

Systemic Mastocytosis: A review of gynaecological implications

Dr Yugisha Gurung, Dr Alexander Mortimer

Case Study:

A 20 year old woman presented with a rare case of vulval mastocytosis. Her symptoms posed the question of this being SM.

From a young age, patient suffered from Urticaria Pigmentosa, causing intense itching of armpits and neck.

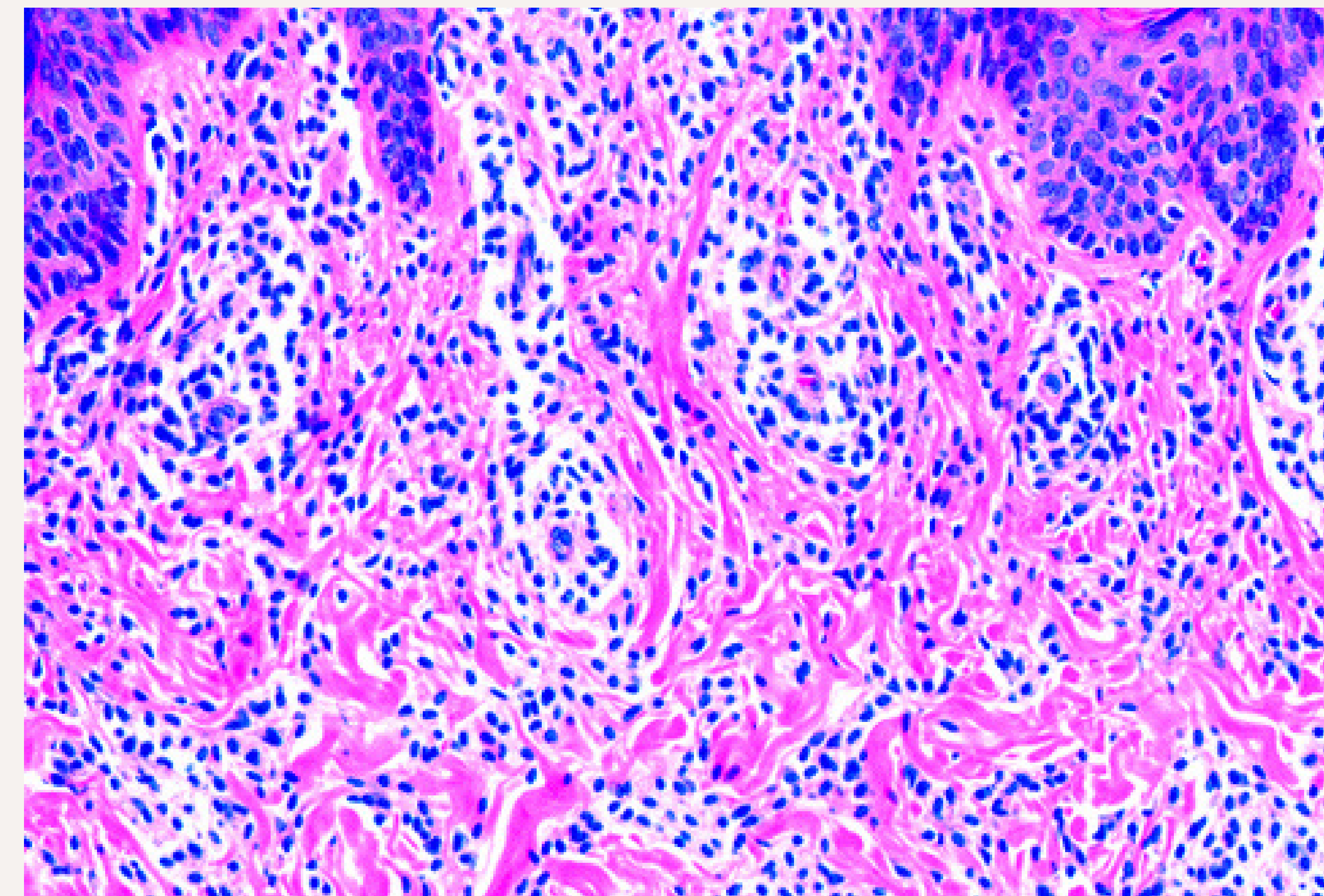
She also reported of abdominal pain and bloating, which initially was attributed to Irritable Bowel Syndrome, but in hindsight, these may have been due to mastocytosis.

Patient then developed vulval lesions, and when lesions were irritated, patient experienced flushing and pre-syncope. Biopsy of the vulval lesions showed mast cell infiltrates.

This unique case lead us to review existing literature, analysing the implications of mastocytosis on women's reproductive health.

Literature search was performed using keywords, including *mastocytosis, contraception, contraceptive, sexual health, antenatal, intrapartum, anaesthetics, and anaesthesia*.

We primarily focussed on publications released in the last 30 years.



Hale CS., Hamodat M. Cutaneous mastocytosis.
<https://www.pathologyoutlines.com/topic/skintumornonmelanocyticmastcell.html>.

What is Mastocytosis?

Mast cells are derived from myeloid stem cells and play an active role in immunity, tissue repair, and angiogenesis (1, 2). Mast cells are present in skin, lungs, GI tract, blood vessels, lymph nodes, and nerves. When activated, mast cells release pre-made and synthesised *de novo* pro-inflammatory mediators and vasodilators, including histamine, heparin, and prostaglandins (1, 3).

Mast cells are also found in the myometrium and placenta, and they have important roles during puberty, pregnancy, and maternal health. Mast cells have oestrogen and progesterone receptors, and during pregnancy, surge in these hormones result in proliferation and activation of mast cells (4).

Mast cells contribute and promote implantation of the blastocyst, and development of the placenta. They are also involved in myometrium contractions, and remodelling of uterine tissues after labour (3).

Mastocytosis is a heterogenous group of disorders that is defined by abnormal accumulation, proliferation, and activation of mast cells in response to stimuli. It has an estimated prevalence of 10-13 in 100,000 (5).

Mastocytosis can be divided into two main categories: cutaneous, and systemic. Cutaneous mastocytosis is confined to the dermis, whilst systemic mastocytosis is more aggressive. Whilst systemic mastocytosis can involve the skin, it mainly affects bone marrow, liver, lymph nodes (6).

Symptoms of cutaneous mastocytosis include flushing, urticaria, whereas systemic mastocytosis can result in more severe symptoms such as breathlessness, hypo/hypertension and even anaphylaxis (4, 7).

Discussion:

Multiple studies have shown impact of mastocytosis on patients' sexual health, with one quoting that 24% of patients with mastocytosis report of sexual impairment, including dyspareunia in women. These impairments cause patients to perceive themselves as disabled, affecting their ability to engage in sexual relationships. Sexual impairment was found to be more severe in systemic mastocytosis, with patients reporting their symptoms as intolerable/severe (8, 9).

During pregnancy and delivery, hormonal changes and stress of labour can cause irregular mast cell activity. Approximately, 20 - 33% of patients with mastocytosis experience exacerbation of their pre-gestational mastocytosis symptoms. Intensity of symptoms included pruritis, flushing, gastrointestinal symptoms, and fatigue. Some patients report of new symptoms which are not present in their pre-gestational phase (3, 10, 11).

However, some patients showcase an improvement in their symptoms during pregnancy, with some patient having complete resolution of their pre-gestational symptoms. Use of H1 and H2 antagonists, along with corticosteroid help with symptoms control (10).

More alarming and severe consequences of mastocytosis are vascular compromise and anaphylaxis (12). Patients with systemic mastocytosis are at an increased risk of anaphylaxis, compared to those with cutaneous mastocytosis. There are evidences of anaphylaxis in pregnant patient with mastocytosis (10, 13).

In the REMA study, 11 out of 138 pregnancies had anaphylaxis, with 7 of them being idiopathic. Idiopathic triggers included medications such as Diclofenac, Aspirin, Codeine, and antibiotics (10). Anaphylaxis may also be triggered by anaesthetics used in labour, including epidural. In population with no background of mastocytosis, incidence of anaphylaxis with peri-operative analgesia ranges from 0.009% to 0.01%, but in those with mastocytosis, incidence of anaphylaxis could be up to 0.3% (7).

Symptoms of anaphylaxis range from sudden hypotension, loss of consciousness, cardiorespiratory collapse, and coagulopathy (7). Increased activity of the coagulation pathway is observed in pregnant patients with mastocytosis, with no background of haematological abnormalities (11).

Our review of literature also shows mastocytosis associated preterm labour and delivery, with one extreme case of preterm delivery occurring at 27 weeks (11, 14).

Conclusion:

Mastocytosis can cause complications to patient's mental, emotional, sexual, and physical wellbeing. Symptoms should be thoroughly investigated and tightly managed when particularly involving antepartum, intrapartum, and postpartum health.

Whilst our patient was not pregnant at time of attendance to clinic, patient was prescribed H1, H2 antagonists, and Motelukast for symptom relief. More importantly, patient was given Epipen in case of any events of anaphylaxis, secondary to mastocytosis.

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