms and soles characterized by marked hyperkeratosis. It usually begins in early childhood with redness of the skin of the palms and soles which gradually becomes thicker and develops a yellowish, waxy appearance. Increased sweating is quite common and there is a tendency to fungal and bacterial infections of the feet. This condition may affect the knuckle pads and nails but usually does not extend beyond the hands and feet. It was first described as a syndrome by Herrmann Arthur Thost (German physician, 1854–1937) and Paul Gerson Unna (German dermatologist, 1850–1929). The case is presented here of a 92-year-old man who was admitted in a state of confusion due to a urinary tract infection.
in a humid atmosphere, with notable improvement during fishing periods. He had never visited a doctor for this condition, as he was poor, and had no health problems other than the “thick skin”. When he was younger he used to “peel” the excess skin. The frequency of peeling varied considerably, from once yearly to two, three or even four times a year. In addition to the hyperkeratotic lesions and the urinary tract infection, he had a notable Dupuytren’s contracture on his left hand (fig. 4, also noted in fig. 5). The remainder of the general examination was essentially normal. X-ray of the hands and feet was normal with mild degenerative changes due to his age (figures 5, 6) apart from Dupuytren’s contracture on the left hand (fig. 5). Chest X-ray showed degenerative changes of his thoracic spine with osteophytes and kissing osteophytes (fig. 7). X-ray of his abdomen, showed osteophytic degenerative changes

Figure 1. Typical lesions in tylosis palmaris et plantaris.

Figure 2. Hyperkeratotic lesions.

Figure 3. Hyperkeratotic lesions.

Figure 4. Dupuytren’s contracture on left hand.

Figure 5. Dupuytren’s contracture.
of his lumbar spine with kissing osteophytes and narrowing between the vertebral spaces (fig. 8). The laboratory findings were not significant.

Family members

The father and two brothers had the same condition and two other brothers left the country on a young age, so there is no information about their own condition or that of their children. Figure 9 presents the patient’s family tree. No other family member is reported to have the same condition. It appears that the condition is determined by a single dominant gene which in this family was transmitted entirely in accordance with theoretical expectations through four generations. No other person had Dupuytren's contracture.

Figure 6. X-ray of the feet.

Figure 7. Chest X-ray.

Figure 8. Abdominal X-ray.

Figure 9. Pedigree of TPEP (tylosis palmaris et plantaris)
DISCUSSION

It is evident from the literature that the condition described is an example of TPEP, a rare but poorly defined clinical entity, which has been given various names, including diffuse nonepidermolytic palmoplantar keratoderma, palmoplantar keratoderma diffusa circumscripta, congenital keratoderma of the palms and soles, hereditary palmoplantar, hyperkeratosis palmaris et plantaris, keratosis palmarum et plantarum, keratoma palmarae et plantae, and ichthyosis palmarum et plantarum. Some authors have reported TPEP to be associated with esophageal cancer, squamous cell carcinoma of the lung, deafness, congenital heart disease and hereditary optic atrophy, but the patient presented here had no additional findings other than Dupuytren’s contracture on his left hand.

TPEP appears at an early age and is usually symmetrical in distribution. It is characterized by thickening of the skin on the palms and the soles of feet but it may affect the palms to a lesser degree. Progression of the disease leads to marked thickening of the palmoplantar surfaces with variation in disease patterns leading to morphological differences in the skin lesions. The palmoplantar thickening has a significant impact on activities of daily living such as walking and grasping, because of pain, contractures and reduced tactile sensitivity.

The diagnosis may be made on clinical examination alone but biopsy may be of help in the histological identification of specific syndromes. Although this condition falls in the realm of dermatology, general practitioners, internists, plastic and reconstructive surgeons and other specialists may be involved, depending on the complications that the disease presents for the patient.

Treatment is non-specific and aims to alleviate pain, prevent complications, minimize hyperkeratosis and improve function. Emollients, keratolytics and retinoids have been used, with some success, to soften the thickened skin. In advanced disease more aggressive treatment may be required. Surgical management consists of excision of the keratotic lesions and grafting, or treatment of secondary complications such as contracture formation by release procedures. When the sole of the foot is involved, total excision of the skin of the sole, including dermal components to the level of the plantar aponeurosis or deep subcutaneous fat, with subsequent grafting has been documented, with reduced recurrence of keratoderma on the treated sole, enabling patients to regain the ability to walk. Unfortunately, despite initial favorable results, surgical management does not appear to be the definitive treatment modality, because recurrence of the disease in the area treated is high.

There is no single answer for the treatment of TPEP and multidisciplinary treatment planning is essential.
References


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