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Successful perioperative care in systemic mastocytosis

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Dear Editor,

Systemic mastocytosis is a rare condition. The rate of immediate reactions related to anesthesia and surgery in patients with mastocytosis is only 0.4%.[1] The physiological stress of surgical procedures and the use of anesthetic and analgesic agents may be triggers of acute hypersensitivity reactions in this condition. Patients with mastocytosis require careful surgical management, especially when general anesthesia is contemplated.[2-5] We present a successful case in which intensive perioperative care was applied.

A 35-year-old Caucasian woman presented with a long history of indolent systemic mastocytosis and diffuse urticaria pigmentosa (Figure 1). She had elevated serum tryptase of 35.6 ng/mL (normal value: <11.4 ng/mL). Her blood KIT D816V mutation was negative. Past medical history included several episodes of perioral angioedema involving the lips and right nephrectomy for hypernephroma, which was considered cured with normal renal function. She had never experienced anaphylaxis. She had concomitant ulcerative colitis well controlled with once daily oral mesalamine (1200mg). About 5 years earlier, she had undergone an uneventful cesarean section with spinal anesthesia. Her mastocytosis was well controlled with daily oral medications: 20mg cetirizine, 300mg ranitidine, 10mg montelukast, 800mg cromolyn sodium, and 2mg ketotifen. She had also been prescribed 0.3mg epinephrine autoinjector (EpiPen), which she never used, and prednisone 20mg tablets as needed for angioedema. She also presented with rectus abdominis diastasis secondary to previous surgical procedures. This complication led to persistent abdominal wall pain that greatly disrupted her quality of life. The patient underwent a successful corrective plastic surgery with general

Figure 1
Urticaria pigmentosa in systemic mastocytosis

anesthesia using vecuronium and propofol as anesthetics and fentanyl and tramadol as analgesics. On the day prior to surgery, she was premedicated with oral 80mg prednisone, 30mg cetirizine, 300mg ranitidine, 4mg ketotifen, 1200mg cromolyn sodium, and 20mg montelukast. During surgery, she received 50mg promethazine, 150mg ranitidine, and 100mg hydrocortisone intravenously. Vital signs, electrocardiogram and oximetry signals were normal. The only adverse event occurred 3 hours after completion of surgery: a small bilateral periocular angioedema, without respiratory symptoms, that completely subsided in about 2 hours after administration of 40mg of prednisone orally.

Patients with systemic mastocytosis have shown an increased rate of reactions to general anesthetics and opioids. Elevated tryptase levels have been associated with an increased risk of systemic reactions. Several drugs, including neuromuscular blockers, can potentially activate mast cells through non-IgE MRGPRX2 receptors, thereby inducing mast cell-mediator release and possibly anaphylaxis.[6-7] Patients with systemic mastocytosis should not avoid undergoing required surgical procedures if proper allergy care is provided.

No conflicts of interest declared concerning the publication of this letter.

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Acesso facilitado à medicação com budesonida e formoterol associados

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Prezado Editor,

A prevalência da asma em adultos no Brasil foi recentemente estimada em 4,4% por meio da Pesquisa Nacional de Saúde, sendo que 43% das mulheres afetadas e 30% dos homens afetados tiveram pelo menos uma exacerbação, e 80% de todos afetados usaram medicação para asma nos últimos 12 meses[1]. Este estudo conclui que são necessárias políticas para obter um melhor controle da asma em nosso país.

As mudanças introduzidas pela Iniciativa Global Contra a Asma (GINA) em 2019, atualmente desaconselham o uso de beta-agonistas de ação curta isoladamente para o tratamento da asma por não tratar o processo inflamatório pulmonar e aumentar o risco de exacerbações de asma e visitas médicas de urgência. A recomendação para o tratamento da asma leve preconiza o uso de corticoesteroides inalados em associação com beta-agonistas de longa duração, sendo a associação preconizada, a de budesonida com formoterol para indivíduos acima de 12 anos de idade. Alternativamente, pode ser usado beta-agonista de curta duração, desde que usado de forma concomitante com corticosteroides inalados, ambos a serem usados conforme a necessidade percebida pelo paciente, sem um esquema de administração continuada.

Entre as vantagens da abordagem com a associação de corticoides com beta-agonista de longa duração por demanda na asma leve são a praticidade de manter somente uma medicação para uso conforme a necessidade do paciente, sem correr o risco de baixa adesão ao uso do corticoide inalado, o que levaria, novamente, ao uso de beta-agonista isolado.