Abstract

Solitary mastocytosis in adulthood is a rare finding. Only two such lesions have been reported in the head and neck. We describe a 27-year-old woman who had a 10-year history of a forehead swelling that had fluctuated in size. Light trauma or pressure on the lesion resulted in an increase in its size. A mass was found to be situated just below the galea and was successfully removed surgically using a high forehead lift. Histologically, the specimen contained predominantly mast cells. A systemic mastocytosis was excluded by a multidisciplinary diagnostic approach and measurement of the 24-h urinary excretion of histamine metabolites. After 36 months of follow-up there has been no recurrence.

Key words  Head and neck solitary lesions · Mastocytosis · Histamine · Surgery

Introduction

Mastocytosis is an uncommon benign disorder consisting of a proliferation of mast cells [5]. Mast cells play a number of regulatory roles in the inflammatory response. The skin is the most frequent site of organ involvement in patients with both systemic and localized mastocytosis. About 15% of patients with cutaneous mastocytosis can present with a localized lesion, which is termed “mastocytoma” or “solitary mastocytosis.”

Solitary mastocytosis has been described as a round or oval slightly elevated plaque or nodule or macule and varies from light brown to red in color [3]. The lesion may fluctuate in size, and rubbing the lesion can produce Darier’s sign, which is a transient itching, erythema, and edema. Most mastocytomas appear on the extremities within the first 3 months of life [6]. Lesions often disappear spontaneously within 2–3 years and are rarely described in adults [1].

In some patients with solitary mastocytosis systemic symptoms occur as a generalized spontaneous flushing as the result of the release of mast cell-derived mediators, such as histamine, leukotrienes, and cytokines [5]. In such cases the determination of the histamine metabolites methyl-histamine (MH) and methylimidazole acetic acid (MIAA) is a more specific and sensitive method of diagnosis than the determination of histamine itself [4].

Measurement of histamine metabolites in urine can be used for screening purposes, although a persistent elevation can be found in patients with chronic myelocytic leukemia and polycythemia vera. Furthermore, the consumption of histamine-rich foods may involve a higher excretion of histamine metabolites. We report our findings in the management of a young woman with solitary mastocytosis of the forehead.

Case report

A 27-year-old white housewife was seen in the outpatient clinic of University Hospital, Nijmegen, for evaluation of a swelling at the middle of her forehead which had recurred periodically over the preceding 10 years. Approximately four or five times a year the swelling appeared spontaneously and then disappeared approximately 5 days later. At times the maximum size of the swelling was 2–3 cm, making it difficult for the patient to wear a motorcycle helmet. Rubbing the swelling seemed to make it bigger and strain the skin. There was no pruritus, pain, or redness. Previous computed tomography of the frontal sinus showed no abnormalities. Intracutaneous allergen tests had not revealed allergy. There was no flushing or increased dermographia.

On examination a 3 cm soft-tissue swelling was palpable on the middle of the forehead. This seemed to be attached to the skin and not to the underlying skull. The overlying skin did not show any
discoloration or blisters. General physical and otorhinolaryngological examinations showed no abnormalities. Hepatosplenomegaly and lymphadenopathy were absent. Dermatological evaluation did not reveal any cutaneous lesions. An ultrasound image of the forehead showed a solid subcutaneous swelling, while magnetic resonance imaging clearly showed the lesion to be localized adjacent to the frontal bone (Fig. 1). A fine-needle aspirate revealed a nonspecific inflammatory reaction. Routine blood investigations included an erythrocyte sedimentation rate, complete blood count, and liver function tests that were all normal.

Surgical removal of the forehead lesion was planned. A peritrichal incision was made from one ear to the other. A myocutaneous scalp-forehead flap was developed anterior in the subgaleal loose connective tissue plane until the tumor was reached. Tumor was found to be under the galea and attached to it, and was located above the pericranium. Although it had caused some pressure changes in the bone, it was not attached to it (Fig. 2). Tumor was removed without complications; the diameter was approximately 20 mm and thickness 4 mm. The wound was closed without incident.

Light microscopy of the hematoxylin-eosin section of the excised specimen showed that the tumor mainly consisted of a densely...