

Pachydermodactyly: a Case Report and Literature Review

Mirjana PARAVINA^{1,2}, Milenko STANOJEVIĆ^{1,2},
Dragan JOVANOVIĆ^{1,2} Dragana LJUBISAVLJEVIĆ²

¹ Medical Faculty, University of Niš, Serbia

² Clinic of Skin and Venereal Diseases, Clinical Center of Niš, Serbia²

*Correspondence: Mirjana Paravina, E-mail: mirjanaparavina@gmail.com

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Abstract

Pachydermodactyly is a rare, benign form of digital fibromatosis, characterized by asymptomatic and progressive, periarticular and usually symmetrical soft tissue finger swelling, specifically on the lateral aspects of the proximal interphalangeal joints mainly of the second, third, and fourth fingers; it mostly affects young adolescents and is probably due to repeated mechanical injury of the skin (such as repeated clasping or rubbing of crossed fingers), sometimes as a result of obsessive-compulsive disorder, which must be distinguished from obsessive "chewing pads". This paper presents a male patient aged 19, who presented with first symptoms at the age of 12, and was diagnosed with periarticular hypertrophy: localized soft tissue thickening around the proximal interphalangeal joints of all fingers except the thumbs; slight hypertrophy of the skin; absence of subjective complaints; normal joint function. Dermatological status on admission revealed: symmetrical soft tissue swelling of all fingers of both hands except the thumbs at the level of the proximal interphalangeal joints; normal appearance of the distal parts of all fingers; thickening at the level of the proximal interphalangeal joints, bilateral, almost symmetrical hypertrophy (ulnar and radial) of phalanges of the affected fingers except both index fingers, affecting only the ulnar side. The skin lesions were pain-free on palpation, with homogeneous texture and elastic consistency, freely movable over underlying structures. The affected joints showed no functional deficit. The test results, sonography, radiography and histopathology confirmed our clinical diagnosis - pachydermodactyly. The review of the currently available literature, published between 1973 and 2014, including 99 papers and 160 patients, provided important insight into the characteristics and variations of the disease.

Key words

Fibroma; Hand Dermatoses; Hand Deformities; Fingers; Soft Tissue Neoplasms; Cumulative Trauma Disorders

Pachydermodactyly (PDD) is a benign digital fibromatosis characterized by asymptomatic and progressive, periarticular and usually symmetrical soft tissue finger swelling, specifically on the lateral aspects of the proximal interphalangeal joints mainly of the second, third, and fourth fingers: thus, it produces a symmetrical, diffuse swelling of the skin around the dorsal and lateral aspects of the proximal phalanges of the index, ring and middle fingers (1).

It particularly affects young adolescents and is probably due to repeated mechanical skin injury, sometimes as a result of obsessive-

compulsive disorder, which must be distinguished from obsessive "chewing pads" (1, 2, 3). Diffuse swelling of the digits, which includes the dermis as well as epidermis, clearly distinguishes PDD from "knuckle pads", calluses, occupational callosities, etc. Affected families have been reported, and pachydermodactyly and knuckle pads may coexist (1).

Pachydermodactyly was first described by Bazex et al. in 1973 as "pachydermie digitale" (4). Verbov named this entity as pachydermodactyly from the Greek pachy (thick), dermo (skin), and dactyly (finger) in 1975 (5).

Case Report

This paper presents a male student aged 19, who developed thickening of the third and fourth fingers of the left hand at the age of 12. By the age of 14, he presented with the same changes on all fingers except the thumbs, and since then the condition has not changed. He associated these changes with hitting the ball, as well as with the habit of repeated clasping or rubbing the fingers. After he underwent evaluation by an orthopedist, physiatrist and rheumatologist, he was referred to our Clinic with the diagnosis of periarticular arthropathy in order to exclude psoriasis and/or seborrheic scalp dermatitis. On admission, the patient denied any other disease or a family history.

On admission, the patient's general condition was good. Dermatological examination revealed: symmetrical soft tissue swelling of all fingers of both hands except the thumbs at the level of the proximal interphalangeal joints; normal appearance of the distal parts of all fingers; thickening at the level of the proximal interphalangeal joints, bilateral, almost symmetrical hypertrophy (ulnar and radial) of phalanges of the affected fingers except both

index fingers, affecting only the ulnar side (Figure 1). The skin lesions were pain-free on palpation, with homogeneous texture and elastic consistency, freely movable over underlying structures. The affected joints showed no functional deficit.

Laboratory and other test results

All relevant laboratory test results were within reference values, including factors of inflammation, serum levels of immunoglobulins, C3 and C4 complement components, presence of antinuclear antibodies and immunoglobulins.

Ultrasonography of the hand joints: osteoarticular surfaces without signs of defects; there are no periarticular focal lesions, except for imbibition of the soft tissues; synovial thickening of the medial interphalangeal joints; intra-articular synovial fluid volume is increased.

X-rays of the hands (both directions) did not reveal any involvement of the bones structures (Figure 2).

Histopathological findings: severe epidermal hyperkeratosis and acanthosis; dermoepidermal junction is flat; dermis is thickened and hypocellular,

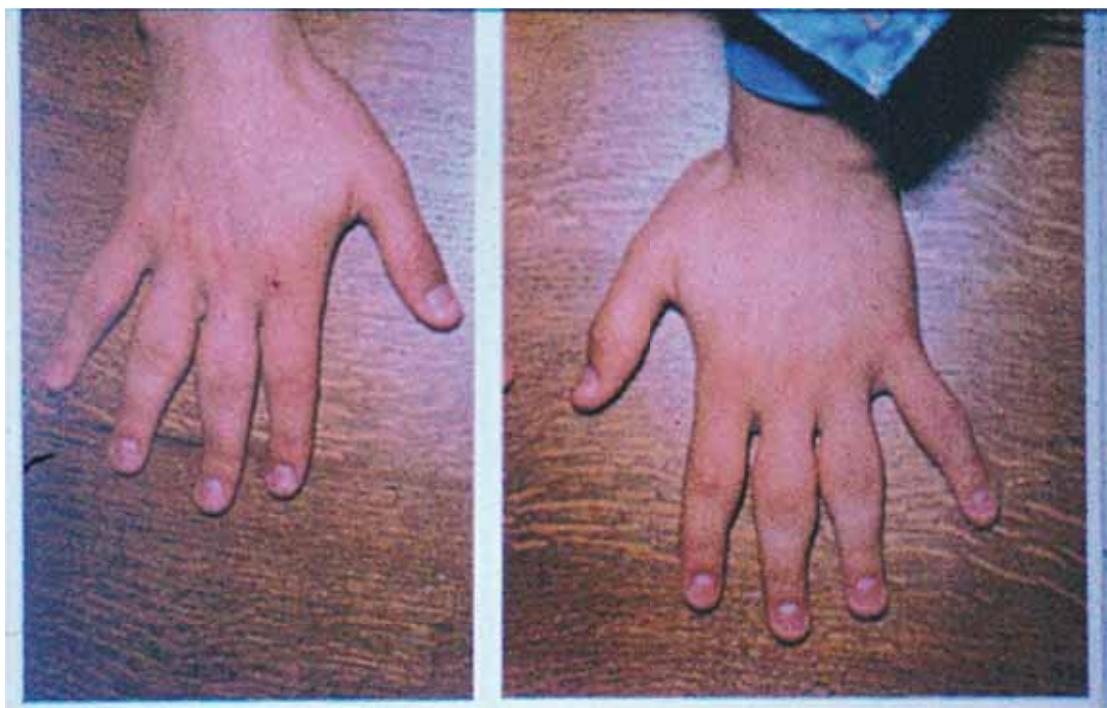


Figure 1. A symmetrical, diffuse swelling around both lateral (ulnar and radial) aspects of the proximal phalanges of the small, ring and middle fingers; proximal phalanges of both index fingers affected only around the ulnar aspects; distal parts without involvement



Figure 2. Hand x-ray: preserved bone structure

rare fibroblasts accompanied by hyalinized collagenous stroma; absence of adnexal skin structures.

Therapy

Intralesional triamcinolone solution injections during three months resulted in slight reduction of swelling of the treated finger. The patient was advised to avoid mechanical trauma, rubbing and cracking of the fingers.

Discussion and literature review

Pachydermodactyly is a condition which mostly occurs in adolescents and is characterized by asymptomatic soft tissue swelling, specifically on the lateral aspect of the proximal interphalangeal (PIP) joints of the hands, and skin lesions. Its diagnosis and treatment requires expertise of various health professionals (rheumatologists, orthopedic surgeons, pediatricians, dermatologists, surgeons). Papers on PDD are published in journals that are not exclusively focused on dermatology, probably because patients with PDD require a multidisciplinary approach (Table 1). Two papers represent a comprehensive analysis of previously

published papers: the first published in 2009 by Beltraminelli et al. (2), and the second published in 2014 by Dallos et al. (3). The first paper analyzed 55 papers including 88 patients, while the other included 17 more papers, with a total of 121 patients reported until then. Our research found 27 more papers (41, 46, 48, 49, 54, 55, 57, 59, 62, 69, 70, 73, 74, 78, 79, 83, 85, 86, 88, 89, 92, 93, 95-98, 100) including 39 patients. So, since Bazex and associates to date, at least 99 papers including 160 patients have been published on PDD. Our case report is the 100rd, and our patient the 161st. These papers are listed in the references, mostly in order of publication, except for authors from the beginning of this paper.

The characteristic features of the disease, described in certain number of patients, are shown in Table 2, based on the table of Dallos et al. (3). Although PDD may be considered a rare condition, one must not neglect clinical features that are at the same time factors responsible for not recognizing the disease (62), such as the absence of subjective symptoms, self limiting course, regression during adolescence, absence of permanent sequelae. For this

Table 1. Types of journals reporting patients with pachydermodactyly

| Journal | No. of reports | References |
|--------------|----------------|------------|
| Dermatology | 60 | 2, 4-62 |
| Rheumatology | 18 | 3, 63-79 |
| Surgical | 4 | 80-83 |
| Pediatric | 6 | 84-89 |
| Orthopedic | 3 | 90-92 |
| Other | 8 | 93-100 |
| Total | 99 | 2-100 |

reason, the actual prevalence of PDD is estimated to be much higher (2, 3).

The disease commonly affects boys around puberty, as in our patient (2, 44, 76); PDD at older age may be due to non-recognition of the disease (3). The medical literature describes two cases of PDD since birth (13, 27), and only four cases in the elderly (36). The condition occurs sporadically, but familial cases have also been reported (8, 21, 24, 36, 42, 93), and in three cases it was a transgradient form of PDD (14, 19, 36).

The exact etiology of pachydermodactyly is unknown; it may include genetic predisposition, probably essential, associated with precipitating factors (3). It is probably due to repeated mechanical injury such as tics, stretching, rubbing or cracking of the fingers, both in healthy or people with obsessive-compulsive disorders (19, 20, 26, 35, 38, 39, 47, 51, 62, 84, 85, 96). PDD may occur due to repeated mechanical trauma in poultry processing workers, farm workers, those whose work is computer-related (52, 56, 58, 77); in athletes (football, handball, fencing) (61, 71), or musicians playing the guitar or flute (69); excessive use of computers by adolescent boys also contributes to appearance of PDD (53, 76). However, there are reports on patients with PDD unpreceded by friction or trauma (8, 10, 49, 74, 100). PDD is significantly more common in boys at the beginning of puberty compared to females and older

children, supporting the role of androgen hormones in its pathogenesis; the possible impact of the growth hormone and hypothyroid function should not be excluded (3).

Due to frequent occurrence of PDD in persons with obsessive compulsive and neurological disorders, there are authors who think that PDD should be on the list of skin manifestations of psychological problems (2, 26, 51). Our patient presented with a habit of rubbing and cracking of fingers, but had no psychological disorders.

Although a typical clinical picture includes benign asymptomatic soft tissue thickening on the lateral (ulnar and radial) proximal interphalangeal joints of the II, III and IV fingers of both hands, there are deviations, as in our patient whose both little fingers were affected, as well as the ulnar sides of both index fingers. Dorsal and/or ventral sides of fingers can also be affected (23, 29, 31, 94). Distal involvement has also been described affecting distal phalanges (37, 42, 45), as well as metacarpophalangeal joints (16, 40), with fibromatous thickening or rigid nodes (83). These lesions are usually symmetrical (88), but may be asymmetrical to a certain point, varying in the number and extent of affected joints on the left and right hand (76, 97) or affecting only some joints, the so-called localized form of PDD (13). There are reports on the monoarticular form of PDD (32, 65). The thumbs and fifth fingers are rarely

Tabela 2. Reported patients with pachydermodactyly

| Reference | Dallos T, et al. (3) | Current review |
|--|----------------------|----------------|
| Demographic data | | |
| No. of patients | n=121 | n=31 |
| male/female | (79)/(21) | 26 (84)/5 (16) |
| Disease characteristics | | |
| No. of patients | n=121 | n=22 |
| Disease onset: median age (y) (range) | 14 (5-74) | 13 (4-20) |
| Course of disease: median (y) (range) | 2.5 (0.25-15) | 3.5 (1-12) |
| Clinical signs and symptoms | | |
| No. of patients | n=121 | n=22 |
| Thickening: n (%) | 121 (100) | 31 (100%) |
| Restriction of movement (n %) | (2.5) | 0 (0) |
| Subjective complaints: n (%) | (5) | 0 (0) |
| Itching: n (%) | (2.5) | 0 (0) |
| Pain: n (%) | NA | 0 (0) |
| Etiology | | |
| No. of patients | n=121 | n=31 |
| Identified mechanical injury: n (%) | (44) | 12 (39) |
| Psychological and/or psychiatric diseases: n (%) | (15) | 8 (26) |
| Positive family history: n (%) | (5) | 0 (0) |

n, number of patients; y, year; NA, not available

affected (6, 8, 21, 50, 80, 99). Toes are never affected, whereas plantar pachydermia was described in a patient with PDD and acrocyanosis (39). Sometimes the skin shows lichenification, fine desquamation or hyperkeratosis, even moderate erythematosis (27),

but it is rarely painful (9, 10, 17). No cases with hypersensitivity, itching, burning, morning stiffness or reduced mobility have been reported (3, 48, 73, 91). Only one case of deforming PDD was reported with nonerosive interphalangeal joint subluxation

(72). Our patient presented with involvement of both lateral aspects, ulnar and radial parts of the proximal interphalangeal joints and proximal phalanges of the II, III, IV and V fingers of both hands, except the index fingers, where only ulnar sides were affected. The skin surface was slightly hyperkeratotic. He had no pain or functional limitations.

Pachydermodactyl has been described in association with Dupuytren's contracture (90), Asperger syndrome (38), Ehlers-Danlos syndrome (33), carpal tunnel syndrome and tuberous sclerosis (13, 33, 36), gynecomastia (10), foot syndactyly (2), acute atrophy maculosa varioliformis (25), and Tourette's syndrome (59).

Due to the heterogeneous clinical picture and described familial cases, a classification with five different forms was proposed (36):

1. classical pachydermodactyl (several proximal interphalangeal (PIP) joints affected, frequently associated with mechanical trauma)
2. localized (one joint affected - mono-pachydermodactyl)
3. transgrediens pachydermodactyl (extension to the palms or metacarpophalangeal joints)
4. familial pachydermodactyl (affecting several family members)
5. pachydermodactyl associated with tuberous sclerosis.

Diagnosis

Laboratory findings show no specific changes. Plain radiography shows soft tissue thickening, without bone, tendon or capsular changes such as periostosis, periarticular osteoporosis, erosions, cysts or osteophytes (3, 55). Magnetic resonance imaging reveals only soft tissue swelling, and typical fusiform swelling (without effusion, synovitis, tendonitis, hypervascularization and without bone involvement) (53, 57, 68, 70, 71).

Histologically, there is hyperkeratosis, acanthosis, thickening of the dermis, increase in fibroblasts and collagen deposits; increase in the thickness of basal membrane and of eccrine sudoriparous glands, intense deposition of mucopolysaccharides, poor demarcation between the papillary and reticular dermis, mucin deposition between collagen fibers (7, 11, 30, 34, 43, 53, 60, 70, 82, 83, 90, 95, 96). Types III and V

collagen are increased with a reduction of collagen type I. Electron microscopy shows an increased number of fine-diameter collagen fibers (21, 30, 34, 36), less uniform (12, 64). Some authors believe that histology is not mandatory for diagnosis (55), as well as many useless and expensive diagnostic tests (54, 89).

Chen and associates (99) proposed the following diagnostic criteria:

- the patient has no symptoms
- morning stiffness is absent
- pain on motion and tenderness to palpation is absent
- finger swelling is radial or ulnar in location, rather than circumferential
- laboratory test results are unremarkable
- plain radiographs show only soft tissue swelling.

With these typical findings, additional investigations, such as MRI or skin biopsy, are rarely needed to establish a diagnosis of PDD.

It is of great importance to distinguish PDD from other diseases of this localization. Differential diagnosis should include: rheumatic diseases, primarily juvenile idiopathic arthritis, rheumatoid arthritis, rheumatoid nodules; bone diseases such as secondary pachydermoperiostosis, ostitis cystoides multiplex Jüngling, spina ventosa; skin diseases, often knuckle pads with circumscribed keratoses that overly the finger joints with highly hyperkeratotic epidermis and hyperplastic dermal connective tissue, pseudo knuckle pads, foreign-body granulomas, collagenous plaques of the hands, infantile or juvenile digital fibromatosis, progressive nodular skin fibrosis, psoriatic acropachydermodactyl, connective tissue nevi; endocrine disorders, thyroid acropathy, acromegaly; tumors, primarily fibromas, sarcomas, paraneoplastic acropachydermodactyl; hereditary diseases, such as tuberous sclerosis, primary pachydermoperiostosis, Thiemann disease (3, 41, 55, 66, 67). PDD is commonly misdiagnosed as a rheumatologic condition (100), namely juvenile idiopathic arthritis (63, 69, 70, 78, 89, 86), which may lead to unnecessary treatment (78). Pereira and associates discussed similarities (mechanical irritation, favorable effects of intralesional triamcinolone injections) and differences (epidermal and dermal response) between knuckle pads and PDD (41).

Treatment

There is no effective medical treatment for PDD at this time (2, 25). Surgical excision of fibrous tissues may be a good therapeutic option (3, 46, 80) without recurrence (75). Good results are obtained using intralesional triamcinolone injections (10, 22, 100), while topical therapy with corticosteroids is mostly ineffective. Higuchi reported good therapeutic results after using tranilast, an anti-allergy agent which inhibits collagen synthesis in human skin fibroblasts (300 mg daily for 6 months) (92). Elimination of mechanical irritation may lead to spontaneous regression (2, 15, 18, 20, 71, 81, 91, 99). Patients with PDD should avoid mechanical irritation, receive adequate care and monitoring, whereas invasive procedures are not recommended (3, 74). Protective gloves and bandages are proposed to prevent mechanical irritation (44). Patients with obsessive compulsive disorders may need psychiatric help.

Pachydermodactyly is a benign condition with a chronic course. The prognosis is variable. After long-term progression, regression may occur at a later age (97).

Conclusion

We report a case of a male patient who developed symptoms of pachydermodactyly around puberty as a progressive soft tissue thickening around the proximal interphalangeal joints of all fingers except the thumbs probably due to stretching, rubbing or cracking of the fingers. There was no family history. Intralesional triamcinolone solution injections during three months resulted in slight reduction of swelling of the treated finger. The patient was advised to avoid mechanical trauma, rubbing and cracking of the fingers. A review of the available literature from 1973 to 2014 provided important insight into the characteristics and variations of the disease.

Abbreviations

- PDD - pachydermodactyly
- PIP - proximal interphalangeal
- MRI - Magnetic resonance imaging

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Pahidermodaktilija – prikaz slučaja i pregled literature

Sažetak

Uvod. Pahidermodaktilija (PDD) je benigna digitalna fibromatoza za koju je karakteristično asimptomatsko progresivno lateralno zadebljanje periartikularnog mekog tkiva, obično simetrično lokalizovanog oko proksimalnih interfalangealnih zglobova (PIP) drugog, trećeg i četvrtog prsta ruku: tako dolazi do simetričnog, oticanja kože dorzalnih i lateralnih strana proksimalnih falangi kažiprsta, srednjeg prsta i domalog prsta. Najčešće se javlja kod mladih adolescenata i verovatno je posledica ponovljene mehaničke stimulacije; nekad se javlja i kao posledica opsesivno-kompulzivnog poremećaja, ali se mora razlikovati od obsesivnog ‘chewing pads’ (eng. grickanje prstiju). Difuzno oticanje zahvaćenog prsta koje uključuje dermis pored epidermisa, jasno odvaja PDD od ‘knuckle pads’ (bokserski jastučići), kalusa, profesionalnih kalozita, itd. Opisani su slučajevi u kojima su oboleli članovi unutar iste porodice, PPD i ‘knuckle pads’ mogu biti istovremeno prisutni kod jedne iste osobe.

Prvi opis bolesti dali su Baseks (Baseks) i saradnici 1973. kao pachydermie digitale. Termin pachydermodactyly predložio je Verbov 1975. i potiče od grčkih reči pachy (thick – zadebljanje), dermo (koža) i dactilos (prst).

Prikaz slučaja. U radu je prikazan muškarac star 19 godina, po zanimanju student, koji je u 12. godini života primetio povećanje obima trećeg i četvrtog prsta leve ruke. Do kraja 14. godine bili su izmenjeni svi prsti sem palčeva na obema rukama i od tada je stanje uglavnom ostalo isto. Pojavu ovih promena povezao je sa udaranjem lopte, kao i sa tim da ima običaj da trlja i „lomi“ prste. Nama je upućen sa uputnom dijagnozom periartikularne artropatije posle pregleda ortopeda, reumatologa i fizijatra, a radi isključenja psorijaze i/ili seboroičnog dermatitisa kapilicijuma. Pacijent je na prijemu negirao postojanje drugih oboljenja i izjavio da u porodici nije bilo obolelih srodnika.

Opšte stanje pacijenta na prijemu bilo je u fiziološkim granicama. Dermatološki status je ukazao na: izmenjen oblik i obim svih prstiju obe šake, osim oba palca, u nivou proksimalnih falangi; normalan izgled distalnih delova svih prstiju; u nivou prvih interfalangealnih zglobova zadebljanje; hipertrofija, skoro simetrična na obe lateralne strane (ulnarna i radijalna) falangi

svih zahvaćenih prstiju, osim oba kažiprsta, gde je hipertrofija zahvatila samo ulnarnu stranu (Slika 1). Palpatorno, promjenjeni delovi su bili bezbolne, homogene teksture i elastične konzistencije, pokretni u odnosu na susedne koštane strukture. Svi zglobovi su pokazivali nepromenjenu funkciju.

Svi ispitivani relevantni laboratorijski parametri pokazali su vrednosti koje su bile u granicama referalnih vrednosti, uključujući faktore inflamacije, serumski nivo imunoglobulina, C3 i C4 komponente komplementa, prisustvo antinuklearnih antitela i imunoglobulina.

Ehosonografija zglobova šaka: osteoartikularne površine bez ehosonografskih znakova za defekte; ne uočavaju se periartikularne fokalne promene, osim inbibicije mekih tkiva; sinovije medijalnih interfalangealnih zglobova zadebljane; povećanje intraartikularne sinovijalne tečnosti.

RTG šaka u oba pravca: koštana građa prvog reda očuvana (Slika 2).

Patohistološki nalaz: u epidermisu izražena hiperkeratoza i akantoza; epidermo- dermalna granica zaravnjena; dermis zadebljao, hipocellularan, retki fibroblasti okruženi su širokim hijalinizovanim trakama kolagene strome; adneks kože odsutan.

Intralezionalno ubrizgavanje rastvora triamcinolona u toku tri meseca dovelo je do neznatnog smanjenja obima tretiranog prsta. Savetovali smo pacijenta da izbegava mehaničke traume, trljanja i „pucanja“ prstiju.

Diskusija i pregled literature. Kako je PDD oboljenje koje se javlja u pubertetu i manifestuje se uglavnom asimptomatskim promenama u predelu proksimalnih interfalangealnih zglobova (PIP) ruku u vidu uvećanja tkiva oko zglobova, sa mogućim promenama na koži, to su u dijagnostiku i lečenje uključeni lekari različitih specijalnosti (reumatolozi, ortopedi, pedijatri, dermatolozi, hirurzi). Radovi koji se odnose na PDD, objavljeni su i u časopisima koji nisu isključivo usmereni samo na dermatologiju, a to se može tumačiti multidisciplinarnim pristupom obolelima (Tabela 1). Dva rada predstavljaju opsežne analize do tada objavljenih radova: prvi iz 2009. objavili Beltraminelli (Beltraminelli) i saradnici, a drugi su

2014. godine objavili Dalos (Dallos) i saradnici. U prvoj studiji izvršena je analiza 55 radova sa 88 bolesnika, dok je druga obuhvatila još 17 objavljenih radova, sa ukupno 121 bolesnikom registrovanim do tada. Novim pretraživanjem registrovali smo još 28 radova sa 45 obolelih. Znači, od Baseksa i saradnika do danas, objavljeno je o PDD najmanje 100 radova u kojima je prikazano 165 bolesnika. S našim prikazom, to bi bilo 101 rad sa 166 bolesnika.

Odsustvo subjektivnih tegoba, samolimitirajući tok, zaustavljanje progresije oboljenja u adolescentnom dobu, odsustvo trajnih sekvela, predstavljaju faktore zbog kojih se pretpostavlja da je stvarna prevalencija PDD realno viša, te da PDD nije tako retka. Bolest se najčešće javlja u ranom pubertetu kod osoba muškog pola, kao što je to slučaj kod našeg pacijenta: u starijem uzrastu, može biti posledica neprepoznavanja oboljenja. U literaturi su opisana dva slučaja bolesti od samog rođenja i samo četiri slučaja kod starih osoba. Bolest se javlja sporadično, ali su opisani i familijarni slučajevi, a u tri slučaja se radilo o transgredijentnoj formi PDD. Etiologija bolesti nije u potpunosti razjašnjena: na moguću individualnu genetsku predispoziciju, koja je verovatno esencijalna, nadovezuje se više precipitirajućih faktora. Najznačajnija je ponavljanja mehanička trauma usled pokreta sličnih tikovima, kao što je istezanje, trljanje ili „pucketanje“, „lomljenje“ prstiju, bilo da se radi o zdravim osobama ili onima sa opsativno-kompulzivnim poremećajem. U literaturi su objavljeni slučajevi PDD čijoj pojavi nisu prethodile ni trauma ni frikcija. Značajno češća pojava PDD kod dečaka na početku puberteta u odnosu na osobe ženskog pola i starijeg uzrasta, ukazuje na patogenetske uloge androgenih hormona; ne isključuje se i mogući uticaj hormona rasta i tiroidne hipofunkcije). Zbog česte pojave PDD kod osoba sa opsativno-kompulzivnim i neurološkim poremećajima postoje zagovornici da se PDD stavi na listu kutanih manifestacija psiholoških poremećaja. Kod našeg pacijenta je postojala navika da trlja i „lomi“ prste, ali nije imao nikakve psihogene poremećaje.

Pored tipične kliničke slike, mogu biti zahvaćene i dorzalne i/ili ventralne strane prstiju. Opisana je i distalna varijanta, sa zahvatanjem distalnih falangi i metakarpofalangealnih zglobova, sa fibromatoznim zadebljanjem ili sa čvrstim nodusima. Promene su obično simetrične, ali mogu biti asimetrične u raznom

stepenu, od različitog broja i stepena zahvaćenih zglobova na levoj i desnoj ruci do zahvatanja samo pojedinih zglobova u tzv. lokalizovanoj formi PDD. Opisana je i monoartikularna varijanta PDD. Peti prst i palac su retko zahvaćeni; prsti nogu nikada nisu zahvaćeni, a plantarna pahidermija je opisana kod bolesnika sa PDD koji je imao akrocijanozu. Koža na obolelim prstima može biti lihenificirana, sa lakovom deskvamacijom ili hiperkratozom, nekad i umereno eritematozna, retko bolna. Nije registrovana pojačana osetljivost, svrab, žarenje jutarnja ukrućenost ili redukovana pokretljivost. Objavljen je samo jedan deformantni slučaj PDD sa neerozivnom sublukasacijom interfalangealnog zgloba. Kod prikazanog bolesnika bila su zahvaćena oba lateralna aspekta, ulnarni i radijalni delovi kože proksimalnih interfalangealnih zglobova i proksimalnih falangi II, III, IV i V prsta obeju ruku, sem kažiprsta, gde su promene bile samo na ulnarnim stranama. Površina kože je bila lako hiperkeratotična. Nije imao poremećaj funkcije, niti bolove.

Zbog heterogenosti kliničke slike i opisanih familijarnih slučajeva, izvršena je klasifikacija bolesti na pet tipova: 1. klasični tip (zahvaćeno više PIP kao posledica mikrotraume); 2. lokalizovani tip (zahvaćen jedan zglob); 3. transgredijentni (zahvata metakarpofalangealnu regiju); 4. familijarni (zahvaćeno više članova iste porodice) i 5. udružen sa tuberoznom sklerozom.

Prema Čenu (Chen), dijagnostički kriterijumi su: pacijent je bez simptoma; jutarnja ukrućenost je odsutna; bol pri pokretima i osetljivost na palpaciju su odsutni; zadebljanje prsta je radijalno ili ulnarno, retko cirkumskriptno; laboratorijski testovi nisu značajni; radiografski se nalazi samo zadebljanje mekog tkiva. Sa ovim tipičnim nalazom dodatna istraživanja retko su potrebna za postavljanje dijagnoze PDD.

U lečenju PDD nema efikasnog medikamentnog tretmana. Operativnom resekcijom fibroznog tkiva postignuti su dobri rezultati bez recidiva. Dobre rezultate dala je i intraleziona aplikacija triamcinolona, dok su pokušaji lokalne aplikacije topikalnih kortikosteroida uglavnom ostali bezuspešni. Higuči (Higuchi) je objavio dobre rezultate posle primene antialergijskog leka tranilasta, koji izaziva inhibiciju sinteze kolagena u koži, u dozi od 300 mg dnevno u toku 6 meseci. Eliminacija mehaničke stimulacije može dovesti do spontane regresije. Bolest je benignog karaktera sa hroničnim tokom. Prognoza je varijabilna.

Moguća je stabilizacija u kasnijim godinama posle višegodišnje progresije.

Zaključak. Prikazana je osoba muškog pola kod koje se pahidermodaktilija manifestovala na proksimalnim falangama svih prstiju osim palčeva obe šake kao posledica trljanja i „pucketanja“ zglobova prstiju. U

porodici nije bilo obolelih. Intralazona aplikacija triamcinolona na jednom prstu dovela je do poboljšanja. Dat je savet da se prekine sa mehaničkom traumom. Pregled dostupne literature od 1973. do 2014. godine omogućio je bolje sagledavanje kliničkih varijanti oboljenja.

Ključne reči

Fibrom; Dermatoze šake; Deformateti šake; Prsti; Neoplazme mekih tkiva; Kumulativni traumatski poremećaj