

Telangiectasia macularis eruptiva perstans: an old terminology, still frequently used

Kelly Wilmas^{1,2} BS, Iris Wohlmuth-Wieser^{2,3} MD, Madeleine Duvic² MD

Affiliations: ¹John P. and Kathrine G. McGovern Medical School at the University of Texas Health Science Center, Houston, Texas ²Department of Dermatology, The University of Texas, MD Anderson Cancer Center, Houston, Texas ³Department of Dermatology, Paracelsus Medical University Salzburg, Austria

Corresponding Author: Kelly Wilmas, BS, John P. and Kathrine G. McGovern Medical School, University of Texas Health Science Center, 6431 Fannin Street, Houston, Texas 77030, Tel: (682)225-4986, Email: Kelly.M.Wilmas@uth.tmc.edu

Abstract

The term telangiectasia macularis eruptiva perstans (TMEP) was originally used to describe a rare form of cutaneous mastocytosis (CM) that was limited to the skin with lesions consisting of irregular, telangiectatic macules ranging in color from red to brown. Recent guidelines, however, recommended that the sole presence of telangiectasias should not form the basis of a distinct variant of CM. We conducted a review of the literature from 1930 to 2017 and found 76 cases that were reported as TMEP. Owing to a general misconception about diagnosis of CM and SM, there is a need for further discussion and awareness of the newly proposed World Health Organization (WHO) guidelines.

Keywords: TMEP: telangiectasia macularis eruptiva perstans; CM: cutaneous mastocytosis; SM: systemic mastocytosis; WHO: World Health Organization; MPN: myeloproliferative neoplasms; MC: mast cells; MPCM: maculopapular cutaneous mastocytosis; UP: urticaria pigmentosa; DCM: diffuse cutaneous mastocytosis; ISM: indolent SM; SSM: smoldering systemic mastocytosis; AHN: systemic mastocytosis with associated hematologic neoplasm; ASM: aggressive systemic mastocytosis; MCL: mast cell leukemia MCS: mast cell sarcoma; MIS: mastocytosis in the skin

Introduction

Mastocytosis represents a wide variety of disorders with an abnormal proliferation of mast cells that release histamine and inflammatory mediators [1]. These mediators are associated with pruritus, flushing,

nausea, vomiting, abdominal pain, diarrhea, vascular instability, and headache [1]. The clinical presentation of mastocytosis is heterogeneous, ranging from disease limited to the skin that spontaneously resolves, often seen in pediatric cases, to more ominous variants with extracutaneous involvement associated with multiorgan failure and shortened lifespan, more commonly found in adult patients [2]. Cutaneous lesions are usually red or brown and monomorphic in adults with similar color, size, and shape, whereas the lesions in children are frequently polymorphic [3]. Darier sign, characterized by reddening and whealing of lesions upon mechanical stroking or rubbing, is often negative in adults, but commonly positive in children [3].

Historically, mastocytosis was subdivided into cutaneous mastocytosis (CM) and systemic mastocytosis (SM). Telangiectasia macularis eruptiva perstans (TMEP) represented a rare form of CM characterized by small, irregular, reddish, telangiectatic, and maculopapular skin lesions with a background color of light to dark brown, in addition to an abnormal infiltration of mast cells in the superficial dermis [4, 5]. Histologically, diagnosis was made by detection of increased amounts of mast cells around dilated venous plexus capillaries and venules of the upper third of the dermis [5]. No guidelines were established as to whether systemic testing should be performed, given the suspected diagnosis of TMEP. Furthermore, there was a lack of evidence demonstrating that TMEP was a separate subtype of CM, as it could have simply represented a highly vascularized form of maculopapular CM with dilated vessels [6]. Owing to an increasing

number of variants in clinical presentation and prognosis, the criteria for diagnosis of CM and SM evolved [1]. In 2016, an international task force of experts from various medical specialties proposed a revised definition of CM that would remove TMEP as a diagnosis because of overlap with urticaria pigmentosa (UP), [3]. Consequently, the term TMEP should no longer be used as a diagnosis, even if the predominant clinical feature is telangiectasia. The World Health Organization (WHO) refined its classification and diagnostic criteria for mastocytosis in 2016, and an understanding of these updates are essential for correct disease management.

Mastocytosis: Diagnosis and Classification

The WHO now classifies mastocytosis as one of eight subcategories of myeloproliferative neoplasms (MPN), [2]. Infiltration of neoplastic, clonal proliferation of mast cells (MC) that are morphologically and immunophenotypically abnormal into one or more organs causes mastocytosis [2]. Mastocytosis can be characterized by cutaneous mastocytosis (CM), systemic mastocytosis (SM), or a localized mast cell tumor (**Table 1**), [6]. CM is defined by an increased number of mast cells in the skin without involvement of extracutaneous tissues, whereas SM is defined by the proliferation and accumulation of mast cells in the bone marrow (BM) and/or other extracutaneous organs, with or without skin involvement [7].

CM is divided into maculopapular CM (MPCM), which is equivalent to urticarial pigmentosa (UP), diffuse cutaneous mastocytosis (DCM), and solitary mastocytosis of the skin [8]. SM is divided into indolent systemic mastocytosis (ISM), smoldering systemic mastocytosis (SSM), systemic mastocytosis with associated hematologic neoplasm (AHN), and mast cell leukemia (MCL), [8]. Regardless of the subtype of SM, the bone marrow (BM) is involved in nearly all cases [9]. Mast cell sarcoma (MCS) is a separate subtype of mastocytosis [8].

The diagnosis and classification of mastocytosis is determined by morphologic, molecular, and immunophenotypic WHO criteria [2]. Mastocytosis in the skin (MIS) describes patients with skin lesions typical of mastocytosis who have not had full work-up for systemic involvement, including BM biopsy. In patients with clinical signs and symptoms of MIS, first obtain a skin biopsy with histology [7]. If MIS is confirmed, WHO criteria for SM include screening for serum tryptase level, BM histology (tryptase and/or KIT (CD117) immunostaining), MC immunophenotyping (CD25 and CD2 expression), KITD816V mutation screening in the BM, blood, or extracutaneous specimen, and FIP1L1-PDGFR screening in the BM and blood if eosinophilia is present [7]. These patients have a different prognosis than those with SM, so a BM biopsy is necessary and suggested for all adult patients with MIS. If at least one major and two minor or three minor criteria

Table 1. WHO mastocytosis classification 2016.

Cutaneous mastocytosis (CM)
Maculopapular CM (MPCM)/urticaria pigmentosa (UP)
Diffuse CM (DCM)
Solitary mastocytoma of skin
Systemic mastocytosis
Indolent SM (ISM)
Smoldering SM (SSM)
SM with associated hematologic neoplasm (AHN)
Aggressive SM (ASM)
Mast cell leukemia (MCL)
Mast cell sarcoma (MCS)

Table 2. SM criteria defined by the WHO and updated in 2016.

Major SM criterion
Multifocal dense infiltrates of MCs (≥ 15 MCs in aggregates) in BM biopsies and/or biopsies of extracutaneous organs
Minor SM criterion
>25% of MCs are atypical on BM aspirate smears or are spindle-shaped in MC infiltrates detected on sections of visceral organs
Activating KIT point mutation at codon 816 in BM, blood, or other extracutaneous organ
MCs in BM, blood, or other extracutaneous organ express CD2 and/or CD25 in addition to normal mast cell markers
Baseline serum total tryptase level >20 ng/mL

are met, SM is confirmed (**Table 2**), [2, 7]. The major criterion includes multifocal dense infiltrates of MCs in aggregates on BM or other extracutaneous organ biopsy. Tryptase is a vasoactive immunoregulatory mediator found in mast cell granules, and elevated serum total tryptase levels >20 ng/mL is a minor criterion for the diagnosis of SM owing to its elevation in most patients with SM [1, 2, 4]. A positive KIT mutation represents a minor diagnostic criterion of mastocytosis, since the catalytic domain of c-kit can lead to unregulated growth and differentiation of mast cells and has been associated with a large number of adults with mastocytosis [1]. FIP1L1-PDGFR α screening in the presence of eosinophilia distinguish chronic eosinophilic leukemia from SM [7]. Neoplastic cells are characterized by abnormal morphology and immunoreactivity to CD2 and CD25 [7]. After systemic investigation, >95% of adults with mastocytosis are diagnosed with SM [2]. If SM is confirmed, screening for B and C findings should be performed to assess for SSM and ASM (**Table 3**), [7]. SSM requires two or more B-findings and no C-findings whereas ASM requires one C-finding and documentation of local mast cell infiltration [2, 7]. Criteria for associated hematologic neoplasms should also be checked [2, 7]. Nevertheless, some patients have only CM; systemic testing was performed and negative [2]. If SM is not found, follow up should be done with a blood count, serum tryptase level, and observation of clinical symptoms [7]. Although adult-onset mastocytosis tends to be a chronic disease, pediatric mastocytosis in the skin has been reported to spontaneously remit in 70-80% by their teenage years [6]. Therefore, systemic testing is not necessary

in children and the diagnosis of CM is accepted [6].

Treatment

A review of all treatments of mastocytosis is outside the scope of this article. There is no gold-standard treatment for patients with mastocytosis, but the individual patient's symptoms, organ involvement, and clinical presentation should guide treatment [6]. The mediators released from mast cells found in all subtypes of mastocytosis require a broad approach to the treatment of systemic symptoms [1]. Treatment currently includes H1 antihistamines, H2 antihistamines, proton pump inhibitors, anti-leukotriene agents, and injectable epinephrine to improve symptoms [1]. Topical corticosteroids may be effective acutely for CM, and psoralen-ultraviolet A therapy can be used in severe cases [1]. Patients should avoid known triggers that stimulate mast cell degradation, such as exposure to certain foods, sunlight, heat, cold, alcohol, and drugs [4]. In patients with SM, corticosteroids, ketotifen, cromolyn sodium, or leukotriene antagonists may be used [8]. Immunotherapy or IgE-depleting agents can be used for patients with SM and severe symptoms from allergy to bee or wasp venom [8]. Drugs targeting mutant KIT forms are in development and may be a novel therapeutic treatment for SM in addition to allogeneic stem cell transplant for refractory cases [8].

Discussion

Reports on the epidemiological aspects of mastocytosis are few. The prevalence of mastocytosis in the pediatric population is not known. The disease is likely to be underdiagnosed or misdiagnosed

Table 3. WHO "B- and C-" findings in SM.

B findings
BM biopsy with >30% infiltration by MCs and total serum tryptase >200 ng/mL
Signs of dysplasia or myeloproliferation in non-MC lineage, but insufficient criteria for definitive diagnosis of a hematopoietic neoplasm, with normal or slightly abnormal blood counts
Hepatomegaly without impairment of liver function, and/or palpable splenomegaly without hypersplenism, and/or lymphadenopathy on palpation or imaging
C findings
Bone marrow dysfunction manifested by one or more cytopenia, but no obvious non-MC hematopoietic malignancy
Palpable hepatomegaly with impairment of liver function, ascites, and/or portal hypertension
Skeletal involvement with large osteolytic lesions and/or pathological fractures
Palpable splenomegaly with hypersplenism
Malabsorption with weight loss due to gastrointestinal MC infiltrates

owing to the complicated symptoms, varying systemic involvement, and a lack of awareness among medical professionals. The prevalence of disease will likely increase with greater awareness of updated diagnostic recommendations.

We conducted a search of the literature on PubMed from 1930 to 2017 and identified 61 relevant articles (with 76 reported single cases) in 8 different languages (English, Spanish, German, French, Dutch, Italian, Hungarian, and Portuguese) describing patients with a diagnosis of TMEP [10-70]. The median age at diagnosis was 46, ranging from 0.5 to 86 years, and a female to male ratio of 1.1:1. Systemic testing was only done in 26 out of 76 subjects (34.2%) and was positive in 6 (23.1%). Of the 26 cases with systemic testing done, none were children. These findings demonstrate the need for increased awareness and usage of WHO guidelines so that patient survival and quality of life can be greatly improved.

Conclusion

Mastocytosis is a rare, but complex disease with many subtypes and clinical variants. Owing to the difference in prognosis, ranging from no symptoms with a normal life expectancy to severe symptoms and death within weeks, a thorough understanding of the disease and correct diagnostic algorithm is needed. Additionally, an increasing number of patients seek information about their disease on the internet, and they may encounter outdated,

incorrect, or irrelevant information that may raise unnecessary concerns. Increased acceptance and awareness of WHO diagnostic criteria for patients with signs and symptoms of mastocytosis can lead to earlier diagnosis and treatment, which produces better health outcomes.

References

1. Pettigrew HD, Teuber SS, Kong JS, Gershwin ME. Contemporary challenges in mastocytosis. *Clin Rev Allergy Immunol* 2010; 38(2-3): 125-34. [PMID: 19639428].
2. Pardanani A. Systemic mastocytosis in adults: 2017 update on diagnosis, risk stratification, and management. *Am J Hematol* 2016; 91:1147-1159. [PMID: 27762455].
3. Hartmann K, Escribano L, Grattan C, et al. Cutaneous manifestations in patients with mastocytosis: Consensus report of the European Competence Network on Mastocytosis; the American Academy of Allergy, Asthma & Immunology; and the European Academy of Allergology and Clinical Immunology. *J Allergy Clin Immunol* 2016; 137(1): 35-45. [PMID: 26476479].
4. Costa DLM, Moura HH, Rodrigues R, et al. Telangiectasia Macularis Eruptiva Perstans: A Rare Form of Adult Mastocytosis. *J Clin Aesthet Dermatol* 2011; 4(10): 52-54. [PMID: 22010057].
5. Severino M, Chandesris M, Barete S, et al. Telangiectasia macularis eruptiva perstans (TMEP): A form of cutaneous mastocytosis with potential systemic involvement. *J Am Acad Dermatol* 2016; 4(5): 885-91.e1 [PMID: 26899198].
6. Broesby-Olsen S, Dybedal I, Gülen T, Kristensen T.K., Møller M.B., Ackermann L. i inni: Multidisciplinary Management of Mastocytosis: Nordic Expert Group Consensus. *Acta Derm Venereol* 2016, 96, 602-612. [PMID: 26694951].
7. Lange M, Ługowska-Umer H, Niedozytko M, et al. Diagnosis of Mastocytosis in Children and Adults in Daily Clinical Practice. *Acta Derm Venereol* 2016; 96:292-297. [PMID: 26270728].
8. Valent P, Akin C, Hartmann K, et al. Advances in the Classification and Treatment of Mastocytosis: Current Status and Outlook toward the Future. *Cancer Res* 2017; 77(6):1261-1270. [PMID: 28254862].
9. Valent P, Akin C, Metcalfe D. Mastocytosis: 2016 updated WHO

- classification and novel emerging treatment concepts. *Blood* 2017; 129(11):1420-1427. [PMID: 28031180].
10. Sokol KC, Ghazi A, Kelly BC, Grant JA. Omalizumab as a desensitizing agent and treatment in mastocytosis: a review of the literature and case report. *J Allergy Clin Immunol Pract* 2014; 2: 266–70. [PMID: 24811015].
 11. Unterstell N, Lavorato FG, Nery NS, et al. Dermatoscopic findings in telangiectasia macularis eruptiva perstans. *An Bras Dermatol* 2013; 88(4): 643-645. [PMID: 24068144].
 12. Vannorsdall EJ, Collins JA, Chen QC, et al. Symptomatic response to imatinib mesylate in cutaneous mastocytosis associated with chronic myelomonocytic leukemia. *Curr Oncol* 2013; 20(4): e349-e353. [PMID: 23904774].
 13. Ragi J, Lazzara DR, Harvell JD, Milgraum SS. Telangiectasia Macularis Eruptiva Persians Presenting as Island Sparing. *J Clin Aesthet Dermatol* 2013; 6(4): 41-42. [PMID: 23630642].
 14. Troeltzsch MM. Cutaneous mastocytosis as a rare differential diagnosis for unilateral chronic facial pain and erythema: a case report. *J Orofac Pain* 2013; 27: 367-371. [PMID: 24171187].
 15. Mihalik N, Hidvégi B, Hársing J, et al. Klinikai tapasztalataink cutan mastocytosisban. *Orv Hetil* 2013; 154: 1469-1475. [PMID: 24016753].
 16. Noda SS. Unilateral telangiectasia macularis eruptiva perstans with a clear demarcation at the midline of the trunk. *Eur J Dermatol* 2012; 22: 407-408. [PMID: 22640982].
 17. PaTel:NN. Unilateral telangiectasia macularis eruptiva perstans of the breast. *Cutis* 2012; 90: 26-28. [PMID: 22908729].
 18. Kagimoto Y, Mizuashi M, Iizawa O, Aiba S. Linear telangiectasia macularis eruptiva perstans mimicking acquired nevoid telangiectasia in a 5-year-old girl. *Int J Dermatol* 2012; 51(1): 69-71. [PMID: 22182381].
 19. Altiner A, Tzu J, PaTel:R, et al. Telangiectasia macularis eruptiva perstans. *Dermatol Online J* 2011; 17(10): 7. [PMID: 22031633].
 20. Costa DL, Moura HH, Rodrigues R, et al. Telangiectasia Macularis Eruptiva Perstans: A Rare Form of Adult Mastocytosis. *J Clin Aesthet Dermatol* 2011; 4(10): 52-54. [PMID: 22010057].
 21. Watkins CE, Bokor WB, Leicht S, et al. Telangiectasia macularis eruptiva perstans: more than skin deep. *Dermatol Reports* 2011; 3(1): e12. [PMID: 25386256].
 22. Sotiriou E, Apalla Z, Loannides D. Telangiectasia macularis eruptiva perstans successfully treated with PUVA therapy. *Photodermatol Photoimmunol Photomed* 2010; 26(1): 46–47. [PMID: 20070840].
 23. Cengizlier R, Hücümenoğlu S, Ozen A, Tülin Sayli R. Treatment of telangiectasia macularis eruptiva perstans with montelukast. *Allergol Immunopathol (Madr)* 2009; 37(6): 334-6. [PMID: 19913347].
 24. Oliveira CR, Albuquerque GC, Simon EF, et al. Caso para diagnostic. *Telangiectasia macularis eruptiva perstans*. *An Bras Dermatol* 2009; 84(1): 87-89. [PMID: 19377767].
 25. Kowalczyk L, Eickenscheidt L, Seidel C, et al. Telangiectasia macularis eruptiva perstans, a form of cutaneous mastocytosis, associated with malignant melanoma. *J Dtsch Dermatol Ges* 2009; 7: 360–362. [PMID: 19054427].
 26. Akay BN, Kittler H, Sanli H, et al. Dermatoscopic findings of cutaneous mastocytosis. *Dermatology* 2009; 218(3): 226-30. [PMID: 19060465].
 27. Turchin I, Barankin B, Schloss E. Unusual cutaneous findings of urticaria pigmentosa and telangiectasia macularis eruptiva perstans associated with marked myelofibrosis. *Int J Dermatol* 2006; 45(10): 1215-1217. [PMID: 17040445].
 28. Kalay Erbay S, Stewart C, Hassanein A, et al. Cutaneous mastocytosis in a patient with primary Sjögren's syndrome. *J Rheumatol* 2006; 33: 1697–1700. [PMID: 16881127].
 29. Rishpon AA. Telangiectasia macularis eruptiva perstans: unusual presentation and treatment. *Skinmed* 2006; 5: 300-302. [PMID: 17086000].
 30. Lee HW, Jeong YI, Choi JC, et al. Two cases of telangiectasia macularis eruptiva perstans demonstrated by immunohistochemistry for c-kit (CD 117). *J Dermatol* 2005; 32(10): 817-20. [PMID: 16361734].
 31. Ruiz Villaverde R, Hernández Jurado I, Sánchez Cano D. Telangiectasia macular eruptiva Perstans y síndrome de Conn. *An Med Interna* 2005; 22(8): 399. [PMID: 16358431].
 32. Neri I, Guareschi E, Guerrini V, Patrizi A. Familial telangiectasia macularis eruptiva perstans. *Pediatr Dermatol* 2005; 22(5): 488-9. [PMID: 16191009].
 33. Nguyen NQ. Telangiectasia macularis eruptiva perstans. *Dermatol Online J* 2004; 10(3): 1. [PMID: 15748571].
 34. Martin LK, Romanelli P, Ahn YS, Kirsner RS: Telangiectasia macularis eruptiva perstans with an associated myeloproliferative disorder. *Int J Dermatol* 2004; 43: 922–924. [PMID: 15569020].
 35. Pascual JC, Bañuls J, Albares MP, et al. Presentation of telangiectasia macularis eruptiva perstans as a long-standing solitary plaque associated with renal carcinoma. *J Cutan Med Surg* 2003; 7(5): 399-402. [PMID: 14973646].
 36. Suzuki KK. Telangiectasia macularis eruptiva perstans in polycythemia rubra vera. *Eur J Dermatol* 2002; 12: 201-203. [PMID: 11872425].
 37. Kalaycayan AK, Kotogyan A. Telangiectasia macularis eruptiva perstans. *J Eur Acad Dermatol Venereol* 2001; 15: 263–264. [PMID: 11683296].
 38. Chang A, Tung RC, Schlesinger T, et al. Familial cutaneous mastocytosis. *Pediatr Dermatol* 2001; 18: 271–276. [PMID: 11576397].
 39. Chung-Leddon J. Telangiectasia macularis eruptiva perstans. *Dermatol Online J* 2000; 6(1): 6. [PMID: 11328616].
 40. Vidal CC. Telangiectasia macularis eruptiva perstans presented as a pseudoallergic food reaction. *J Investig Allergol Clin Immunol* 2000; 10: 248-250. [PMID: 11039843].
 41. Betti RR. Guess what! Telangiectasia macularis eruptiva perstans involving the upper arms in an adult male. *Eur J Dermatol* 2000; 10: 563-564. [PMID: 11056433].
 42. Bachmeyer C, Guillemette J, Blum L, et al. Telangiectasia macularis eruptiva perstans and multiple myeloma. *J Am Acad Dermatol* 2000; 43(5 Pt 2): 972-4. [PMID: 11044837].
 43. Soter NA. Mastocytosis and the skin. *Hematol Oncol Clin North Am* 2000; 14: 537–545. [PMID: 10909039].
 44. Gibbs NF, Friedlander SF, Harpster EF. Telangiectasia macularis eruptiva perstans. *Pediatr Dermatol* 2000; 17(3): 194–197. [PMID: 10886750].
 45. Biedermann T, Rueff F, Sander CA, Przybilla B. Mastocytosis associated with severe wasp sting anaphylaxis detected by elevated serum mast cell tryptase levels. *Br J Dermatol* 1999; 141: 1110–1112. [PMID: 10606862].
 46. Tebbe B, Stavropoulos PG, Krasagakis K, Orfanos CE. Cutaneous mastocytosis in adults. *Dermatology* 1998; 197(2): 101-108. [PMID: 9732155].
 47. Sarkany RP, Monk BE, Handfield-Jones SE. Telangiectasia macularis eruptiva perstans: a case report and review of the literature. *Clin Exp Dermatol* 1998; 23(1): 38–39. [PMID: 9667109].
 48. Ellis DL. Treatment of telangiectasia macularis eruptiva perstans with the 585-nm flashlamp-pumped dye laser. *Dermatol Surg* 1996; 22(1): 33-7. [PMID: 8556255].
 49. Donahue JG, Lupton JB, Golichowski AM. Cutaneous mastocytosis complicating pregnancy. *Obstet Gynecol* 1995; 85(5 Pt 2): 813-5. [PMID: 7724121].
 50. Cohn MS, Mahon MJ. Telangiectasia macularis eruptiva perstans. *J Am Osteopath Assoc* 1994; 94(3): 246-8. [PMID: 8200829].
 51. Granier F, Bourgeois-Droin C, Pedreiro JP, et al. Mastocytose systémique à type de telangiectasa macularis eruptiva perstans d'aspect poikilodermique. *Ann Dermatol Venereol* 1991; 118: 817-

818. [PMID: 1789642].
52. Clark DP, Buescher L, Havey A. Familial Urticaria Pigmentosa. *Arch Intern Med* 1990; 150(8): 1742-1744. [PMID: 2383167].
 53. Milgraum SS, Headington JT. A Subungual Nodule of Recent Onset. *Arch Dermatol* 1988; 124(3): 429-430. [PMID: 3345093].
 54. Fried SZ, Lynfield YL. Unilateral facial telangiectasia macularis eruptiva perstans. *J Am Acad Dermatol* 1987; 16(1 Pt 2): 250-2. [PMID: 3819061].
 55. Bonnefoy M, Rouchouse B, Clavreul C, et al. Pertes de connaissance prolongées révélatrices d'une mastocytose. A type de telangiectasia eruptiva macularis perstans. *Ann Dermatol Venereol* 1986; 113(3): 259-62. [PMID: 3752866].
 56. Grænerus G, Olafsson JH, Roupe G. Studies on histamine metabolism in mastocytosis. *J Invest Dermatol* 1983; 80(5): 410-6. [PMID: 6841997].
 57. Menzel I, Milbradt R. Teleangiectasia macularis eruptiva perstans Weber. *Hautarzt* 1980; 31(3): 145-9. [PMID: 7399904].
 58. Monheit GD, Murad T, Conrad M. Systemic mastocytosis and the mastocytosis syndrome. *J Cutan Pathol* 1979; 6(1): 42-52. [PMID: 438392].
 59. Cammer W, Norton WT. Disc gel electrophoresis of myelin proteins: new observations on development of the intermediate proteins (DM-20). *Brain Res* 1976; 109: 643-648. [PMID: 1276940].
 60. Klotz L. Teleangiectasia macularis eruptiva perstans. Beitrag zur Kenntnis einer Sonderform der Urticaria pigmentosa. *Hautarzt* 1970; 21(8): 372-5. [PMID: 5518372].
 61. Beltrani GG. Teleangiectasia macularis eruptiva perstans con mastocitosi. *Minerva Dermatol* 1966; 41: 436-442. [PMID: 5997511].
 62. Cramer HJ. Teleangiectasia macularis eruptiva perstans, eine besondere Form der Urtikaria pigmentosa. *Hautarzt* 1964; 15: 370-4. [PMID: 14251426].
 63. de BERGH, Mali JW. Teleangiectasia macularis eruptiva perstans. *Ned Tijdschr Geneesk* 1962; 106: 1050-1052. [PMID: 13867710].
 64. Moursun MP, Hirschmann VR. Telangiectasia macularis eruptiva perstans; Review of the Literature, Report of a Case and Discussion of the Etiology and Pathology of Generalized Telangiectasia. *AMA Arch Derm Syphilol* 1951; 63(2): 232-249. [PMID: 14789240].
 65. Mitchell-Heggs GB, Crow KD. Telangiectasia macularis eruptiva perstans. *Proc R Soc Med* 1946; 39(11): 686. [PMID: 21001838].
 66. Warren CM. Telangiectasia Macularis Eruptiva Perstans. *Proc R Soc Med* 1945; 38(4): 146. [PMID: 19993022].
 67. Forman L. Urticaria Pigmentosa in an Adult (Telangiectasia Macularis Eruptiva Perstans, Barber and Parkes Weber Type). *Proc R Soc Med* 1943; 36(6): 291. [PMID: 19992635].
 68. Goldsmith WN. Telangiectasia Macularis Eruptiva Perstans: Case for Diagnosis. *Proc R Soc Med* 1932; 25(12): 1742-3. [PMID: 19988982].
 69. Barber HW. Telangiectasia Macularis Eruptiva Perstans (Parkes Weber). *Proc R Soc Med* 1932; 25(7): 1029-1030. [PMID: 19988736].
 70. Weber FP. Telangiectasia Macularis Eruptiva Perstans—probably a telangiectatic variety of urticaria pigmentosa in an adult. *Proc R Soc Med* 1930; 24(2): 96-97. [PMID: 19987891].