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Cutaneous mastocytosis in children: anaesthetic considerations

Despite considerable information and discussion about the risk of serious complications in patients with systemic mastocytosis undergoing general anaesthesia, little is known specifically about the risk to patients with isolated cutaneous mastocytosis. The experience of 29 general anaesthetics in 12 children with urticaria pigmentosa and three with solitary cutaneous mastocytoma was reviewed. No major complications were encountered and the four minor problems seen were self-limiting. The data from this study do not suggest that patients with urticaria pigmentosa or solitary cutaneous mastocytoma are at increased risk of life-threatening complications under general anaesthesia.

Urticaria pigmentosa and solitary mastocytomas are the uncommon cutaneous manifestations of a group of disorders known as the mastocytoses.^{1,2} In these conditions mast cell degranulation, which releases histamine, may result from a variety of factors, including drugs (morphine, d-tubocurarine, codeine, acetylsalicylic acid), stress, anxiety and temperature extremes.^{3,4} Reactions in cutaneous mastocytosis are characterized by symptoms of intense pruritus, generalized flushing, nausea, vomiting and diarrhoea. Syncope, hypotension and tachycardia have also been reported.¹

A number of studies discuss the risk, prevention and treatment of potentially life-threatening complications in patients with systemic or generalized mastocytosis during general anaesthesia.⁴⁻⁷ One case report has been published involving an uneventful anaesthetic in a child with

isolated urticaria pigmentosa, a four-year-old girl undergoing ligation of patent ductus arteriosus.⁸ However, the discussion of this case leaves the reader with the distinct impression that this group of patients should be considered at increased risk with any "invasive intervention."⁸

To obtain a clearer perspective of these issues, we retrospectively reviewed the hospital records of children with urticaria pigmentosa or solitary cutaneous mastocytoma who have undergone general anaesthesia at The Hospital for Sick Children, Toronto, over the past 25 years.

Methods

With the approval of the Committee for Human Research we reviewed the records of all patients with a diagnosis of cutaneous mastocytosis who presented at this centre between 1960 and 1985. Only the 42 patients with a diagnosis confirmed by biopsy or by a positive Darier's sign (urtication with light mechanical irritation of lesions) were included in the study.¹ The diagnosis of the subtypes of cutaneous mastocytosis was made by the attending dermatologist (B.R.K.). There were 29 cases of cutaneous pigmentosa, five of solitary mastocytoma, and four of diffuse cutaneous mastocytosis.

The anaesthetic records of the 15 patients who had received a general anaesthetic (mean age 4.9 years) were scrutinized for drugs administered and intra- and post-operative problems encountered. If necessary, clarification of details was sought from parents and attending physicians.

Results

The 15 children in our study group had undergone a total of 29 general anaesthetics: 12 patients with urticaria pigmentosa had 26, and the three children with a solitary cutaneous mastocytoma had one each. None of the four children with diffuse cutaneous mastocytosis, all of whom were receiving chronic drug therapy, had received a general anaesthetic.

Two of the patients with urticaria pigmentosa received preoperative prophylactic treatment with H₁- and H₂-blocking antihistamines on one occasion each. One of

Key words

ANAESTHESIA: general; ANAESTHESIA: paediatric;
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TABLE Details of perioperative drug administration in patients having cutaneous mastocytosis and undergoing general anaesthesia

	<i>Urticaria pigmentosa</i>		<i>Solitary cutaneous mastocytosis</i>	
	<i>No. of patients</i>	<i>No. of general anaesthetics</i>	<i>No. of patients</i>	<i>No. of general anaesthetics</i>
General anaesthesia	12	26	3	3
Chronic antihistamine administration	1	1	0	0
Preoperative antihistamine prophylaxis	2	2	0	0
Atropine	12	26	3	3
Succinylcholine	10	16	0	0
d-Tubocurarine	3	3	0	0
Morphine	2	2	0	0
Postoperative codeine	8	13	2	2

these patients was also receiving chronic antihistamine treatment for allergic rhinitis. None of the other patients having a general anaesthetic had received either preoperative prophylaxis or chronic antihistamine therapy. Details of all perioperative drugs administered are shown in the Table.

Only four perioperative complications were reported, all in children with urticaria pigmentosa. Two reactions associated with the administration of codeine were recognized: in one child a bullous eruption followed intramuscular administration in the postanesthetic recovery room, and in the other an urticarial eruption followed postoperative oral administration on the ward. No therapy was needed for either reaction. The other two problems occurred in the operating room and were nonspecific: laryngospasm following extubation, and a generalized flush following intravenous atropine administration. There were no reported side effects following the intravenous administration of morphine in two patients and d-tubocurarine in three patients. No problems were reported in either of the two patients who had received preoperative prophylaxis, either during the operation for which they were given prophylaxis or for the two procedures each underwent without prophylaxis.

Discussion

Patients with urticaria pigmentosa have been considered to be at increased risk of life-threatening cardiovascular collapse during general anaesthesia, due to histamine release.⁵ However, the only published case history of isolated urticaria pigmentosa does not appear to us to support this conclusion.⁸ In addition, in our series of patients with isolated cutaneous mastocytosis (i.e., urticaria pigmentosa or solitary mastocytoma) we found no evidence to suggest an increased risk of serious adverse reactions during general anaesthesia. There were no major complications, and the minor problems were self-limiting even though only two patients had received

any special precautions or prophylaxis, on one occasion each. Although several provocative drugs (e.g., codeine, morphine, d-tubocurarine) were administered (Table), only two minor reactions occurred (to codeine), neither requiring medical management.

Pretreatment of patients with systemic mastocytosis using combinations of prophylactic H₁, H₂ and prostaglandin blockers has been recommended.⁷ This is not without its own potential complications. Cimetidine has been associated with serious (if uncommon) side effects: cardiac rhythm abnormalities, inhibition of metabolism of certain drugs and decreased threshold for bronchospasm.⁹ Acetylsalicylic acid, although used by some workers in the perioperative drug therapy of patients with mastocytosis, actually liberates histamine.^{1,7,8}

Our data do not suggest that major adverse reactions are common in patients with isolated cutaneous mastocytosis undergoing general anaesthesia. In our opinion the indications for and benefits of the use of H₁, H₂ and prostaglandin blockers as prophylaxis have not been demonstrated and they may not be indicated. These drugs might even increase the risk to these patients. If prophylactic antihistamines were to be used, both H₁ and H₂ blockers would have to be given to fully block the systemic effects of histamine release.¹⁰

We believe that a conservative approach (monitoring closely, keeping the full range of therapeutic and resuscitative drugs readily available and avoiding known histamine-releasing drugs) may prove to be the optimal management for patients with isolated cutaneous mastocytosis undergoing general anaesthesia. A prospective multicentre study may be helpful in augmenting the available data for these rare conditions.

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Résumé

Malgré des informations et discussions considérables sur les risques de complications sérieuses lors de l'anesthésie des patients atteints de mastocytose systémique peu d'informations concernent les risques potentiels reliés à la mastocytose cutanée isolée. Notre expérience avec 29 anesthésies générales chez 12 enfants atteints d'urticaire pigmentée et trois atteints de mastocytose cutanée isolée est revue. Aucun de ces enfants n'était sur une thérapie chronique ou a reçu un traitement prophylactique pré-opératoire. Aucune complication majeure n'est survenue et les quatre problèmes minimes rencontrés se sont résolus par eux-mêmes. Les données de cette étude démontrent que les patients atteints d'urticaire pigmentée ou de mastocytose cutanée ne présentent pas un risque de complication grave lors de l'anesthésie.