

SHORT COMMUNICATIONS

Unique Presentation of Urticaria Pigmentosa as a Subcutaneous Mass

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Urticaria pigmentosa (UP) is one of the most common clinical variants of cutaneous mastocytosis. It can present clinically as multiple red-brown macules and rarely plaques and nodules. Childhood cases have a good prognosis usually without systemic involvement.¹ Our case describes UP uniquely presenting as a subcutaneous mass in the perineum of a pediatric patient.

A 9-year-old male with history of tan-brown macules starting at age 4 with punch biopsy consistent with UP (Figure 2) was managed with antihistamines (levocetirizine and montelukast) and close observation until he presented with an asymptomatic lump of the right perineum with no skin surface change. An ultrasound revealed a 1.7 cm hypoechoic solid lesion (Figure 1) for which excisional biopsy stained diffusely positive for CD68 and CD117, with rare positive S100 and CD1a cells, consistent with UP. Serum tryptase levels were within normal limits. The patient received antihistamines and full body 6-week UVB light treatment with significant improvement of his cutaneous lesions and complete resolution of the subcutaneous perineal mass.

Figure 1. Ultrasound of the perineal lump revealed a 1.7cm hypoechoic solid lesion.

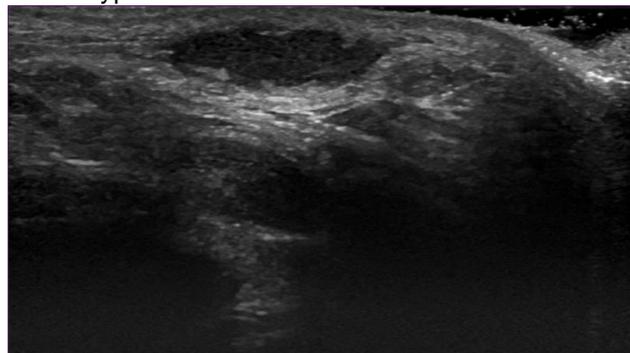


Figure 2. Tan-brown macules involving the posterior auricular area, chest and abdomen.



Urticaria pigmentosa (UP) is the most common presentation of cutaneous mastocytosis in children.¹ Children typically present with widespread distribution of tan macules and rarely with nodules or plaques, most commonly on the trunk.^{1,5} It can present at birth but typically appears in infancy and childhood and gradually improves by adolescence. Darier's sign, urtication of UP lesions following scratching, may occur.¹

There have been few reports of female pediatric patients presenting with a single mastocytosis lesion in the vulvar regions, but no cases of mastocytosis presenting as a subcutaneous mass in the perineum of males have been reported in the literature.^{4,6}

Evaluation and diagnosis of UP are often made clinically but can be confirmed with a biopsy. Systemic tests are rarely needed but can be deemed useful if there is a concern of systemic mastocytosis involvement such as diarrhea, gastric ulcers, flushing reactions, headaches, and failure to thrive.⁵ These tests include a CBC with differential, complete chemistry panel, and serum tryptase level which would be elevated in systemic mastocytosis but negative in cutaneous mastocytosis.¹ UP that begins after age 10 usually persists and can be associated with systemic disease.

Treatment focuses on controlling symptoms associated with the lesions. H₁ blockers can be effective against itching and flushing symptoms. Topical steroids can also be used for symptomatic relief.¹ Phototherapy is a second-line therapy which may be considered when there is poor symptomatic control or progression of disease, such as what occurred in our patient. Avoiding mast cell degranulation is another aim of therapy.

Physical granulators include temperature changes such as extreme heat or cold and

mechanical factors such as trauma or irritation. Chemical granulators include alcohol, aspirin, NSAIDs, and iodinated contrast media.^{1,5} Precautions should be taken when placing a pediatric patient with UP under systemic anesthesia. Morphine and certain neuromuscular blockers such as rocuronium and atracurium should be avoided as they have been associated with a significant incidence of anaphylaxis due to mast cell degranulation.^{2,3} Propofol is considered a safer recommendation for general anesthesia for patients with mastocytosis. Other safe hypnotics agents include etomidate and ketamine.³

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References:

1. Briley LD, Phillips CM. Cutaneous mastocytosis: a review focusing on the pediatric population. *Clinical pediatrics*. 2008;47(8):757-761.
2. Dewachter P, Castells MC, Hepner DL, Mouton-Faivre C. Perioperative management of patients with mastocytosis. *Anesthesiology: The Journal of the American Society of Anesthesiologists*. 2014;120(3):753-759.
3. Klein NJ, Misseldine S. Anesthetic considerations in pediatric mastocytosis: a review. *Journal of anesthesia*. 2013;27(4):588-598
4. Serarslan, Gamze, Esin Atik, and S. Zeteroglu. "Nodular mastocytosis of the vulva: an unusual localisation." *Australian and New Zealand journal of obstetrics and gynaecology* 45.4 (2005): 335-336.
5. Zitelli, Basil J., Sara C. McIntire, and Andrew J. Nowalk. *Zitelli and Davis' Atlas of Pediatric Physical Diagnosis E-Book: Expert Consult-*

- Online*. Elsevier Health Sciences, 2017:8, 275-340.
6. Shuangshoti S, Shuangshoti S, Pintong J, Piyayotai V, Sukpanichnant S. Solitary mastocytoma of the vulva: report of a case. *Int J Gynecol Pathol*. 2003; 22: 401– 403.