

CASE REPORT ΕΝΔΙΑΦΕΡΟΥΣΑ ΠΕΡΙΠΤΩΣΗ

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.

ARCHIVES OF HELLENIC MEDICINE 2015, 32(1):106–110
ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2015, 32(1):106–110

E. Pelechas,¹
A. Kavvadias,²
P. Karagianni,³
N. Tsigaridas,⁴
C. Manatou,²
P. Tsapogas²

¹Department of Internal Medicine,
Derriford Hospital, Plymouth, United
Kingdom

²Second Department of Internal
Medicine, General Hospital of Corfu,
Corfu

³Department of Microbiology, University
Hospital of Ioannina, Ioannina

⁴Department of Cardiology, "Hatzikosta"
General Hospital, Ioannina, Greece

Οικογενής παλαμο-πेलματιαία
τύλωση σε συνδυασμό με
σύσπαση Dupuytren: Παρουσίαση
μιας σπάνιας περίπτωσης

Περίληψη στο τέλος του άρθρου

Key words

Dupuytren's contracture
Ectodermal anomaly
Hyperkeratosis

Υποβλήθηκε 28.4.2014
Εγκρίθηκε 24.7.2014

Tylosis palmaris et plantaris familiaris (TPEP) is a rare familial ectodermal anomaly of the palms and soles characterized by marked hyperkeratosis.¹ 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.² 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).² 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.

CASE REPORT

A 92-year-old, peasant/fisherman, was admitted to the Corfu General Hospital, Greece with confusion due to a urinary tract infection. Typical hyperkeratotic lesions of the soles (figures 1, 2) and the palms were present (fig. 3), with extension of the lesions to the dorsal surfaces of the index and little fingers and the lateral surfaces of the median and parameian fingers. The patient's wife gave a good description of tylosis in other male members of her husband's family. The condition had become evident in early childhood and had undergone flare-ups and remissions during his life. It was aggravated by cold, dry weather, which resulted in gross thickening, cracking and even bleeding, especially of the palmar skin. Remission occurred

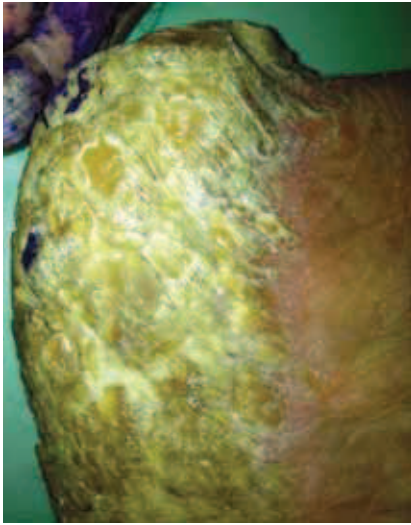


Figure 1. Typical lesions in tylosis palmaris et plantaris.



Figure 2. Hyperkeratotic lesions.



Figure 3. Hyperkeratotic lesions.

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 4. Dupuytren's contracture on left hand.



Figure 5. Dupuytren's contracture.



Figure 6. X-ray of the feet.

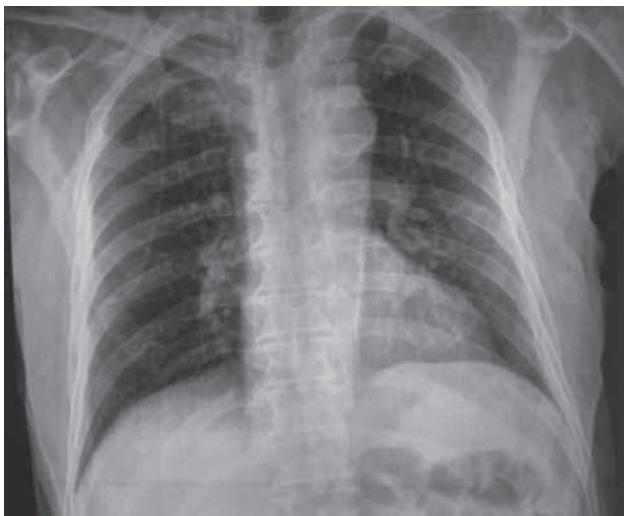


Figure 7. Chest X-ray.

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 8. Abdominal X-ray.

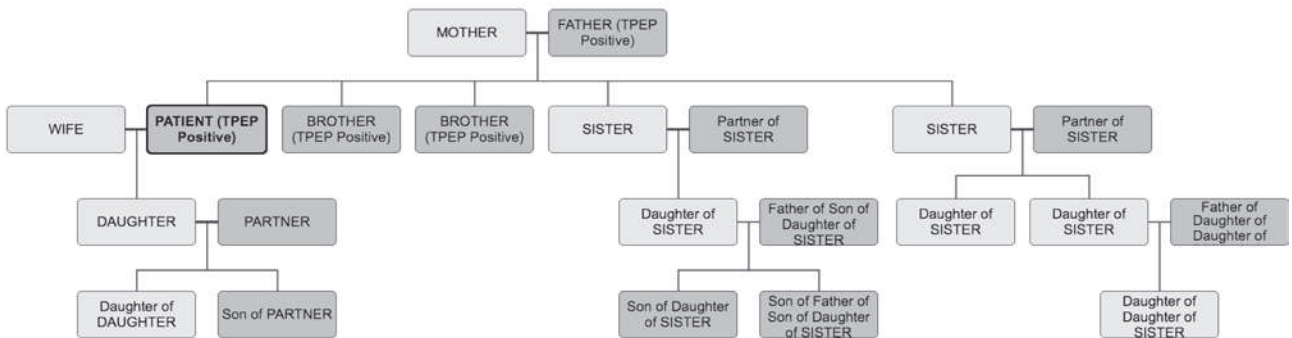


Figure 9. Pedigree of TPEP (tylosis palmaris et plantaris)

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.^{3,4} Some authors have reported TPEP to be associated with esophageal cancer,⁵ squamous cell carcinoma of the lung,⁶ deafness,⁷ congenital heart disease⁸ and hereditary optic atrophy,⁹ 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.¹⁰ It is characterized by thickening of the skin¹¹ on the palms and the soles of feet but it may affect the palms to a lesser degree.¹² 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.¹³

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.¹⁴

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.¹⁵ Surgical management consists of excision of the keratotic lesions and grafting, or treatment of secondary complications such as contracture formation by release procedures.¹⁶ 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.¹⁷ 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.

ΠΕΡΙΛΗΨΗ

Οικογενής παλαμο-πελματιαία τύλωση σε συνδυασμό με σύσπαση Dupuytren: Παρουσίαση μιας σπάνιας περίπτωσης

Ε. ΠΕΛΕΧΑΣ,¹ Α. ΚΑΒΒΑΔΙΑΣ,² Π. ΚΑΡΑΓΙΑΝΝΗ,³ Ν. ΤΣΙΓΑΡΙΔΑΣ,⁴ Χ. ΜΑΝΑΤΟΥ,² Π. ΤΣΑΠΟΓΑΣ²

¹Παθολογική Κλινική, Νοσοκομείο Derriford, Plymouth, Ηνωμένο Βασίλειο, ²Β΄ Παθολογική Κλινική, Γενικό Νοσοκομείο Κέρκυρας, Κέρκυρα, ³Τμήμα Βιοπαθολογίας, Πανεπιστημιακό Νοσοκομείο Ιωαννίνων, Ιωάννινα, ⁴Καρδιολογική Κλινική, Πανεπιστημιακό Γενικό Νοσοκομείο «Χατζηκώστα», Ιωάννινα

Αρχεία Ελληνικής Ιατρικής 2015, 32(1):106–110

Ένας άνδρας, 92 ετών, εισήχθη στο νοσοκομείο με σύγχυση. Κατά την κλινική εξέταση παρατηρήθηκαν υπερκερατωτικές αλλοιώσεις στις παλαμιαίες και τις πελματιαίες επιφάνειες των χεριών και των ποδιών του. Επίσης, παρατηρήθηκε σύσπαση Dupuytren στο αριστερό του χέρι. Δεν εντοπίστηκαν άλλες αλλοιώσεις κατά την κλινική εξέταση, τον απεικονιστικό και τον εργαστηριακό έλεγχο. Από το οικογενειακό ιστορικό αποκαλύφθηκε ότι ο πατέρας του και οι δύο αδελφοί του παρουσίαζαν τις ίδιες υπερκερατωτικές αλλοιώσεις.

Λέξεις ευρητηρίου: Εκτοδερμική ανωμαλία, Σύσπαση Dupuytren, Υπερκεράτωση

References

1. NAGAI H, EMI M. Palmoplantar keratosis. *Jpn J Clin Med* 2000, 58:1501–1504
2. KLINTWORTH GK, ANDERSON IF. Tylosis palmaris et plantaris familiaris associated with clinodactyly. *S Afr Med J* 1961, 23:170–175
3. VOHWINKEL KH. Keratoma hereditarium mutilans. *Arch Derm Syph* 1929, 158:354–364
4. ITIN PH, FISTAROL SK. Palmoplantar keratodermas. *Clin Dermatol* 2005, 23:15–22
5. HOWEL-EVANS W, McCONNELL RB, CLARKE CA, SHEPPARD PM. Carcinoma of the oesophagus with keratosis palmaris et plantaris (tylosis): A study of two families. *Q J Med* 1958, 27:413–429
6. NOMORI H, HORIO H, IGA R, FUYUNO G, KOBAYASHI R, MORINAGA S. Squamous cell carcinoma of the lung associated with palmo-plantar hyperkeratosis. *Nihon Kyobu Shikkan Gakkai Zasshi* 1996, 34:76–79
7. FITZGERALD DA, VERBOV JL. Hereditary palmoplantar keratoderma with deafness. *Br J Dermatol* 1996, 134:939–942
8. HOEGER PH, YATES RW, HARPER JI. Palmoplantar keratoderma associated with congenital heart disease. *Br J Dermatol* 1998, 138:506–509
9. DIMSDALE H. Hereditary optic atrophy in family with keratoderma palmaris et plantaris (tylosis). *Proc R Soc Med* 1949, 42:796
10. SYBERT VP, DALE BA, HOLBROOK KA. Palmar-plantar keratoderma. A clinical, ultrastructural, and biochemical study. *J Am Acad Dermatol* 1988, 18:75–86
11. CHRISTIANO AM. Frontiers in keratodermas: Pushing the envelope. *Trends Genet* 1997, 13:227–233
12. DENCER D. Tylosis. *Br J Plast Surg* 1953, 6:130–140
13. ATABAY K, YAVUZER R, LATIFOĞLU O, OZMAN S. Keratoderma hereditarium mutilans (Vohwinkel syndrome): An unsolved surgical mystery. *Plast Reconstr Surg* 2001, 108:1276–1280
14. AHN CS, ZOUMARAS J, FERNANDES A. Congenital tylosis and the role of the plastic and reconstructive surgeon. *Eur J Plast Surg* 2011, 34:493–496
15. MEVORAH B, GOLDBERG I, SPRECHER E, BERGMAN R, METZKER A, LURIA R ET AL. Olmsted syndrome: Mutilating palmoplantar keratoderma with periorificial keratotic plaques. *J Am Acad Dermatol* 2005, 53(Suppl 1):S266–S272
16. ATHERTON DJ, SUTTON C, JONES BM. Mutilating palmoplantar keratoderma with periorificial keratotic plaques (Olmsted's syndrome). *Br J Dermatol* 1990, 122:245–252
17. WYNN-WILLIAMS D. Plantar keratoderma treated by split-skin grafts. *Br J Plast Surg* 1953, 6:123–129

Corresponding author:

E. Pelechas, 17 Plymbridge Lane, Plymouth, PL68AJ, United Kingdom
e-mail: pelehas@yahoo.com