A case report of papular variant of maculopapular cutaneous mastocytosis.

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ABSTRACT:

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Although maculopapular cutaneous mastocytosis (MPCM) is the most common manifestation of cutaneous mastocytosis (CM), papular variant is quite rare. The subtype is clinically characterized by slightly tan or yellowish papules up to 1 cm in diameter.

We report case of a 9-month old male infant presented with multiple discrete brownish papular lesions on the face, trunk and extremities. Darier's sign was positive. The histopathology and special stainings were compatible with cutaneous mastocytosis. Complete physical examination revealed no hepatosplenomegaly. Laboratory investigations and pediatric dermatology consultation for further systemic involvements were performed. Although there was no evidence of systemic involvement at present, long term follow-up is recommended.

Key words: Papular variant of maculopapular cutaneous mastocytosis, cutaneous mastocytosis.

บทคัดย่อ:

มานิตา อัตถสุริยานันท์, โกวิท คัมภีรภาพ รายงานผู้ป่วยโรคแมสเซลล์สะสมที่ผิวหนังชนิดก้อนนูน วารสารโรคผิวหนัง 2560; 33: 167-172.

สถาบันโรคผิวหนัง กรมการแพทย์ กระทรวงสาธารณสุข

โรคแมสเซลล์สะสมที่ผิวหนัง สามารถจำแนกได้หลายกลุ่มตามลักษณะของรอยโรค โดยกลุ่มที่พบได้บ่อยที่สุดคือ maculopapular cutaneous mastocytosis ซึ่งกลุ่มนี้ยังสามารถจำแนกตามรอยโรคเป็นสองแบบ คือ urticaria pigmentosa และ papular variant โดยพบว่าผู้ป่วยที่มีรอยโรคแบบตุ่มนูนจำนวนมาก พบได้น้อย

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รายงานฉบับนี้เป็นการนำเสนอผู้ป่วยเด็กชายอายุ 9 เดือน ที่มาด้วยตุ่มนูนสีน้ำตาลที่ใบหน้า ลำตัวและแขนขา จากผลการตรวจทางพยาธิวิทยาและการย้อมพิเศษ ให้การวินิจฉัยว่าเป็นโรคแมสเซลล์สะสมที่ผิวหนังชนิดตุ่มนูน การตรวจร่างกายไม่พบตับม้ามโต ผลการตรวจนับเม็ดเลือด, ค่าการทำงานของตับ และระดับเอนไซม์ tryptase อยู่ในเกณฑ์ปกติ นอกจากนี้ยังได้ส่งผู้ป่วยไปปรึกษาตจกุมารแพทย์แล้ว ในขณะนี้ยังไม่พบว่ามีการกระจายของโรคไปยังอวัยวะอื่นๆ แต่อย่างไรก็ตามยังมีการนัดเพื่อติดตามอาการในระยะยาวต่อไป

คำสำคัญ: โรคแมสเซลล์สะสมที่ผิวหนังแบบตุ่มนูน, โรคแมสเซลล์สะสมที่ผิวหนัง

Case report

A 9-month old Thai male infant presented with multiple discrete brownish papules on the face, trunk and extremities for 3 months**.** The lesions began to develop as small papules on the face, trunk and extremities since he was 6 months old. He was born full-term and healthy with no apparent illnesses or abnormalities. He has no siblings and none of his family members has a similar condition**.** Physical examination revealed generalized well-defined skin colored to brownish papules scattering on the face, head, neck, trunk and extremities. Darier's sign was positive. Neither hepatosplenomegaly nor lymphadenopathy was observed. Glycerol preparation for scabies was negative, complete blood count (CBC) and liver function tests (LFTs) were also unremarkable. A 4-mm punch biopsy from the back was performed and displayed dense diffuse infiltration of mononuclear cells in the dermis. The epidermis was atrophic due to the massive dermal infiltration. No distinct exocytosis was observed. The cells showed round nuclei with pink cytoplasm. Neither pleomorphism nor increased mitosis was found. Toluidine blue staining demonstrated eosinophilic granules within cytoplasm of the tumor cells together with positive CD117 staining. Thus, the final diagnosis was consistent with papular variant of maculopapular cutaneous mastocytosis. He was treated with an oral antihistamine and topical 0.02% triamcinolone acetonide cream to apply the lesions twice a day. Furthermore, pediatric consultation was sought and no systemic involvement was found.

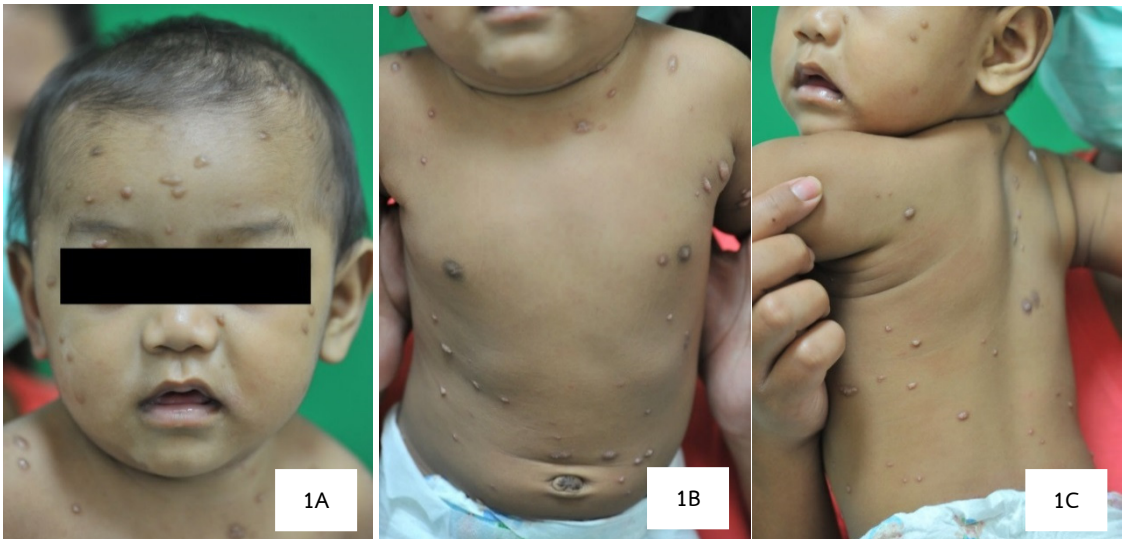


Figure 1A, 1B, 1C reveal generalized well-defined skin colored to brownish papules scattering on the face, head, neck, trunk and extremities

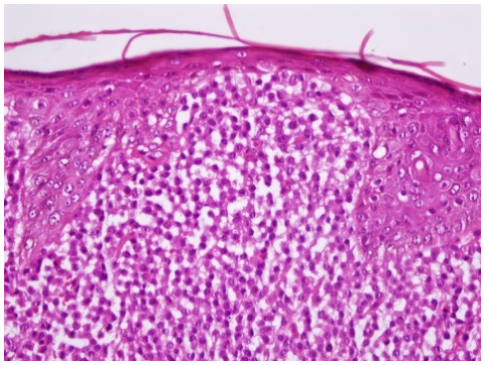
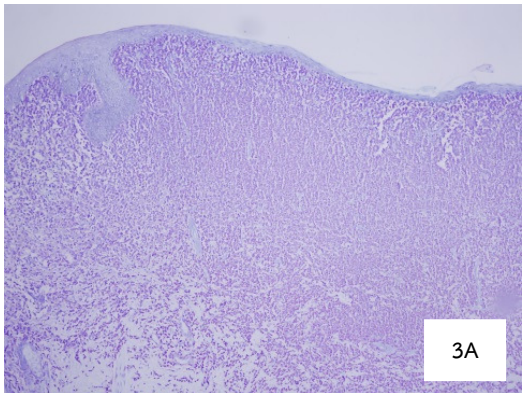


Figure 2 Histopathological image shows dense diffuse infiltration of mononuclear cells in the dermis. The epidermis was atrophic due to the massive dermal infiltration. No distinct exocytosis was found. The cells showed round nuclei with pink cytoplasm. Neither pleomorphism nor increased mitosis was found.

Discussion:

Mastocytosis is defined as a heterogeneous group of disorders characterized by an accumulation of mast cells in one or more organs, particularly in the skin, bone marrow, liver, spleen and lymph nodes.[1] It is traditionally divided into cutaneous mastocytosis (CM) and systemic mastocytosis. The most common clinical presentation is the cutaneous form. In childhood, CM typically manifests in cutaneous form and is transient while the adult onset commonly progresses to a systemic form.



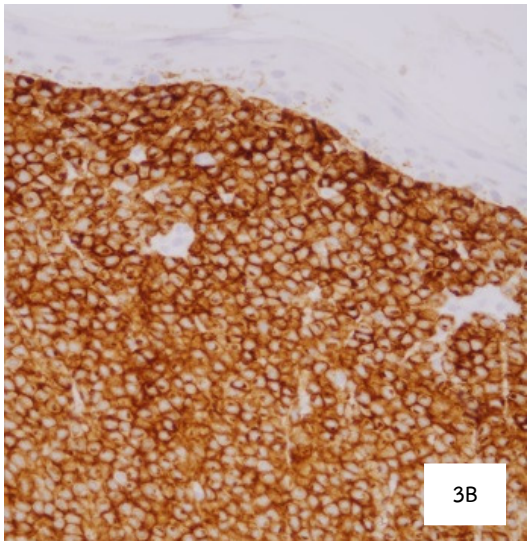


Figure 3A, 3B Toluidine blue staining demonstrated eosinophilic granules within cytoplasm of the tumor cells. CD117 staining was positive.

Regarding a systematic review of 1,747 cases conducted between 1950 and April 2014, 90% of the cases occurred before the age of 2 years, 75% presented with urticaria pigmentosa (UP), 20% with mastocytoma and 5% with diffuse cutaneous mastocytosis.[2]

Typical cutaneous lesions of UP are characterized by generalized eruption of round to oval tan-brown macules or slightly elevated papules. The highest density of lesions is usually on the trunk. Additionally, the face and mucous membranes may be involved while palms, soles, and scalp are often spared.[3] Pruritus is typical, but may be lacking in limited disease form. In 2002, a modified consensus proposal concerning classification of CM was presented by Hartmann and Henz (Table I).[4]

Table I Classification of cutaneous mastocytosis according to Hartmann and Henz[4]

Maculopapular cutaneous mastocytosis
Plaque type cutaneous mastocytosis
Nodular cutaneous mastocytosis/mastocytoma
Diffuse cutaneous mastocytosis
Telangiectatic cutaneous mastocytosis

Maculopapular cutaneous mastocytosis is the most common manifestation of CM. There are 2 clinical subsets namely UP and papular variant. The papular variant of MPCM consists of slightly tan or orange to yellowish papules up to 1 cm in diameter. This type of CM usually occurs in infancy, often within 1 or 2 months after birth. It has about 50% tendency for spontaneous involution before puberty.[5] Moreover; the multiple papular lesions are even rarer to be clinically encountered. The type of initial rash in patients with UP-type lesions was reported in 35% of cases. These were macules (20%), papules (12%), maculopapules (41%), plaques (15%) or nodular-type lesions (12%).[2]

CM is clinically diagnosed and confirmed by skin biopsy showing mast cell infiltration, predominantly surrounding blood vessels in the papillary and upper reticular dermis. Mast cells may be identified using traditional toluidine blue, Giemsa, chloroacetate esterase, or avidin stains. Immunohistochemical staining with tryptase is the most accurate stain for the detection of mast cells in mastocytosis in all tissues.[3] Systemic involvement must always be investigated.

Pediatric mastocytosis was previously considered to be a benign and self-limiting disease, and a literature review shows that partial or complete regression occurs in 67% of children. According to a systematic review, serum tryptase levels do not appear to have a significant prognostic value in pediatric mastocytosis. However, they do appear to be correlated with the extent of skin involvement. About 3% of cases of pediatric mastocytosis are aggressive and progress to mast cell sarcoma (MCS) or mast cell leukemia (MCL) and systemic disease with a fatal outcome.[2]

The treatment for most forms of pediatric mastocytosis is mostly conservative and symptomatic. Patients should be instructed to avoid precipitating causes of mast cell degranulation e.g. food (spicy foods, alcohol, cheese, hot beverages, etc.) and medications (aspirin, NSAIDs, codeine, morphine, opiates, procaine, radiographic dyes, dextran, polymyxin B, D-tubocurarine, etc.).[6]

In our case, the patient was diagnosed with papular variant of MPCM and was referred to pediatric dermatology department of Queen Sirikit National Institute of Child Heath (QSNICH) hospital for further systemic evaluation. Complete physical examination revealed no hepatosplenomegaly. The results of CBC, LFTs, and serum tryptase were within normal limits. Although there is no systemic involvement at the moment, a long term follow up is recommended.

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