PAPULONECROTIC TUBERCULID (CASE REPORT)

MOHAMMED MOHY EL- DIN SELIM, MD And BAHRAM AZADEH, MD, FRC Path.

Abstract

We present in this article a 48 years old female who suffered from papulonecrotic tuberculid for 4 years. The diagnosis is based on the clinical picture, the highly positive tuberculin test, the histopathologic findings and the history of contact with an open pulmonary tuberculosis.

Introduction

Tuberculids are chronic, recurrent skin disorders which affect persons who have had personal or family history of active tuberculosis with a strongly positive Mantoux test (1)

Tuberculids result from hematogenous spread of mycobacteria in a patient with a moderate to a high degree of immunity. The mycobacterium tuberculosis cannot be isolated from the lesion but the mycobacterial DNA could be demonstrated by Polymerase chain reaction (PCR), [2,3,4].

The relation between the degree of the patient immunity and virulence of the mycobacteria determines the development and features of the tuberculids [1,5]. The tuberculids include lichen scrofulosorum, populonecrotic tuberculid (PNT), erythema induratum (nodular vasculitis), erythema nodosum and recently a nodular granulomatous phlebitis of the skin was decribed as a form of tuberculid

Case Report

A female born 1949 from Iraq. She has been suffering from sore red hands and feet. The duration of her complaint was 4 years for the hands and 2.5 years for the feet. Her condition was aggravated in winter or by exposure to cold and gets better in summer. She gave history of being in contact with a case of active pulmonary tuberculosis. On examination she was a normotensive female with good general condition. Both hands were reddish blue with tiny papules (3-4 mm) on the dorsal surface of hands and flexor aspect of both wrists. [Fig. 1,2,3,4,]. Some papules had central necrosis. Both feet showed reddish blue papules dorsally and in tendoacheles region [Fig. 5,6,7,8]. Most of the lesions on clearing leave atrophic pigmented scars.

The condition was provisionally diagnosed as perniosis, erythromelalgia and PNT.

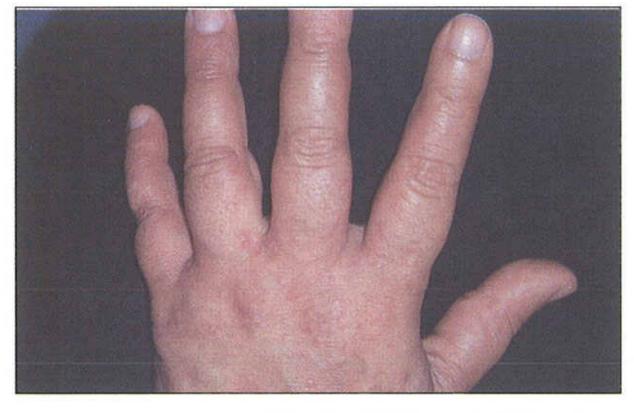
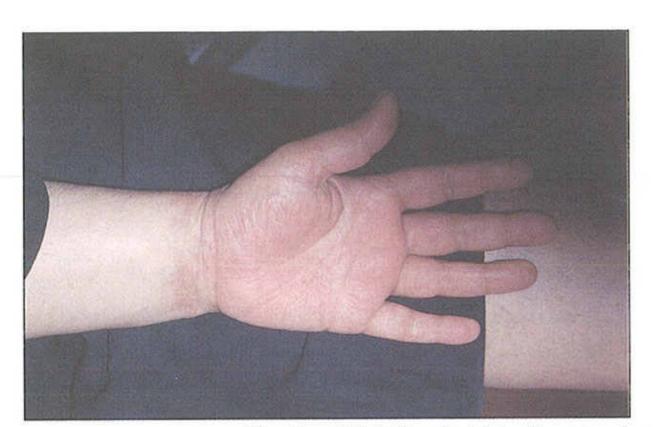




Fig. 1 and 2: left and right hands showing bluish red colour and papular lesions



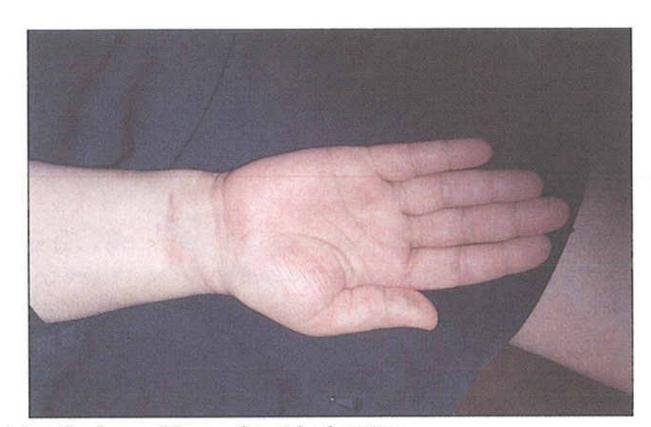


Fig. 3 and 4: left and right palms showing bluish red colour with papules at both wrists.





Fig. 5 and 6: left and right feet showing papular lesions.

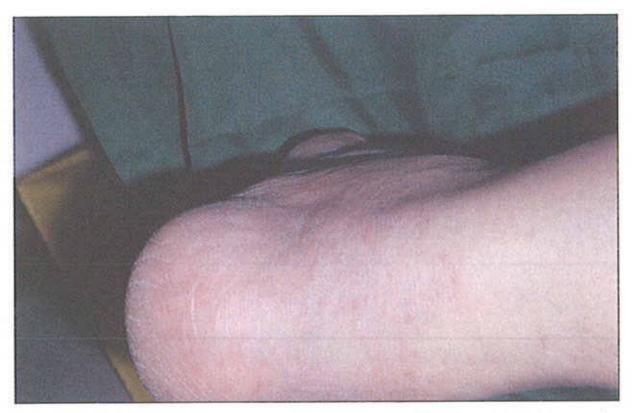




Fig. 7 and 8: left and right tendo-acheles region showing papular lesions.



Fig. 9: highly positive tuberculin test with central bulla and necrosis.

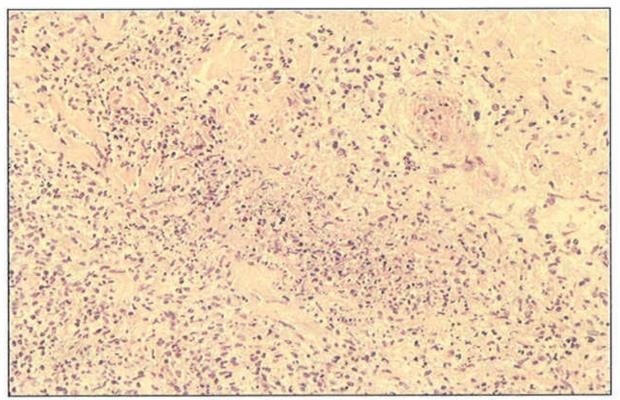


Fig. 11: Acute vasculitis with fibrinoid necrosis, thrombosis on the lumina and perivascular inflammatory cellular inflitrates.

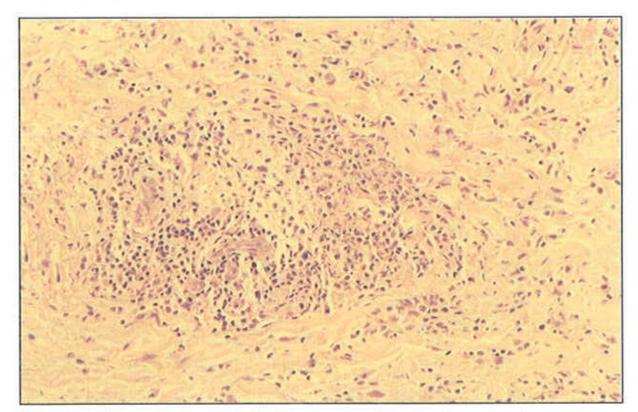


Fig. 13: Chronic lymphocytic vasculites in the periphery of the lesion showing a dense mantle of mononuclear inflammatory cells around a vessel HE x 25.

The investigations done showed the following results -

1-Microcytic anemia with WBC count of 4.3, high monocytes (15.5) low neutrophils (32.6), high eosinophils (6.6) and raised E.S.R. (51).

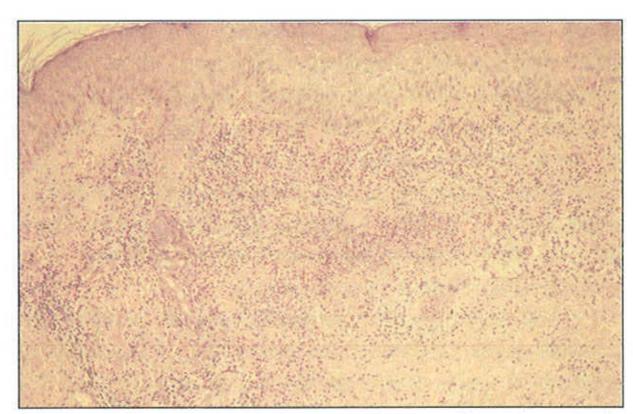


Fig. 10: inflammatory and necrotic changes along the axis of a hair follicle and blood vessels HE x 12.

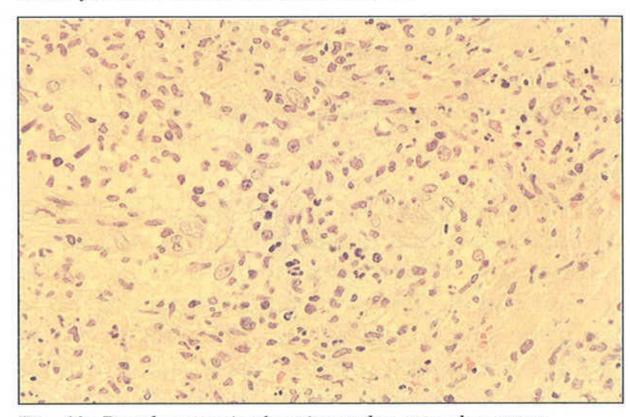


Fig. 12: Poorly organized perivascular granulomatous aggregates consisting of monocytohistiocytic cells HE x 50.

2-Blood biochemistry showed normal blood sugar, kidney function, liver function, blood proteins and high cholesterol (6.17 m.mol/l).

3-Serology and immunology showed normal ANA and anti DNA antibodies, normal C3 and C4, negative rheumatoid factor, cryoglobulins, treponema IgG and negative serology for hepatitis A,B,C. and normal G6PD.

4-Tuberculin test was highly positive (more than 30 mm with central bulla and necrosis [Fig.9]

5-X-ray chest showed prominant hilae with increased bronchovascular marking and reticular shadowing seen on both sides. Both CP angles were clear and cardiac size was within normal limits.

6-Biopsy showed a relatively large, elliptical zone of dermal necrosis with vasculitis, inflammatory cellular infiltrates and ulceration of overlying epidermis. Inflammatory and necrotic changes were centered along philosebaccous follicles and blood vessels [Fig.10]. Vessels adjacent to or in the cen-

ter of the necrotic zone showed fibrinoid necrosis of the walls and thrombotic occlusion of lumina with perivascular infiltrates of histiocytic macrophages, lymphocytes and nuclear debris [Fig.11]. Polymorphs and plasma cells were inconspicious. Histiocytic granulomatous aggregates were present in some foci [Fig.12]. Vasculitic changes in the periphery of the lesion appeared more chronic with a perivascular dense mantle of

lymphocytes and monohistiocytic cells without any noticable filirinoid necrosis [Fig.13]. Scattered epitheloid cells were noticed but not in sufficient numbers to constitute a definite palisade. Well defined epitheloid cell tubercles were not present in multiple level examined. There was no caseation and Ziehl Neelsen staining did not reveal any acid fast bacilli. The histopathologic examination was consistant with the diagnosis of PNT.

Discussion

PNT is characterized by papules eruption affecting extrimities and occuring in more or less symmetric crops. The papules are dusky red, symptomless and usually not itchy but were reported in one case to be severely itchy (7). The sites involved in PNT are the dorsum of hands and feet, the elbows, the knees, the palms, the soles, the buttocks and perineum, the glans peris was reported to be affected (8,9), and perniotic areas may be favoured (10). PNT affects patients who have strongly positive tuberculin test which may even show large bulloe (7).

The diagnosis of PNT in the present case is based on the clinical presentation, the history of contact with a family case of active pulmonary tuberculosis and the highly positive tuberculin test. It was reported that mycobacterial tuberculosis DNA has been detected by PCR in 77.8% of PNT specimens (11). Wilson-Jones and Winkelmonn believe that PNT presents a characteristic clinical and histopathological pattern and that a special granulomatous type of small-vessel vasculites explains the unique pathological features of the disease. Immunohisto-chemical labeling of biopsy specimens (13), revealed a preponderance of T-lymphocytes, monocytes-macrophages and Largerhans cells indicating type IV hypersensitivity reaction. B lymphocytes were sparse. Conditions that may be confused with PNT on clinical or histopathological ground include perforating callagenosis, perforating granuloma annulare,

necrotizing or septic vasculitis, Churg-Strauss granuloma, populonecrotic syphilid, pityriasis lichenoides chronica, lymphomatoid populosis, popular articaria, miliary tuberculosis, chordrodermatitis nodularis, suppurative falliculites and infectuous causes of palisading granulomas (12,13.14).

In the 12 cases described by Wilson-Jones and Winkelmann (12), the age of the patients varied between 7 years and 30 years. The famales predominated (9 of 12) with a female to male ration 3:1 and tuberculin test was positive in 100% of cases. The number of locations affected in each patient varied from one to six with a mean of 3. The principal sites affected were the hands (8 of 12); the legs (5 of 12); the feet and buttocks (4 of 12); the knees, elbows, thighs and arms (3 of 12); fingers, trunk and ears (1 of 12). The duration of the lesion varied from 5 weeks to 15 years.

Our patient, a female 48 years old had the PNT on hands and feet and she winter and improves in summer. We provisionally thought of perniosis and the investigations done were negative to cryoglobulins, hepatitis B, C & A, rheumetoid factor, ANA, and anti DNA antibodies. She had raised ESR and highly positive Mantoux test. Wilson-Jones and Winkelmann (12) thought that one of their patients had chilblains and 6 of the 7 patients they tested for ESR had raised ESR readings which became normal after responding to antituberculous therapy in 3 to 12 weeks and they suggested that ESR could be useful to monitor the effect of treatment. The influence of cold and the predilection of PNT to perniotic areas was stressed (10).

PNT was reported to evolve in lupus vulgaris (15), or may be associated with erythema induration (12,15,16). Extracutaneous tuberculosis was found in 30-40% of cases with high incidence of nodal affection (15,17). Therefore in patients with PNT it is important to search for any associated tuberculosis in lymph nodes, lungs and bones (12).

Some consider PNT the most common tuberculid (18). Tuberculids in general are treated with antituberculous drugs (1), treatment is indicated even in absence of M. tuberculosis in histologic specimens or culture of skin lesion (19). Triple agent chemotherapy (rifampicin 900mg/d, isoniazed 400 mg/d and ethambutol 1200 mg/d), could lead to remission in two months with maintenence of treatment for 12 months (20). Long term triple regimen of treatment is recommended for PNT (21).

REFERENCE

- 1-Rademaker-M; Lowe DG; Munro-DD:Erythema Induratum (Bazin's disease).J. Am. Acad, Dermatol 1989; 21:740-5
- 2-Degitz-K; Steidl-M; Plewig-G. et al: Aetiology of tuberculids. Lancet 1993; 341:239-40.
- 3-Kox-L.F.F.; Rhienthong-D; Medo Miranda-A et al: a more reliable PCR for detection of Mycobacterium tuberculosis in clinical samples. J. of Clin. Microbiol. 1994; 32: 672-78.
- 4-Degitz-K: Detection of mycobacterial DNA in the skin Etiologic insights and diagnostic perspectives. Arch-Dermatol 1996; 132:71-75.
- 5-Wolff-K, Tappeiner-G: Mycobacterial diseases: Tuberculosis and atypical infection, in Dermatology in General medicine. Third edition by Fitzpatrick TB, Eisen-AZ; Wolff-K; Freedberg-IM and Austen-KF, 1987 Mc Graw Hill. Book company P 2152-80.
- 6-Hara-K; Tsuzuki-T; Takagi N et al: Nodular granulomatous phlebitis of the skin: a fourth type of tuberculid. Histopathology 1997; 30: 129-34.
- 7-Iden-DL; Rogers-RS and Schroeter-AL: Populonecrotic tuberculid secondaryto mycobacterium bovis.Arch-Dermatol. 1978; 114: 564-66
- 8-Iscraelewicz-S; Dharan-M; Rosenman-D. et al: Populonecrotic tuberculid of the glans penis. J. Am. Acad Dermatol. 1985; 12: 1104-106
- 9-Nakamura-S, Aoki-M, Nakayama-K, el al: Penis tuberculid (Papulonecrotic tuberculid of the glans penis): treated with a combination of rifampicin and an extract from tubercle bacilli (T.B. vaccine). J. Dermatol. 1989; 16: 150-3
- 10-Savin-J.A.: Mycobacterial infections, in Rook, Wilkinson, Ebling text book of dermatology-fifth edition edited by R.H. Champion, J.L. Burton and F.J.G. Ebling Blackwell scientific

- publications 1992, Page 1050-56.
- 11-Baselga-E; Margall-N; Barnadas-MA et al: Mycobacterium tuberculosis.
- Arch Dermatol. 1996, 132: 92-94.
- 12-Wilson-Jones-E and Winkelmann-RK: Papulonecrotic tuberculid: A neglected disease in Western countries.J. Am. Acad. Dermatol. 1986; 14: 815-26.
- 13-Jordaan-HF; Von-Niekerk-DJ, Louw-M: Papulonecrotic tuberculid. A clinical, histopathological and immunohistochemical study of 15 patients. Am-J. Dermatopathol. 1994; 16: 474-85.
- 14-Su-WP; Kuechle-MK, Peters MS, et al: Palesading granuloma caused by infectuous diseases. Am.J. Dermatopathol 1992; 14:211-5.
- 15-Morrison JGL, Fourie ED: The papulonecrotic tuberculid from Arthus reaction to lupus vulgaris. Br. J. Dermatol. 1974; 91: 263-70.
- 16-Wong Ko, Lee KP, Chin SF: Tuberculosis of the skin in Honk Kong: A review of 160 cases. Br. J. Dermatol. 1968; 80:424:39.
- 17-Schwarz-K; Ott-F; Eichenberger-H: Papulonecrotic tuberculid. Lymph node tuberculosis. Dermatologica 1967; 134: 323-4.
- 18-Visser-AJ; Heyl-T: Skin tuberculosis as seen at Ga-Rankuwa Hospital. Clin Exp. Dermatol. 1993; 18: 507-15
- 19-Hassoun-PM; Shepherd-KE; Flotte-TJ, et al: Erythema induratum and active pulmonary tuberculosis Am.J.Med, 1988; 84:784-5.
- 20-Ollert-MW; Thomas-P; Korting-H.C., et al: Erythema induratum of Bazin. Evidence of T-lymphocyle hyperresponsiveness to purified protein derivative of tuberculin- Report of two cases and treatment Arch.Dermatol 1993; 129:469-73.
- 21-Kullavanijaya-P; Sirimachan-S; Suwantaroj-S: Papulonecrotic tuberculid- Necessity of long term triple regimens. Int. J. Dermatol. 1991; 30: 487-90.