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Nodular Mastocytosis of the Vulva and Coexisting Urticaria Pigmentosa

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Abstract

Nodular forms of mastocytosis are rather rare skin diseases, especially when localized on the vulva. A 9-year-old girl presented with urticaria pigmentosa type lesions since her 4th year, associated with several solitary or confluent vulvar nodules, varying in size from a pea to a walnut, and mild systemic symptoms. Diagnosis of mastocytosis was confirmed by histology, and apart from splenomegaly, no signs of systemic spread or associated hematologic disorders were detected. Therapeutic response of nodular lesions was rather poor, and further follow up is necessary.

Mastocytoses are a group of disorders with an increased number of mast cells in the bone marrow, skin, spleen, liver, lymph glands, or gastrointestinal tract. The skin is the most frequently affected organ, with several forms of cutaneous presentations: urticaria pigmentosa (maculopapular), nodular, diffuse and telangiectatic (1-3). Nodular mastocytosis is rare, localized usually on the limbs, scalp or trunk. There have been a few reports in the literature about mastocytosis localized on the vulva (4-7).

Case report

A 9-year-old girl presented with a 5-year previous history of rash on her head and trunk, and nodules in the genital area. Skin lesions appeared for the first time at the age of 4, with sore, red and itchy nodules on the vulva. Papular pruritic eruptions and blisters on the head and trunk appeared later, with residual macular pigmentation. Erythema of the trunk, with intense pruritus, used to occur after warming. On several occasions, heart palpitation lasting for 2-3 minutes, was accompanied by blushing of the face, eyes and trunk, but resolved spontaneously. After administration of the Ibuprofen syrup, for febrile upper respiratory tract infection, a generalized itchy erythematous eruption appeared, with wheals over the pigmented patches.

The skin lesions on the vulva presented with multiple skin-coloured nodules, pea to walnut-size, solitary or confluent, in tumorous formations on her right labium majus (Figure 1). Pigmented maculopapules with positive Darier's sign were visible on the trunk, forehead, scalp and limbs (Figure 2.). Apart from eosinophilia (0,793x10^9/L) and elevated total serum protein level of 85.9 g/L (normal value up to 80 g/L), other laboratory and biochemical findings, were within normal values. Peripheral blood smear, sternal punctate, bone radiography, control values of differential white blood count (eosinophiles 0.348x10^9/L) were within normal values. Abdominal ultrasound scans revealed splenomegaly (spleen diameter 98x30 mm, normal value up to 88 mm). Ultrasound examination of the pelvis revealed normal anatomy of internal genital organs. Echocardiography showed an aberrant chord of the left ventricle, with normal electrocardiogram.

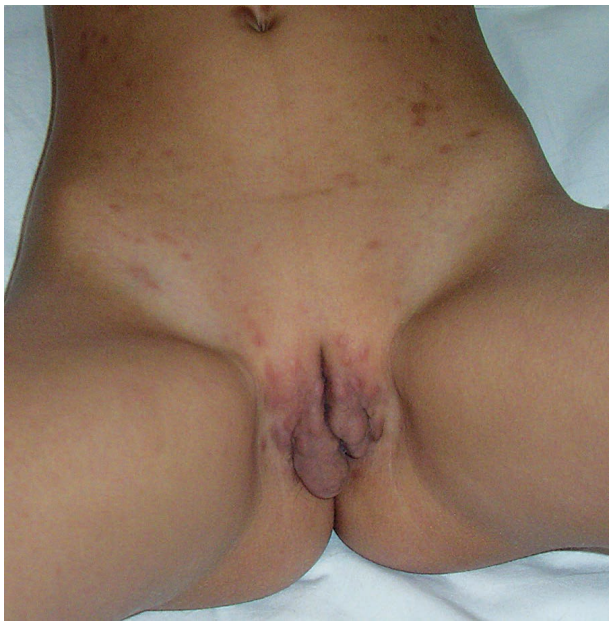

Figure 1. Numerous skin-coloured nodules of the vulva, pea to walnut-size, solitary and confluent in tumorous formations on the right labium majus

Pathohistological findings of the maculopapular truncal lesions and vulvar nodules were almost the same (Giemsa stain): perivascular dermal mast cell infiltration of medium density, with discrete epidermal hyperkeratosis (Figure 3).

Peroral hydroxyzine, 3x12.5 mg daily, and betamethasone dipropionate 0.05% ointment were applied to the vulvar lesions. Avoidance of key triggers of mast cell degranulation, such as extreme temperatures, certain medications (Ibuprofen) and insect stings was recommended. On control visit, 2 months later, better control of flushing and pruritus was achieved, while the local status on the vulva remained unchanged. Further follow-up was advised.

Discussion

Mastocytosis is characterized with abnormal accumulation of mast cells, associated with wide spectrum of local or systemic symptoms. Although this accumulation may affect all tissues, it most frequently affects the skin. It affects both sexes equally, with bimodal distribution and two peaks: in early childhood and in adulthood, during the second to fourth decade. About 2/3 of patients are children, one half are under the age of 2 years, and about 10% are children 2 to 14 years of age (8). The clinical picture differs in childhood and adulthood, both regarding the course and association with c-kit ligand mutation (9).

Among different cutaneous types (maculopapular urticaria pigmentosa, nodular, diffuse and telangiectatic), urticaria pigmentosa is the most frequent (70 - 90%) type in children, followed by solitary mastocytoma (1-3). Urticaria pigmentosa most frequently affects infants between the age of 3 and 9 months, in the form of numerous red-brownish pigmented maculas, papules and nodules located on the trunk (8). Nodular mastocytosis is a rare type, localized on limbs, face, scalp or trunk (10-12).

The diagnosis of mastocytosis, based on clinical picture and Darier's sign, was confirmed by presence of metachromatic granules in the mast cells with Giemsa staining of skin biopsy samples. In patients with general symptoms, additional laboratory analyses are recommended, but invasive diagnostic methods (bone marrow biopsy) are seldom necessary (13,14). In our patient, splenomegaly and eosinophilia raised the possibility of a systemic disease. Hepatosplenomegaly may be a sign of systemic mastocytosis in children. Cases of urticaria pigmentosa, associated with hematological malignancies have been reported, most frequently

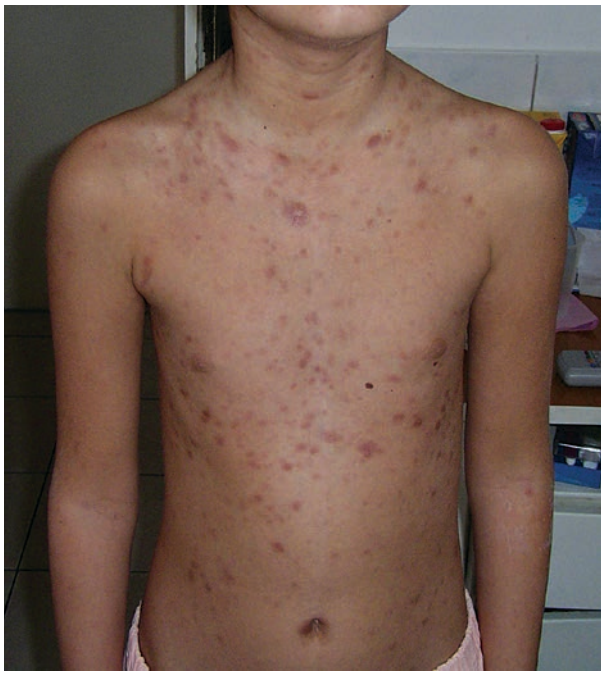

Figure 2. Urticaria pigmentosa lesions on the trunk

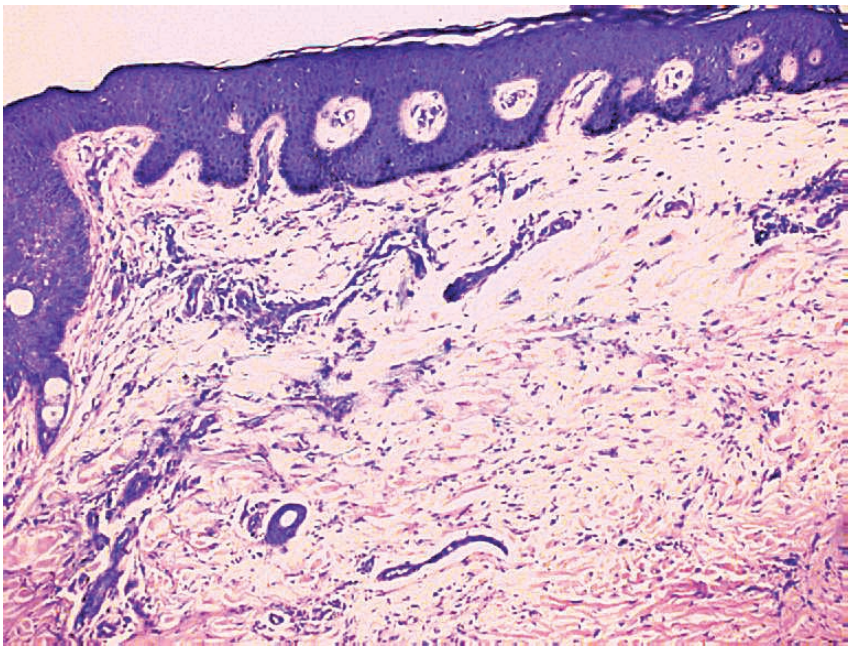

Figure 3. Perivascular dermal mast cell infiltration of medium density, discrete epidermal hyperkeratosis (Giemsa stain, x 100)

with acute myeloid or acute lymphoblastic leukosis (15). A nodular skin mastocytosis, associated with systemic spread, was described in literature (16). In our patient sternal punctate was performed; normal findings excluded the impairment of the hematopoetic system and associated hematological disorders.

Multiple nodular lesions of mastocytosis are rare, usually localized on limbs, face, scalp, trunk, or palms and soles (10,11). Nodular mastocytosis of the vulva is exceedingly rare. Since vulva is an unusual localization, mastocytosis should be considered in the differential diagnosis of such nodular lesions. Schuangshoti et al. (5) described a solitary mastocytoma on labia majora, without truncal lesions. Serarslan et al. (6) first described multiple nodular lesions of the vulva, similar to xanthomata, associated with urticaria pigmentosa. In our case nodules on the vulva were multiple, of normal color of the overlying skin.

Mastocytosis in children can be associated with systemic or local symptoms of mediator release: itch, palpitations, headache and episodic flushing or wheals that can arise spontaneously or after mechanical, physical or chemical stimulation. In our patient, after Ibuprofen administration, generalized erythema of the skin occurred, with a wheal over the pigmented macules.

This patient was diagnosed with benign cutaneous mastocytosis. Associated general symptoms are not predictive for systemic mastocytosis, but splenomegaly associated with increased eosinophil count and nodular skin lesions requires further follow up.

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Nodularna mastocitoza vulve udružena sa urtikarijom pigmentozom

Sažetak

Uvod: Nodularna mastocitoza je redak oblik mastocitoze, posebno kada je lokalizovana na vulvi. Prikaz bolesnika: Prikazujemo devojčicu uzrasta 9 godina, sa promenama tipa pigmentne urtikarije, koje traju od njene četvrte godine, udružene sa pojedinačnim i slivenim nodusima na vulvi, veličine zrna graška do veličine oraha. U više navrata imala je osećaj lupanja srca u trajanju od 2-3 minuta, uz crvenilo i intenzivan svrab očiju, lica i tela, koje je spontano prolazilo. Osim eozinofilije (0,793x10^9/l) i ukupnih proteina 85,9 g/l, laboratorijski nalazi su u granicama referentnih vrednosti. Izuzev splenomegalije, nisu utvrđeni znaci sistemske mastocitoze ili udruženih hematoloških oboljenja. Dijagnoza mastocitoze potvrđena je patohistološkim nalazom kožnih promena na vulvi i trupu. Terapijom hidroksizinom per os i lokalnom aplikacijom betametazon dipropionata 0,05% na noduse na vulvi, postignuta je bolja kontrola „napada crvenila“ i svraba, dok je nalaz na vulvi ostao nepromenjen.
Zaključak: Kod devojčice je postavljena dijagnoza benigne kutane mastocitoze. Opšti simptomi sami po sebi ne ukazuju na sistemsku bolest, ali je zbog prisustva spelomegalije i eozinofilije potrebno dalje praćenje bolesnika.