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Rodica-Elena Perciun, Anuța Telcian

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Case Report: This clinical case of a 50-year-old insulin-treated female, presented with plantar and palmar fibromatosis (Ledderhose disease and Dupuytren disease), cheiroarthropathy, knuckle pads (Garrod's pads), Huntley's papules, acanthosis nigricans and large dorsal cutaneous scleredema. Diagnosis has engaged clinical and imaging correlation. Planning the therapeutic approach has been adapted accordingly.

Conclusion: In this case, the soft tissue's abnormalities have challenged our clinical and imaging skills as well. Long-term follow-up is claimed due to the diabetic condition and the potential aggressiveness of lesions.

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Keywords: Diabetes mellitus, Insulin-treated, Soft tissue pathology, Therapeutic approaching

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INTRODUCTION

Some soft tissue (ST) abnormalities are frequently seen in diabetic individuals, rather in isolation than in association. A non-invasive imaging becomes undoubtedly helpful because they often present clinically undetectable features [1]. Certain ultrastructural similarities suggest a potential identical root at least for superficial proliferative disorders (palmar and plantar fibromatosis, Dupuytren disease, knuckle pads, Peyronie disease or keloids) [2]. We are describing a 50-year-old diabetic insulin-treated female presenting bilateral Ledderhose's disease (LD) and Dupuytren disease (DD), frequently referred to as plantar and palmar fibromatosis. The knuckle pads, cheiroarthropathy, scleredema and acanthosis nigricans (AN) (insulin-resistance cutaneous marker) were diagnosed too.

In our opinion, this case with dysmetabolic grounds and multiple clinical ST abnormalities required a careful consideration and complementary imagistic approach for diagnosis. To our knowledge no parallels between clinical and imaging studies have been performed in a similar case. An original aspect consists in some 'histological' related data that high frequency ultrasound (HFUS) described.

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CASE REPORT

Our patient is a 50-year-old female with a 12-year history of insulin-treated diabetes mellitus (DM). A microvascular disease (incipient retinopathy, nephropathy and neuropathy) goes along with hypertension, hypercholesterolemia, autoimmune thyroid disease and obesity. Neither notable infectious fever, tightness in neck and shoulders nor a history of familial LD, drinking or phenytoin treatment have been recorded. Her current medication included hypotensors (ACE-inhibitors), hypolipidemics (statins), thyroid hormones substitutes (euthyrox), bed time basal insulin (glargine) and metformin. Blood analysis was within accepted ranges, excepting the mean of HbA1c which was, for years, 7.9%. No monoclonal gammopathy evidence was found. Three years ago she had been exclusively clinically examined for the left plantar mass having no more than 1.5 cm diameter, but never for skin pathology either in hands or trunk.

Clinically, we have noticed bilateral medial mid-third LD. On the left foot an irregular, non-inflamed, more prominent mass (about 3.5 cm diameter) without contractures of toes was described. (Figure 1). The 10 MHz gray scale and Doppler ultrasound assessment, as conventional ultrasound (CUS), has shown various aspects according with sequences. The split screen transversal scan (left plantar lesion) showed a pseudolobular image of 1.9/1.1 cm without vascularity (Figure 2). The split screen longitudinal view depicted an inhomogeneous (left) and vascularized echostructure (right) (Figure 3). A split screen plantar image showed a cross-sectional pseudocystic echostructure of 1.5/0.8 cm and other two infracentimetric isoechoic nodules (arrows) (left); an oblique longitudinal view exhibited its largest dimensions (3.1/1.8 cm) (right) (Figure 4). The magnetic resonance imaging (MRI) scan detailed its heterogeneity (various cellularity and collagen content), size and invasiveness (fascial tails, cutaneous and subcutis) (Figures 5A–C).

The patient also presented an incipient, bilateral form of DD with small nodules beneath the palmar skin without fingers' contracture yet (Figure 6). The co-existence of limited joint mobility (LJM) affecting the patient's hands (cheiroarthropathy) was proven by the presence of the 'prayer sign'. The thickened tight skin determined the hand stiffness impairing the extension of interphalangeal (IP) and metacarpophalangeal joints (MCP) (Figure 7). Conventional ultrasound depicted an irregular hypoechoic subcutis nodule as DD's anatomical substructure (Figures 8 and 9). The hypoechoic thickened flexor tendon sheath (1.5 mm vs and <1 mm the normal) defined the cheiroarthropathy substructure, here in association with DD (Figure 9). High frequency 20 MHz Derascan C (Cortex technology) (HFUS) penetrates as deep as 2.5 cm. The echogenicity emerges from the low (LEP), medium (MEP) and high echogenic pixels (HEP). The normal volar skin shows the epidermis (E) - 0.29 mm, dermis (D) - 2.05 mm, integument (E+D) - 2.34 mm

with intra-dermal scattered echos and the underlying hypoechoic subcutis (SC) (Figure 10A). Using a similar gain, the affected skin showed E- 0.53 mm with bilaminar intensively hyperkeratosis echostructure, thus preventing the deeper dermal scanning (Figure 10B). However, enhancing the gain curve (to compensate the posterior attenuation artifact), we detailed: the subepidermal low echogenic band (SLEB) and integument thickness of 4.5 mm being more hypoechogenic than normal either in upper dermis (UD) (84% vs 45%) or lower dermis (LwD) (94% vs 61%) (Figure 10C).

Another ST abnormalities are exhibited, over the dorsum of the IP and MCP as firm, painless nodules named 'knuckle pads' with tiny Huntley's papules on ('cobblestone') (Figure 11).

Split screen (gray-scale CUS) image showed the normal MCP dorsal subcutis (left) and the heterogeneously hypoechoic, thickened counterparts within overgrowth knuckle pads (right) (Figure 12). HFUS scanned the normal E-0.13 mm compared with intensively hyperkeratosis thickened E-0.28 mm, not allowing the deeper penetration (Figure 13A–B).

Despite the lack of complaints, the upper thorax dorsal area exhibited an erythematous, indurated skin suggesting scleredema. (Figure 14). The lower blue "X" painted sign conveys to the thickest cutis area of 8.1 mm that CUS expressed (Figure 15). Other areas measured no more than 4.8 mm.

Acanthosis nigricans (AN) mimicked a dark velvety appearance over some flexural areas as shown by the patient's dorsal neck flexure (Figure 16). HFUS scanned the normal skin (sex, age, and anatomical site counterparts), scleredema and AN integuments to be compared. The normal cutis with E-0.20 mm, D-3.18 mm, integument-3.34 mm expressed a dominant medium/low echogenic UD and slightly hypoechogenic LwD (Figure 17A). Scleredema exhibited thicker layers: E-0.29 mm, D-5.21 mm, integument-5.5 mm with hyperechogenic UD and dominant hypoechogenic LwD (Figure 17B). Acanthosis nigricans showed irregular thickened E- 0.39 mm, ill-defined E-D junction, intensively hyperechogenic D-3.17 mm (slightly regressive in the LwD) and thickened integument-3.56 mm (Figure 17C).

DISCUSSION

Usually, the ST pathology in diabetics can be clinically diagnosed. Nevertheless, some abnormalities might be overlooked if attention is not paid.

There is an agreement over the relationship between the superficial fibromatosis entities (LD, DD, knuckle pads and Peyronie's disease) and the long-standing poorly controlled DM associated with microvasculopathy [1–3]. Although, the DD and LD have different clinical, temporal and prevalence exposures, some histochemical and ultrastructural similarities connect them to the same root [2]. Nevertheless, some particular histology might also be found [4].

Figure 1: Clinical aspect of bilateral medial mid-third plantar fibromatosis; on the left foot an irregular, more prominent but noninflammatory mass can be described.

Figure 2: Split screen image-B-mode conventional ultrasound (CUS) of the left plantar fibroma. The transverse image depicts a pseudolobular lesion of 1.9/1.1 cm without Doppler signal.

Figure 3: Split screen image (CUS) of the left plantar fibroma. These longitudinal gray-scale (left) and color Doppler (right) images depicted other anatomical sequences heterogeneously echostructured.

Figure 4: A split screen plantar image showing a cross-sectional pseudocystic echostructure of 1.5/0.8 cm and other two infracentimetric isoechoic nodules (arrows) (left); an oblique longitudinal view exhibited its largest dimensions of 3.1/1.8 cm (crosses and arrows) (right).

Ledderhose disease (LD) is a benign fibroproliferative disorder usually within medial and central aspects of aponeurosis plantaris, being bilateral up to 60%. The patient has developed bilateral, asymmetrical non-inflammatory nodules located within medial mid-thirds. The patient's complaints consisted in a palpable lump and pain while walking, mainly because the left

Figure 5: MRI assessment of the left lesion (encircled) detailed its heterogeneous signal (various cellularity and collagen content), size and invasiveness (fascial tails, cutaneous and SC). (A): Sagittal T1-weighted sequence showing the lesion with some small foci low signaled, (B): Short axis T1-weighted post contrast sequence, (C): Sagittal T1-weighted fat saturation post contrast sequence depicting the same enhancing lesion.

Figure 6: Photograph of the incipient palmar fibromatosis (DD) (encircled) along with cheiroarthropathy

lesion's size increased for the last three years. According to literature, LD is more confidently diagnosed when clinic and imagistics corroborate [2, 5]. CUS scanned a multifaceted potentially aggressive lesion asking for MRI interrogation. The heterogeneous structure with some low signaled foci suggested hypocellularity and higher collagen amount in contrast to the rest of the mass. The differentials from neurofibroma or fibrosarcoma should be considered but MRI scan is helpless in differentiating

Figure 7: Clinical positive 'prayer' sign of cheiroarthropathy.

Figure 8: Gray-scale conventional ultrasound showing irregular hypoechoic subcutis nodule (white arrows) of palmar fibromatosis.

LD, clear cell sarcoma or desmoid tumor on the basis of their high signal intensity on T-weighted images [6]. Generally, when lesions are disabling the biopsy, excision, chemotherapy or radiotherapy should be considered [2]. Our patient rejected any other diagnostic and therapeutic maneuvers.

The connective tissue is probable irreversible damaged by chronic hyperglycemia. This process is consistent

Figure 9: Gray-scale conventional ultrasound evidence of cheiroarthropathy hypoechoic thickening of the flexor tendon sheath of 1.5 mm (white arrows and crosses). An oval shaped hypoechoic subcutis mass is the substructure of the associated palmar fibromatosis (colored arrows).

Figure 10 (A): Normal palmar skin: E-0.29 mm, Integument-2.34 mm, (B): The cheiroarthropathy bilaminar, hyperkeratosis and thickened E-0.43 mm (similar gain) thus preventing the deeper dermal scanning, (C): Gain enhancement imaging shows: A thickened hyperkeratosis E-0.57 mm, an underlying thin hypoechoic band of edema (SLEB) (arrow), a thickened integument-4.5 mm more hypoechoic either in UD or in LwD.

Figure 11: Photograph of the hands with knuckle's pads (Garrod's nodules) over their interphalangeal (IP) and metacarpophalangeal (MCP) dorsal aspects (arrows) and pebbled knuckles (Huntley's papules) (encircled).

with: a high level of reactive oxygen species, the advanced glycated endproducts, some immunobiochemicals, cytotoxic edema and microvasculopathy. Subsequently,

Figure 12: Split screen (gray-scale CUS) image of the normal MCP dorsal subcutis (left); the heterogeneous and thickened subcutis layer within overgrowth knuckle pad (encircled) (right).

Figure 13: High frequency ultrasound (HFUS) views of the dorsal MCP aspect. (A): Normal E-0.13 mm, (B): An irregular hyperkeratosis thickened E-0.28 mm, that impedes the deeper penetration.

Figure 14: Photograph of the upper dorsal skin exhibiting an indurated, erythematous aspect. The lower blue 'X' painted sign corresponds to the thickest skin.

Figure 15: Gray scale conventional ultrasound view of the 8.1 mm skin thickness (between crosses).

Figure 16: Photograph of Acanthosis nigricans area; its typical dark velvety appearance engages the dorsal neck flexure.

Figure 17: High frequency ultrasound (HFUS) of the normal and affected skin. (A): Normal cutis. E-20 mm; D-3.18 mm; Integument-3.34 mm. A dominant medium/low echogenicity in the UD and slightly hypoechogenic LwD, (B): Scleredema. Thicker layers with E-0.29 mm; D-5.21 mm; Integument-5.5 mm. Relatively higher UD echogenicity and intensively hypoechogenic LwD, (C): Acanthosis nigricans. An irregular thickened E-0.39 mm; ill-defined E-D junction; D-3.17 mm intensively hyperechogenic but slightly regressive in the LwD; Integument-3.56 mm.

hypoxia will ultimately increase collagen and glycosaminoglycan synthesis. The genetic should also be discussed [7].

Our patient exhibited within this context (HbA_{1c} higher than the accepted range: 7.9 vs 6.5–7%) the DD, LJM (their prevalence ranges between 8–58% compared with 4–26% in non-diabetics) and knuckle pads (50–75% of diabetics). The CUS differentiated the ST anatomical substructures and discerned the knuckle pads from synovitis, rheumatoid nodules and pachydermodactyly [8–11]. Using HFUS technique, the skin polychromatic structures express various echogenicities (extracellular matrix densities): white/yellowish - hyperechogenicity (HEP); yellow/red - medium echogenicity (MEP); dark green or black - hypoechogenicity (LEP). Therefore, a correlation between skin histology and HFUS imaging is quite possible [12].

Using the HFUS, the volar skin (no photoaged area) showed inflammatory edema and fibers abnormalities harbored within SLEB, besides the hyperkeratosis and thickened skin.

Regarding scleredema, the patient's data raised the suspicion and disjointed the differentials. Both operating US systems measures skin thickness: 8.1 mm CUS vs 5.5 mm HFUS. The discrepancy emerged from their different image construction abilities [12]. The HFUS described dermal thickening and dominant LEP that quantifies the inflammatory edema and thickened disorganized collagen fibers interspaced by acid mucopolysaccharides.

According to literature, AN substructures are: tiny papillomatosis, hyperkeratosis, hypertrophic epidermis and thickened dermis (collagen fibers abnormalities) being hyperinsulinemic promoted [13]. We found no particular CUS features but the HFUS scanned the dominant MEP and HEP components suggesting: proteic synthesis and probable a higher degree of promoting microfibrils. Nevertheless, the intrinsic ageing process cannot be discerned.

The treatment of the hands' pathology implies a tight glycemic control from the beginning. Other therapeutics are more or less successful: antioxidants, some local steroid injection, physical therapy. Regarding scleredema, certain attempts were adapted to cases: immunosuppressants, psoralen with ultraviolet -light (PUVA), tamoxifen, electron beam, physical therapies and so on [14, 15]. For our patient having no complications of scleredema (restrictive dyspnea, shoulder motion disturbances) or evidence of malignancy, a strict glycemic control with periodical exams were considered.

CONCLUSION

Such a great diversity of soft tissue pathology generally suggests, in diabetics, both the poorly metabolic control and microvasculopathy. The imagistics add valuable and complementary anatomo-histological related data; therefore, an earlier diagnosis and a holistic perception of the ST pathology could initiate a better understanding of its intrinsic processes and a preventive strategy related to microvasculopathy.

ABBREVIATIONS

ST: soft tissue. LD: Ledderhose's disease. DD: Dupuytren's disease. US: ultrasonography. MRI: magnetic resonance imaging. LJM: limited joint mobility. E: epidermis. D: dermis. SC: subcutis. AN: acanthosis nigricans. UD: upper dermis. LwD: lower dermis. LEP: low echogenic pixels. MEP: medium echogenic pixels. HEP: high echogenic pixels. MCP: metacarpophalangeal joints. IP: interphalangeal joints.

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Author Contributions

Rodica-Elena Perciun – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Ancuța Telcian – Acquisition, analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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