

URTICARIA PIGMENTOSA TOGETHER WITH BRONCHIAL ASTHMA : A CASE REPORT

Nazan TOMAÇ, M.D., Abdurrahman KARA, M.D., Ayşegül OKSAL*, M.D.,
Alev EKEN**, M.D., Kenan BEK, M.D.

Sami Ulus Children's Hospital, Departments of Pediatrics, Pathology*, and Dermatology**,
Ankara, Turkey

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SUMMARY :

Despite the putative role of the mast cell in causing both bronchial asthma and urticaria pigmentosa, the occurrence of asthma together with urticaria pigmentosa is uncommon. In this case report we describe an 11 year-old-girl with bronchial asthma and urticaria pigmentosa.

Key Words: *Urticaria Pigmentosa, Bronchial Asthma.*

INTRODUCTION

Mastocytosis is a disorder of mast cell proliferation that occurs in both cutaneous and systemic forms. It may be due to altered cutaneous metabolism of mast cell growth factor, and may represent a hyperplastic rather than a neoplastic disorder(1). The most frequent site of organ involvement in patients with any form of mastocytosis is the skin, in which a variety of clinical manifestations have been described. Cutaneous lesions include mastocytoma (single or multiple macules, plaques or nodules), urticaria pigmentosa (multiple macules, papules, and plaques), diffuse and erythrodermic forms including bullous mastocytosis, and telangiectasia macularis eruptiva perstans.

Urticaria pigmentosa (UP), is a common cutaneous manifestation of mastocytosis and occurs primarily in infants and children. Asthma and elevated serum IgE levels are uncommonly reported in pediatric-onset mastocytosis. A patient with UP and asthma with hyper IgE is presented and

the relationship between these two conditions is discussed in this report.

CASE REPORT

An 11-year-old Caucasian girl was admitted with a five-year history of rhinitis, coughing, wheezing, and dyspnea attacks. Asthma symptoms were twice weekly and precipitated with nonspecific irritants. There was no family history of atopy. She had red-brown plaques two years ago without any other complaints such as pruritus or flushing.

Physical examination revealed diffuse brown macules, wheezing on lung auscultation and negative Darrier's sign (Fig. 1). A complete blood count, bone marrow specimens, and skeletal X-ray surveys were normal. Serum IgE level was elevated to 793 (normal 0-150) kU/L and allergy skin prick testing (Alk, Allergologisk Laboratorium A/S, Denmark) was positive with grass pollen and animal dander. Pulmonary function tests (Pony spirometry, Cosmed, Italy) showed mild to



Fig - 1 : Both large and small pigmented lesions in the child.

moderate obstruction reflected by a reduction in FEV1 which improved shortly after the administration of a bronchodilator drug. She was diagnosed as bronchial asthma in accordance with the American Thoracic Society's standards (2).

Light microscopy of the lesional skin revealed hyperkeratosis, akantosis in the dermis, and perivascular mononuclear cell infiltration in the superficial dermis. Toluidine blue stain demonstrated excessive mast cell infiltration around blood vessels.

The patient was treated with inhalation, corticosteroids. At follow-ups, both her symptoms and pulmonary function tests improved.

DISCUSSION

UP is an uncommon disease in both children and adults. It is a cutaneous form of mastocytosis. Systemic form of mastocytosis is more common in adults than in children and characterized with lymphoreticular, bone marrow, skeletal, and gastrointestinal system involvement (1).

Azana et al. (3), have reported that the most common initial symptom in pediatric-onset mastocytosis is pruritus. Additional symptoms due to mast cell mediator release include flushing, gastrointestinal complaints (ie, diarrhea, abdominal pain/colic, vomiting), and in children less than 2 years of age, bullae.

Asthma is suggested to be an infrequent symptom in pediatric mastocytosis (4). As far as we

know, the association of UP and bronchial asthma has not been reported in detail, yet. Children with pediatric-onset mastocytosis appear to be at no greater risk of developing allergies than the general population. Measurements of IgE in these children were found within the normal limits (5). In contrast to these reports, some studies have shown that patients with UP and their relatives have increased history of hay fever and asthma (6, 7). Boyle et al. (8) have reported a patient with UP who had non-specific BHR to "Carbachol" and secondary elevation of the total serum IgE. Children with UP, typically have plasma histamine levels that range from normal to several times the normal level (5). Histamine is one of the most important mediators of mast cells and may cause asthma symptoms. The proportion of mast cells and the histamine content of bronchoalveolar lavage (BAL) fluid have been correlated with the degree of airflow obstruction and the level of bronchial hyperresponsiveness (BHR) (9). Current evidence suggests that early asthmatic reaction is predominantly mast cell mediated. BAL, performed on stable asthmatic patients, indicates that there is a significantly higher recovery of mast cells in patients with atopic and nonatopic asthma (10). Mast cells in BAL from asthmatic airways are not only more numerous but are in a heightened state of activation both in the unstimulated and stimulated state. In fact, mast cell activation accounts for the immediate bronchial response and may initiate bronchial inflammation and BHR. So, we expect BHR and asthmatic symptoms more frequent than reported before in patients with UP.

It will be useful to report patients with asthma and UP being together for the correct incidence of asthma in UP. In addition, since the clinical presentations and prognosis of mastocytosis are heterogenous, there may be several mechanisms for the mast cell infiltration in tissues (11). The recognition of mastocytosis syndromes may play an important role in understanding the pathogenesis of bronchial asthma.

Correspondence to : Dr.Nazan TOMAÇ
Dr.Sami Ulus Çocuk Hastanesi
Babür Caddesi
Telsizler
06080 ANKARA - TÜRKİYE
Phone : 312 - 317 07 07 / 364
Fax : 312 - 317 03 53

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