PTERYGIUM UNGUIUM. DISCUȚII DESPRE TREI CAZURI ATIPICE

PTERYGIUM UNGUIUM. DISCUSSIONS ON THREE UNUSUAL CASES

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Rezumat

În lucrarea de față prezentăm 3 cazuri cu Pterygium unguium, o distrofie unghială nu prea des recunoscută. Cazurile sunt: epidermoliza buloasă dobândită, epidermoliza buloasă joncțională forma benignă și sindromul nail-patella, cel din urmă asociat cu multiple displazii osteao-articulare și glomerulonefrită cu depozite membranare. Am luat în discuție patogeneza bolilor mai sus-menționate și implicarea în dezvoltarea distrofiilor unghiale de tip pterygium unguium, cât și importantă pentru dermatolog dar și medici de alte specialități a cunoașterii semnificației onicodistrofiilor.

Cuvinte cheie: *pterygium unghuium, collagen, displazie, autoimunitate.*

Summary

We present 3 patients with Pterygium unguium, a not so common dystrophy of the nail. Their diagnoses are: Epidermolysis bullosa aquisita, junctional Epidermolysis bullosa, benign form and Nail-Patella Syndrome, the last one being associated with multiple osteo-arthro dysplasies and glomerulonephritis with membranar deposits. We discuss about the pathogenesis of the diseases mentioned above and its implication in the development of pterygium unguium as well as the importance of onychodystrophies for the physician.

Key words: *pterygium unguium, collagen, dysplasies, autoimmunity.*

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Introduction

Dorsal pterygium unguium (PU) consists of a gradual thinning of the proximal nail fold with associated extension of the cuticle over the nail plate. The nail plate becomes fissured because of the fusion of the cuticle to the matrix and subsequently to the nail bed; its split portions progressively decrease in size as the pterygium widens. This often results in two small nail remnants if the pterygium process is central. Complete involvement of the matrix and nail bed in the pathological process leads to total loss of the nail plate, with permanent atrophy and sometimes scarring in the nail area. Dorsal pterygium is characteristic of lichen planus and, less often, of peripheral vascular ischaemia. It also occurs in severe bullous dermatoses, radiotherapy and on the hands of radiologists; it may follow injury; rarely congenital forms have been described.

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Ventral pterygium, or pterygium inversum unguis, is a distal extension of the hyponychial tissue which anchors to the under surface of the nail, thereby eliminating the distal groove. Scarring in the vicinity of the distal groove, causing it to be obliterated, may produce secondary pterygium inversum unguis. Ventral pterygium may be seen in scleroderma associated with Raynaud's phenomenon, disseminated lupus erythematosus, and causalgia of the median nerve. The condition may be idiopathic, congenital, and familiar or aquired. A congenital, aberrant, painful hyponychium has been described associated with oblique, deep fractures of the nails. There are recent reports of familial forms of the disease. In one case an unusual aquired association of pterygium inversum unguis and lenticular atrophy of the palmar creases was recorded [1].

Subungual pterygium (non-inflammatory) is analogus to the claw of lower primatess.

In patients suffering from dorsal pterygium (excluding the traumatic or congenital types) the main characteristic is a dilatation in the nail fold capillary loops and the formation of a slender microvascular shunt system in the more dilated loops. These changes are visible by capillaroscopy.

Case Reports

Case1. Female patient 66 years old, for 3 years in our evidence, has been hospitalised , several times, for a disseminated bullous eruption involving the entire skin, the acral areas being most affected. The bullae are accompanied by erosions, atropho-scarring lesions on the dorsum of the hands and on the friction areas; nail dystrophies like onychomadesis during erruptive periods and dorsal pterygium unguium involving the second and third fingernails from both hands; millium cysts on the dorsum of the fingers and the flexor areas of the forearms.

Her family history is negative for any bullous or other diseases with clinical significance.

Her personal history reveales seven pregnancies from which three ended with cesarean section, six children died, from unknown causes, aged between three days and ten months.

In April 1991 she accused pruritus accompanied by erythematous macules and later bullae, initially on the traumatised and friction areas; the evolution was towards scar formation and nail dystrophies: onychomadesis, pterygium unguium dorsalis, anonychia and onychatrophy involving the toes. Without a proper treatment the eruption generalised. The clinical examination revealed a hypostatural type.

The local examination revealed, on the forearms, thigs, hands, feet, calves, the anterior and posterior thorax, spontaneous and posttraumatic bullae, of 0.5-5 cm in diameter. The bullae were big, flat, some of them with haemorrhagic content, the Nikolski sign was positive, evolution was with erosions and scar formation. She presented also a diffuse alopecia; the teeth were abnormal, badly implanted, the palatal vault was high(ogival).

During time the disease extended, new bullae, having the same features, appeared periungual, in the submamary folds, arm pits and crural areas. Anonychia of the second and third fingers from feet was observed during evolution as well as-on the hands- onychomadesis accompaning active phases with nail shedding and pterygium unguium dorsalis on the second and third fingers-without any symmetry; millium cysts were obsered on the dorsum of the fingers.

The paraclinical investigations performed revealed:Hb:10.75g%;WBC:5200; ESR: 80 mm\h; fibrinogen: 650 mg%; gamma globulines 23%; C3 108 mg%; Lta: 36-44%.

The Tzank smear didn`t show any acantholytic cells; AAN were absent. Skin biopsy revealed subbazal split, the inflammatory cellular infiltrate consisted of: lymphocytes, eosinophiles, hystiocites, neutrophils, was located on the bullae`s floor and superficial dermis.

The diagnoses was: Epidermolysis bullosa aquisita (EBA)-dermolytic pemphigoid.

She was treated, locally, with antibiotics and phenitoin; the sistemic treatment consisted in corticosteroids, azathioprin, cyclosporin. She was told to avoid mechanical trauma. For a short period the evolution was favorable, in time the erosions relapsed and the scarring process continued.

Case 2. Female patient, L.A., 23 years old, presented herself to the dermatologist for a variety of dystrophies of the fingernails. From her history we note that the alterations of the hands` fingernails have been presented since birth.

The nail dystrophy consisted of complete absece of fingernails at both tumbs, pterygium unguium and onychatrophy at fingers 2, 3, 4 of both hands and the fifth from the rigt hand. The nail from finger 5 of the left hand was normal.

She followed ambulatory treatment with Fe, Ca and vitaminotherapy but with no results.

The family history didn`t reveal anything important and from her personal history we underline the existence of reccurent dyslocations of the left knee`s patella, for which she suffered a surgical procedure at the age of 10.

Clinical examination revealed a hypostatural (154 cm) female patient but with normal psycho-somatic status. She cannot totally extend the elbow to straighten the arm and has difficulties concerning the movements of supination and pronation from the elbow joint. The patient presented also status post reccurent dyslocation of the left patella, surgically cured.

The nail`s modifications consist of a large variety of lesions:

- the thumbs` fingernails were absent and replaced by pterygium unguium dorsalis with two fragments of onychatrophy;
- the nails from the fingers 2, 3, 4 from both hands and fifth from the right presented pterygium unguium dorsalis and onychatrophy of decreasing severity from index to the small finger;
- the nail from the fifth finger of the left hand is normal;
- fingers 2, 3, 4 from both hands presented an axial deviation;
- the feet` fingernails were normal, without any lesions in the patient`s history;

From the paraclinical investigations that had beed performed (WBC, RBC, ESR, proteins, serum electrophoresis, glucose blood level, thymol, serum ALAT, ASAT, calcium blood level, urine analysis, Addis sediment, VDRL) we noticed AII blood group, eosinophilia of 6%, sideremia of 28.2 umol\l(normal values 6.6-26 umol\l), proteinuria under 200 mg\24 h.

X-rays examinations revealed the following changes:

1. X-ray of the hands

Swelling of soft tissue; the right fingers 2, 3 and left fingers 3, 4 presented an axial deviation at the level of proximal interphalangeal joint; cubital deviation of the metacarpophalangeal joint- left finger 3,4; osteosclerosis at the proximal apophysis- first phalanx right hand, the second phalanx of the fifth fingers are shorter; structural changes of the carpal bones, especially of the semilunar and left big bone with periarticular osteosclerosis; demineralisation of the distal epyphisiss of the right radius; absence of the styloidee cubital apophysis- bilaterally.

2. X-rays of elbows and underarms

More gracile bone segments with demineralisation of the distal epyphisis of the humerus; spotted demineralisation of the proximal epyphisis of the radius and cubitus- bilaterally; hypoplasia of the radial head and his dyslocation in the joint; altered head position with the olecran having a medial position, the radial head being extremely deviated.

3. Chest x-rays

Modification of aspect and position of the acromial processes, wich present a strong oblicity and show medial deviation, the left having a greater projection surface; assymetry of the acromial processes, the left being greater with a smaller glenoid surface than the humeruse`s head; thickening of the external edge of the scapula which is more convex; slight acromio-clavicular dysjunction on the right side;

4. X-rays of the pelvis

Bilaterally periarticular osteosclerosis, especially concerning the iliac side; the growing cartilages of the iliac crests are not completely linked; sacralisation of the L5; spina bifida; coxa valga of the femural heads which are more gracile; iliac wings are unfolded with the hypertrophy of the postero-inferior iliac spinas.

5. X-rays of the knees

Demineralisation of the periarticular segments with more opaque trabecular bone through the

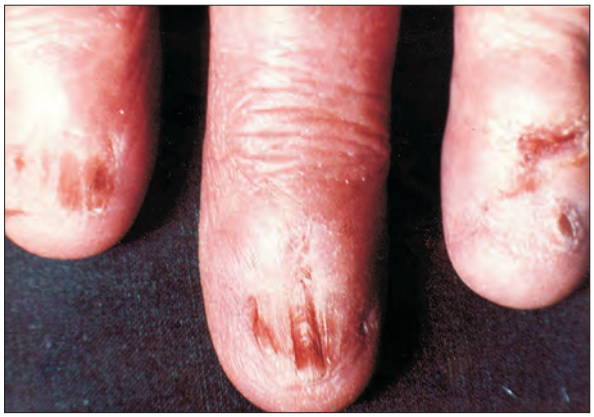

Fig. 1. Pterygium unguium dorsalis, onychoatrophy and anonychia in patient with Junctional Epidermolysis Bullosa benign form

force lines; hypoplasia of the both patellas and of both femural lateral condyles, the left lateral tibial spine is flattened; more evident inter-condilian cavity, slightly demineralisated at it`s border.

6. X-rays of the ankles

Periarticular demineralisation, with more opaque trabecular bone structure along the force lines which continues from the distal epyphisis of the tibia to the astragal; a few small areas of lysis at the distal epyphisis of the right fibulae.

7. X-rays of the feet

Demineralisation of the exposed segments, especially of the periarticular surfaces, metatarsial and phalangeal 2, 3, 4, 5 bilaterally; area of lysis at the proximal right epyphisis of the third metatarsian.

The diagnosis was Nail-Patella syndrome (NPS).

Case 3. M.M, 24 years old female patient is in our evidence with Junctional Epidermolysis Bullosa (JEB)-benign form. Since childhood she has been presented serohaemorrhagic bullae on the sites of minor trauma with an atropho-scarring evolution and with shadding of the nails.

Her family history revealed a brother and a sister diagnosed with Epidermolysis bullosa, unknown type. Her personal history didn`t reveal anything important.

The clinical examination: sero-haemorrhagic bullae on the sites exposed to minor and repeated trauma and on both knees, the pain beeing

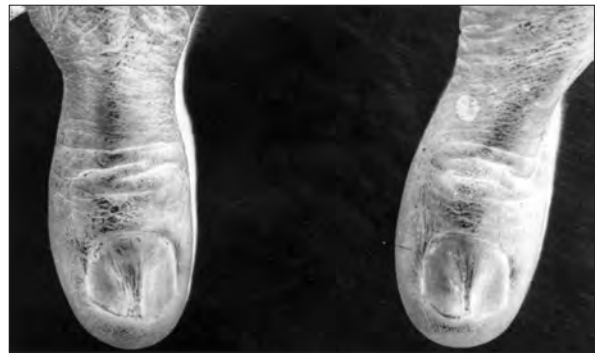

Fig.2 Pterygium unguium dorsalis in patient with Nail-Patella Syndrome

absent. They disappear in few days leaving atrophic, white, thinned, pleated- like onion skin scars. The feet`s nails are absent the nails from the hands showed: dorsal pterygium, a scar from the proximal to the distal edge replacing the nail plate (onychoatrophy, anonychia). The fingers are thin, sclerodactylic (Fig .1).

The biological evaluation was normal, measurement of T3 and T4 excluded hyperty-roidism. She was treated with: isoprinosine, phenitoin; topically with antibiotics. The patient presented a prolonged depressive reaction.

She is still in our observation.

Discussions

The three cases presented above-EBA, NPS, JEB- have, as a common sign, major nail dystrophies, especially pterygium unguium, which can point out anomalies as a part of a syndrome- like in NPS case.

The onychodystrophy described in our second case is typical for the NPS (Fig.2). It consists of lesions in which gravity decreases from the radial extremity to the cubital one of the hand. We mention that fingers 2, 3, 4 from both hands present a deviation of the axes which gives them a peculiar aspect. Although we mention that the nails from the feet are normal.

The dysplasia of the fingernails has been presented since birth or since the first years of childhood, in a great number of patients. Occasionally only the ulnar side of the nails has been affected. Even if the nail is totally absent the nail bed is still present [2] The capillaroscopy

showed, in pterygium dorsal, a major characteristic, a dilated capillary loop in the nail fold and the formation of a mycrovascular system between the dilated loops.

Other abnormalities that have been described in NPS are: onychoschysis, hyponychium, anonychy and triangular lunula.

The second major feature present in our case is the bone dysplasia of the knee with hypoplasia of the patella and of the lateral femoral condyles; our patient had suffered a surgical cure for recurrent luxation of the left patella, frequent in these patients [3].

The bone dysplasia of the elbow consisting in: hypoplasia of the capitelum, and of the radial head, that didn`t permit, in our case the complete extension of the elbow joint, as well as difficulties of the supination and pronation movements in this articulation.

Another characteristic feature of the NPS would be the presence of iliac horns from the posterior iliac crests. This iliac horns were discovered many times by chance at an X-ray examination, but in our case they were missing. We still observed the fact that the growing cartilages of the iliac crests were not completely linked, fact that makes us consider the possibility of the appearance of iliac horns in the evolution of the case.

There were described other bone dysplasias associated to NPS, also reported: the congenital absence of the fibula, shoulder dysplasia, thickening of the lateral side of the scapula, spina bifida as well as a dysplasia of the superior epyphisis of the radius, partially present in our case. Our case presented other bone dysplasias not mentioned before in the literature: the absence of both cubital styloide apophysis; the asymmetry of the acromial processes with a more reduced glenoid cavity, as well as the absence of the iliac horns, despite of the aspect of the one pelvis which is typical for the NPS.

The inheritance of the syndrome is autosomal dominant with complete penetration and variable expressivity, but there were not excluded some translocations. The linkage is possible between the locus that controles the NPS and that of the blood group [4].

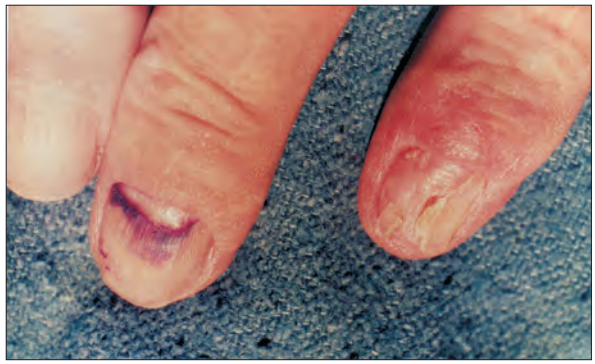

Fig. 3. Pterygium unguium dorsalis and onychomadesis in patient with Epidermolysis Bullosa Aquisita

In association with the NPS there has been described a characteristic glomerulopathy that consists in deposits of a collagen like material in the glomerular basement membrane which is evidenced by electronooptic microscopy [5]. Proteinuria is asymptomatic in 60 % of cases, and in 5.5%- 8% of cases the disease leeds to renal failure and the necessity of hemodialysis. Our case presented only physiological proteinuria, less than 200 mg\ 24 h. The presence of the syndrome suggested, in our patient, a renal biopsy, but it was not performed (no acceptance from the patient). In one case Korting and Gerhart evidenced increased excretion of muccopolysacharides.

Other abnormalities of the uro-genital system were: renal dysplasia, double ureter, renal failure, nephrotic syndrome, Goodpasture syndrome [6, 7, 8].

Ocular disorders that had been described: heterochromia of the irises, glaucoma, mycrocorneea.

Returning to the pterygium unguium diagnosed in our first case: it is asymmetric, isolated, involves only one or two hand`s fingernails, beeing linked to the erruptive periods of EBA (Fig.3). The autoantibodies against 290 kD antigenic fraction of collagene are responsible for autoimmunity in EBA, their migration is similar to that seen in Bullous Lupus Erythematosus.

A renal biopsy with immunoelectron-microscopy would be of interest, knowing the fact that glomerular involvement is frequent in SLE.

EBA is an aquired immunological disorder unlike NPS or JEB. The last one presents severe nail dystrophies and loss of the nails since infancy. In our case the hands` fingernails show a dorsal pterygium unguium- a scar between the proximal and distal edges, without the presence of any nail fragments (onychatrophy, micronychia) - as we see in our second case- NPS. In JEB benign form, the target are structures of lamina lucida. The cause of nail dystrophies and renal impairment may be due to structural similarities between the two basal membranes . In both cases (JEB, NPS) the patient is hypostatural.

We have met, also, in the context of an Lichen planus without hepatic involvement (HBs Ag. and Anti-HVC Ab. were negative), a case of sistematised pterygium unguium affecting the hands` fingernails; here the scar splits the nail plate in the middle, fingernails from both hands being involved. Cutaneo-mucous lesions cured with a proper treatment- the nails` aspect hadn`t been influenced, and once again the toe nails hadn`t been affected in this context.

Another case of dorsal pterygium unguium has been observed in a different dermatological context, Urticaria vasculitis. The second, third and fourth fingernails from the right hand were affected by a median split between the proximal and the distal edges, the nail plate being divided in two equal parts, without any tendency to micronychia or onychoatrophia. The nail dystrophies were precipitated by a trauma which affected, probably, the vascularisation of the matrix, inducing, by means of duration and severity, a scar , the matrix becoming unable to fulfill the generating function .

The nail disorders, especially the dorsal pterygium unguium, have to draw the dermatologist`s attention because its major implications in patient`s prognosis.

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