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PERNIOSIS: AN ENIGMATIC DERMATOLOGICAL ENTITY- STUDY OF ITS DEMOGRAPHIC PROFILE AND NAIL CAPILLAROSCOPIC CHANGES

Dermatology

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ABSTRACT

BACKGROUND: Perniosis is a localised inflammatory disorder affecting the unprotected skin regions of individuals exposed to damp, non-freezing cold. The exact pathogenesis has not been elucidated, however, it is thought to be a microvascular disease. **AIMS:** To study the demographic profile of patients presenting with perniosis and examine the nailfold capillaroscopic (NFC) changes. **MATERIAL AND METHODS:** This was an observational, cross-sectional study conducted at a Tertiary care hospital of North India over a period of three months (November 2020- January 2021). Patients with idiopathic perniosis were included in the study. After detailed history and clinical examination, proximal nailfold (PNF) video capillaroscopy was performed in the affected digits. **RESULTS:** A total of 47 patients with idiopathic perniosis were recruited in the study consisting of 35 females and 12 males with female to male ratio of 2.9:1. A majority of individuals belonged to age group 21-35 years (38.29%). Lesions were localized to both hands and feet in 23 patients (48.9%), only hands in 19(40.4%) and only toes in rest 5 (10.6%) patients. Ten (21.2%) patients had family history of similar complaints and history of smoking was present in 2 (4%) patients. The mean BMI recorded in 37 patients aged >20 years was 20.72±4.32kg/m^2. Finger nail videocapillaroscopy performed in 42 patients showed dilated capillaries in 42.8%, tortuosity in 40.4% and microhaemorrhages in 28.5% patients. **CONCLUSION:** In the present study, presence of nailfold capillaroscopic changes suggested that an organic damage to the microcirculation is present in perniosis. Low BMI is an important association for development of perniosis. **LIMITATIONS:** Long term follow up is required for patients with chilblains to assess permanency of nailfold capillaroscopic changes.

KEYWORDS

Chilblains, nailfold capillaroscopy, microhaemorrhages, body mass index (BMI).

INTRODUCTION

Perniosis, also called chilblains, is a localised inflammatory skin manifestation characterised by erythema and swelling of the extremities, persisting for more than 24 hours [1]. The disease is triggered by exposure to non-freezing damp cold usually during late winter or early spring when daily mean temperatures drop below the range of 12 °C to 15 °C [2].Clinically, it presents as oedematous, erythematous or violaceous macules, papules or plaques on the dorsal fingers or toes and less frequently on ears, nose, face or thighs. Lesions may be associated with pruritus, tenderness or burning. Vesicles, ulceration, permanent skin discolouration or scarring may occur in severe cases [3].Chilblains can either be idiopathic or occur in association with autoimmune connective tissue diseases such as systemic lupus erythematosus[4]. Recently, acral chilblain-like lesions have been reported in COVID-19 patients [5-8].

Defective vasodilatory reflex has been attributed to cause perniosis. In normal individuals, cold-induced vasoconstriction is followed by vasodilation, but in patients with perniosis, the initial vasoconstriction is prolonged, resulting in vasospasms [9].Since skin vessels are affected in pernio, potential changes in microcirculation may possibly be demonstrated by nailfold capillaroscopy (NFC). NFC is a non-invasive, reproducible, painless and inexpensive technique for studying proximal nailfold (PNF) capillaries. Changes in nailfold capillaroscopy in patients of perniosis have been demonstrated by Ozmen M et al [4]. However, still there is paucity of studies in India evaluating nailfold capillaroscopic changes in perniosis as this disorder mainly occurs following exposure to non-freezing, cold humid weather during winter or early spring that is mainly experienced in parts of North-western India. Therefore, present study was undertaken to assess nailfold capillaroscopic changes in pernio that can help to assess whether these patients are prone to develop permanent changes in nailfold capillaries later in life and also study demographic profile of patients presenting with this condition.

MATERIAL AND METHODS

This was an observational, cross-sectional study conducted at Government Medical College, Patiala, a tertiary care hospital of Northwest India over a period of three months (November 2020 to January 2021). The study was approved by Institute's Ethical Committee (vide memo number EC/NEW/.INST/2020/997/9413-14) . Patients of idiopathic perniosis presenting to outpatient Dermatology Department were recruited in the study. Patients with history suggestive of connective tissue disease or positive Anti nuclear antibody (ANA) were excluded from the study. COVID-19 RT-PCR was done in all patients to rule out infection. An informed consent was taken from all patients enrolled. Detailed relevant history of each patient was recorded in the proforma specially designed for this study. Data like age, gender, and BMI [weight (kg)/height $(m)^2$] was recorded. Proximal nailfold capillaroscopy was performed in patients with active disease in the involved digits by video USB dermatoscope (Dino-lite AM2111, 10X-220X). Ultrasound gel was used to increase skin permeability. Results of nailfold capillaroscopy were recorded as follow [10] [Table 1]:

Table 1: Definitions of various abnormalities noted on NFC

Abnormality	Definition
Tortuous capillary	Capillary limb curled but not crossing over
Dilated capillary	Width more than two times surrounding normal capillaries
Focal microhaemorrhages	Singularly placed micro petechiae
Diffuse microhaemorrhages	Multiple micro petechiae present in groups
Capillary dropout	Absence of individual capillary loop
Avascular area	Absence of two or more adjacent capillaries from the distal most row

STATISTICAL ANALYSIS: Data was entered into Microsoft Excel and presented as tables.

RESULTS

Out of 47 patients recruited in the study, 35 were females and 12 were males with female to male ratio of 2.9:1. The most common age group affected was 21-35 years (38.29%) with mean age 33±19.3 years (range 13 to 78 years) [Table 2].

Table 2: Age distribution of patients

Parameters	Number (%)
Age group (in years)	
<20	10 (21.2%)
21-35	18 (38.29%)
35-50	14 (29.7%)
>50	5 (10.6%)

Majority of patients (31.9%) had age of onset of chilblains between 21-30 years. Past history of chilblains was present in 32 (68%) patients whereas 15 (31.9%) patients had first episode. Ten (21.2%) patients had family history of similar complaints. History of smoking was present in 2 (4%) patients.

The most common sites involved were both hands and feet in 23 patients (48.9%), followed by only hands in 19(40.4%) and only toes in rest 5 (10.6%) patients. (Figure 1,2)

Fig 1: Erythema, oedema & ulceration of fingers

Fig 2: Erythema & oedema on toes

Two (4.2%) patients had involvement of ears and nose was involved in 1 (2.1%) patient. Amongst 37 patients aged >20 years, the BMI calculated ranged 17-28.5 kg/m^2 with mean BMI 20.72 $\pm$ 4.2 kg/m^2. Erythema and oedema was present in all the patients which was associated with ulceration in 5 (10%) and pigmentation also in 5 (10%) .The examination findings & BMI are depicted in Table 3.

Table 3: BMI & Examination findings of patients

BMI (kg/m^2) (in 37 patients aged >20 years)	
<18.5	10 (27%)
18.5-24.9	21 (56.7%)
25-29.9	6 (16.2%)
30.0 or above	0
Dermatological examination	
Erythema and oedema	37
Erythema, oedema and pigmentation	5
Erythema, oedema with vesicles	0
Erythema, oedema with ulcers	5

Proximal nailfold capillaroscopy of involved fingernails was performed in 42 patients with involvement of hands or both hands and feet. The most common finding was capillary dilation present in 18 (42.8%) patients, followed by tortuosity in 17 (40.4%) and microhaemorrhages in 12 (28.5%) patients [Figure3,4].

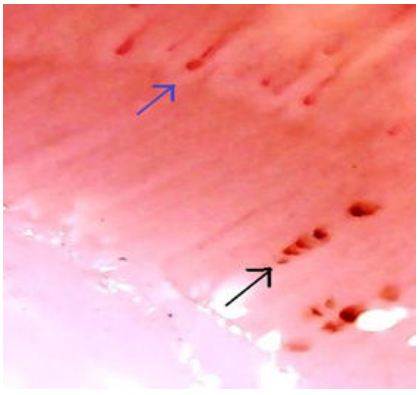


Fig 3:NFC Showing microhaemorhages

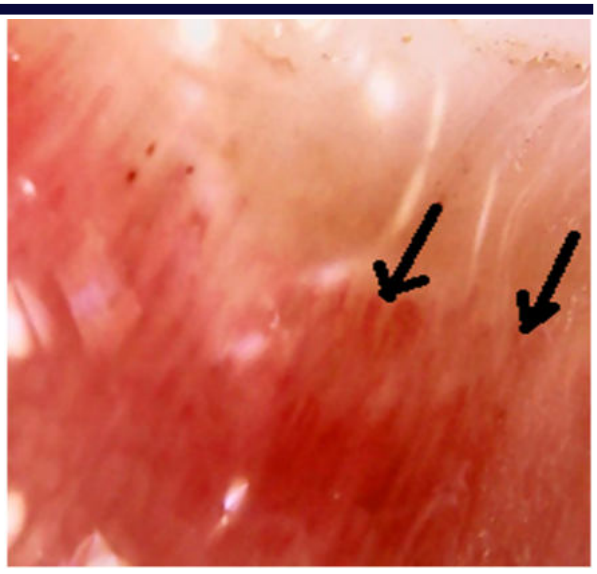


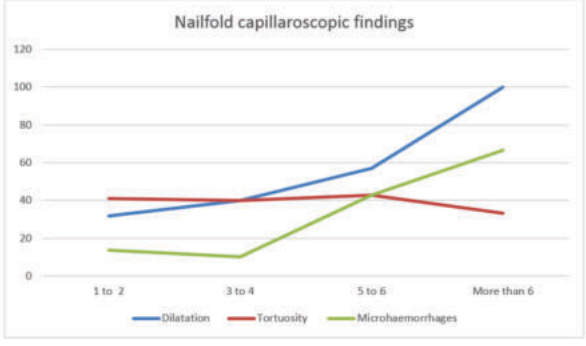
Fig 4 : NFC showing dilated & tortuous vessels

No avascular areas or capillary dropouts were seen. Normal study was present in 15 (35.7%) patients [Table 4].

Table 4: Nailfold capillaroscopic findings:

No. of episodes	Total number of patients	Dilatation	Tortuosity	Microhaemorrhages
1-2	22	7 (31.8%)	9 (40.9%)	3 (13.6%)
3-4	10	4 (40%)	4 (40%)	1 (10%)
5-6	7	4 (57.1%)	3 (42.8%)	3 (42.8%)
>6	3	3 (100%)	1 (33.3%)	2 (66.6%)
Total	42	18 (42.8%)	17 (40.4%)	12 (28.5%)

Table 5: Graphical Representation of Nailfold capillaroscopic findings



DISCUSSION

Perniosis or chilblains is disorder that mainly affects the extremities upon exposure to non freezing, damp cold. Both patient-related and environmental factors play a role in pathophysiology of perniosis. There is no clear explanation why some individuals are more vulnerable to cold than others. However, the role of hyper viscosity, abnormal vasoconstriction and vasculitis has been suggested [4].

In the present study, the mean age of the patients presenting with chilblains was 33$\pm$19.3 years. (range 13 to 78 years) and 37 patients were aged >19 years. However, in a study conducted by Ozmen M et al the mean age was 25 $\pm$ 9 years (range 16–51) and ranged from 12 to 46 years with a mean of 29.73$\pm$7.8 years in the study conducted by Singh GK et al [4,3]. The difference may be due to different study designs or due to regional variations.

Out of 47 patients recruited in the present study, 35 were females and 12 were males with female to male ratio of 2.9:1. The results of our study corroborated with findings of previous studies where chilblains have been described more commonly in females [11,12].

The most common sites involved were both hands and feet in 48.9%

patients followed by hands in 40.4% patients and feet in 10.6% patients. Two patients had involvement of ears and one had involvement of nose. However, the study conducted by Singh GK et al reported feet to be commonest site involved in 41.6% patients followed by both hands and feet in 29.6%, while hands alone were involved in 21.2% patients [3]. The difference may be due to the fact that majority of subjects in their study were soldiers serving at high altitude of Ladakh.

Though smoking has been attributed as a risk factor for development of cold related injuries, only two (4.2%) patient had history of smoking which could be due to the fact that majority of our patients were females and could also be due to regional and cultural differences.

Amongst 37 patients aged >20 years, the mean BMI recorded was 20.72 ± 4.2 kg/m^2. Similar findings were observed by Singh et al in their study where majority of patients (63.8%) had had BMI ranging between 20 and 22 kg/m2 with arithmetic mean of 20.03 kg/m^2 suggesting that patients with lower BMI are more prone to develop perniosis [3]. Similar findings were observed in the studies conducted by Prakash S et al and Singh GK et al where 33.33% patients had BMI ranging from 17 to 19[9,14].

In the nailfold, unlike in other regions, capillaries are located in parallel to the surface of the skin. This allows organic and functional changes in the microcirculation to be assessed by video capillaroscopy. As pernio is a disease that affects mainly the skin vessels, it may be possible to demonstrate the potential changes in the microcirculation by capillaroscopy. Proximal nail fold capillaroscopy was performed in affected fingernails of 42 patients who had involvement of either hands or both hands and feet. The most common finding was capillary dilation present in 18 (42.8%) patients followed by capillary tortuosity in 17 (40.4%) patients and microhaemorrhages in 12 (28.5%) patients. The findings of capillary dilation and microhaemorrhages increased as the number of episodes increased in patients[Table 5]. Patients presenting with fewer episodes of chilblains in the past had less prominent changes on proximal nailfold capillaroscopy. Study conducted by Ozmen et al also found tortuosity and increased capillary diameter in patients with perniosis on nailfold video capillaroscopy and they suggested that these results may indicate organic damage to the microcirculation[4].

LIMITATIONS: Long term follow up is required for patients with chilblains to assess permanency of nailfold capillaroscopic changes.

CONCLUSION

In present study, presence of nailfold capillaroscopic changes in patients with pernisois suggested that an organic damage to the microcirculation is present. We found that as that number of episodes of perniosis increased, more nailfold capillaroscopic changes in the form of dilatation and microhaemorrhages were present. More prospective studies are required to validate these findings. Also**,** it was observed that patients presenting with perniosis had low BMI. Long term studies are needed to study nailfold capillaroscopic changes in patients with perniosis in summer months as well to assess whether the changes are persistent, functional or organic and also to evaluate whether these patients develop connective tissue disorders later in life.

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