Research Paper

Volume : 5 | Issue : 4 | April 2016 • ISSN No 2277 - 8179 | IF : 3.508 | IC Value : 69.48

Histopathological Study of A Rare Cutaneous Lesion- Solitary Mastocytoma

Medical Science

KEYWORDS : Mastocytoma, Solitary, Darier's sign

Dr. Mahendra V. Nagargoje — Assistant professor, Dept of Skin and VD, BKL Walawalkar Rural Medical College, Tal. Chiplun, Dist.Ratnagiri, Maharashtra, India, PIN-415606.

*** Dr.Aarti S. Salunke** — Assistant professor, Dept of Skin and VD, B. J. Govt. Medical College and Sassoon General Hospital, Pune, Maharashtra, India PIN-411001. * corresponding author

ABSTRACT

Mastocytosis is characterized by abnormal proliferation of mast cells, involving the skin (cutaneous mastocytosis) or extracutaneous organs (systemic mastocytosis). Skin is the most commonly involved organ, especially in children. Though often asymptomatic, systemic features may be associated with any clinical pattern of the disorder at any age group. Here we report rare entity, solitary cutaneous mastocytoma confirmed on histopathology and Giemsa stain.

INTRODUCTION

Mastocytosis represents a spectrum of clinical disorders with a common phenotype of tissue mast cell hyperplasia. The signs and symptoms of mast cell diseases are varied,depends on localisation of mast cells in different organs and the local and systemic effects of mediators released from these cells.[1] Cutaneous mastocytosis is heterogenous group of rare disorder and classified as generalised urticaria pigmentosa, solitary mastocytoma, diffuse mastocytosis, telangiectatic macularis eruptive perstance.[2],[3]Herein we describe a rare case of solitary cutaneous mastocytoma presenting as a solitary nodule over the neck.

CASE STUDY

Two and half year old girl brought by mother, who noticed single mildly itchy lesion over right side of neck (Figure-1) since 5 months of her age.

Figure-1 Solitory nodular lesion over lateral aspect of neck

There was no family history of any significant skin disorder. Symptoms like abdominal pain, diarrhoea, palpitation, syncope, bone pain or weight loss were absent. There was no evidence of lymphadenopathy or Hepatosplenomegaly. On cutaneous examination, a solitary, skin coloured nodule of 1.5 x 1 cm was observed over right side of neck. Stroking of lesion resulted in a wheal with flare (positive Darier's sign) (Figure-2).

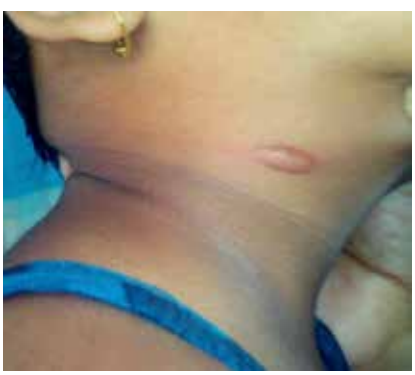

Figure-2 Incresed size and edema over lesion on stroking of lesion (Darier's sign)

This was not associated with blister formation over nodule Differential diagnoses of localized mastocytoma, juvenile xanthogranuloma, pseudolymphoma were considered. Routine investigations such as complete blood count, liver function test, renal function test, blood coagulation profile and urine examination were within normal limits. Chest X-ray, ultra sonography of abdomen-pelvis and bone marrow examination were performed to rule out systemic involvement, which revealed no significant abnormality. The lesion was completely excised, and the specimen send for histopathological examination. Micro sections showed tissue lined by thin keratinized stratified squamous epithelium, and dermis showed abundance of spindle shaped mast cell with eosinophilic cytoplasm infiltrating around the blood vessels, skin appendegeal structures and the entire dermis (Figure-3). The mast cells stained metachromatically with Giemsa stain (Figure-4) suggesting a final diagnosis of solitary cutaneous mastocytoma.

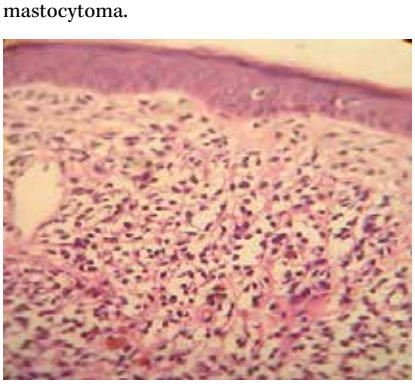

Figure-3 Diffuse infiltration of spindle shaped mastocyte in upper dermis (H&E)

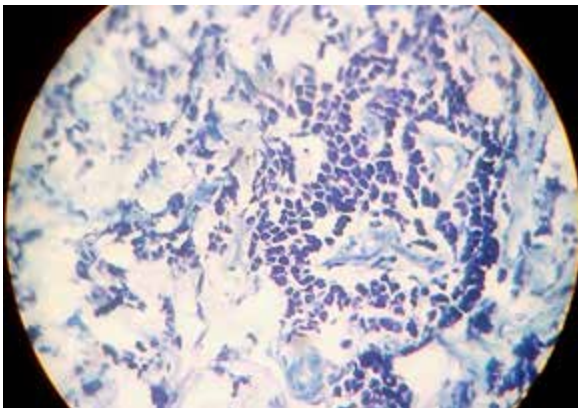

Figure- 4 Mast cells stained metachromatically (Giemsa stain)

DISCUSSION

The prevalence of mastocytosis in the general population is unknown. It has been reported to be present in one of 1,000 to 8,000 new patients evaluated at a dermatology clinic. Mastocytosis occurs in all races, and there is no sex predilection. The peak incidence is during infancy and early childhood, with a second peak occurring in middle age. The disease may be benign, with minor transient symptoms and signs that never cause the patient to consult a physician, or it may be life-threatening.[4] It is traditionally divided into cutaneous mastocytosis (CM) and systemic mastocytosis (SM). CM is divided into three major subtypes: urticaria pigmentosa (UP), mastocytoma, and diffuse cutaneous mastocytosis (DCM). Approximately 58~90% of patients with CM have the UP subtype, while 10~40% of affected patients have mastocytoma. DCM is the rarest subtype, accounting for only 1.74% of all cases of CM.[5]

The cutaneous form of solitary mastocytoma (SM) was first described by Nettleship in 1889. Since then, recognition of SM and mastocytosis has increased due to significant increase in the knowledge of these entities. Solitary mastocytomas are brownish nodules, which, when traumatized, may cause systemic symptoms such as pruritus (of varying intensity and severity), flushing, gastrointestinal complaints (nausea, colicky pain, diarrhoea) and headaches. Rarely, gastrointestinal bleed, asthma and hypotension may occur. [3] SM may be present at birth or they may arise during infancy, generally before 6 months of age. These lesions often appear on distal extremities, but can occure in any anatomic location. The symptoms are due to local and systemic release of histamine and other mast cell mediators like leukotriens, prostaglandins and platelet activating factors.[5][3] Rubbing or scratching a lesion of mastocytoma causes swelling, urtication, flare and sometimes blister formation due to release of histamine(Darier's sign).[6]

Cutaneous mastocytosis is mostly sporadic disorder but can be familial occurence. The exact etiology of mast cell proliferation is unknown. Various postulates include c-KIT receptor mutation, excess production of c-kit ligand or increase production of soluble form of mast cell growth factor (MGF).[7] There are indications that mutation in c-kit receptors of which MGF is a ligand may be responsible for chronic course in adults.[8]

The usual differential diagnosis of SM include melanocytic nevi, xanthoma, xanthogranuloma, neurofibroma. Histopathological features include increased number of mast cells with normal cytological appearance, increased melanin production and increased dermal fibroblasts.[9] The mast cells are usually oval or spindle shaped and predominantly found around blood vessels and skin appendages in papillary dermis. Toluidin blue or Giemsa stain usually identifies mast cells by their metachromatic granules in cytoplasm.[10] A skin biopsy should be taken with due care so as not to degranulate mast cells, which may results in false negative report.[11] Various tests such as complete hemogram(to look for anemia, thrombocytopenia, thrombocytosis, leukocytosis, eosinophilia), liver function tests, radiological skeletal survey (osteolytic lesions, osteoporosis, osteosclerosis) are needed to exclude the systemic involvement. These tests were normal in our patient and hence ruled out infiltration of organs other than skin.[12]

Solitary mastocytoma has benign clinical course and heals spontaneously in childhood without leaving any scarring. Dissemination of disease is a rare condition theoretically but there are no case reports in literature.[9] Treatment of solitary mastocytosis has mostly been palliative in the form of H1 and H2 antihistaminics, chromolyn sodium, aspirin, avoidance of triggering factors such as friction, ingestion of alcohol, NSAIDS, or temperature changes. Other modalities tried include topical and intralesional corticosteroid.[13] Resolution of solitary mastocytoma is usually expected in upto 10 years, which would be unacceptable to symptomatic patient.[3] Thus, in case of single or few, symptomatic lesions, surgical excision can be tried as a first line therapy, as done in our case.

CONCLUSION

Thus, to conclude we came across rare case of solitory cutaneous mastocytoma at unusual site. Aim of this case is to increase the awareness about mastocytoma masquerading as a solitary lesion with no other systemic association.

REFERENCES:

1. Andersen CL, Kristensen TK, Severinsen MT, Moller MB, Vestergaard H, Bergmann OJ,et al.,Systemic mastocytosis--a systematic review. Dan Med J. 2012 Mar;59(3):A4397
2. Poulton JK, Kauffman CL, Lutz LL, Sina B. Solitary mastocytoma arising at a hepatitis B vaccination site. Cutis 1999;63:37-40
3. Pandhi D, Singal A, Aggarwal S. Adult onset, hypopigmented solitary mastocytoma: Report of two cases. Indian J Dermatol Venereol Leprol 2008;74:41-3
4. Alto WA, Clarcq L. Cutaneous and systemic manifestations of mastocytosis. Am Fam Physician.1999;59:3047–3054
5. Lee EH, Kim MR, Kang TW, Kim SC. Diffuse Cutaneous Mastocytosis with Generalized Bullae.Ann Dermatol. 2010 Feb;22(1):77-80
6. Madke B, Nayak C. Eponymous signs in dermatology. Indian Dermatol Online J. 2012;3:159-65
7. Kitamura Y, Tsujimura T, Jippo T, Kasugai T, Kanakura Y. Regulation of development, survival and neoplastic growth of mast cells through the c-kit receptor. Int Arch Allergy Immunol 1995;107:54-6.
8. Longley J. Is mastocytosis a mast cell neoplasia or a reactive hyperplasia? Clues from the study of mast cell growth factor. Ann Med 1994;26:115-6.
9. Zekayi Kutlubay, Yardımcı G, Engin B,TüzünY. Cutaneous Mastocytosis- J Turk Acad Dermatol 2011; 5 (3): 1153r1.
10. Kiszewski AE, Alvarez-Mendoza A, Rios-Barrera VA *et al.* Mastocytosis in children: clinicopathological study based on 35 cases.Histopathology 2007;22:535–539.
11. Inamadar AC, Palit A. Cutaneous mastocytosis: Report of six cases. Indian J Dermatol Venereol Leprol 2006;72:50-3
12. Tamhankar PM, Suvarna J, Deshmukh CT. Cutaneous mastocytosis. Getting beneath the skin of the issue: a case report. Cases J. 2009; 2: 69.
13. Lange M., Bogusław Nedoszytko B., Górska A., Żawrocki A., Sobjanek M., Kozłowski D. Mastocytosis in children and adults: clinical disease heterogeneity. Arch Med Sci 2012; 8:533–541.