

# Images in Allergy and Immunology

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## Extracutaneous mastocytoma

**Editor's note:** This feature, Images in allergy and immunology, is designed to highlight current concepts of the immunopathology of allergic diseases and other common immunologically mediated diseases. The presentation will appear as sets of images that involve cross-pathology, histopathology, and molecular pathology and will cover a range of topics of interest to allergists and immunologists.

Mastocytomas in children are usually cutaneous lesions (urticaria pigmentosa) that consist of solitary brown-tan plaques or nodules larger than 1 cm, typically located on the skin of the extremities, face, scalp, and trunk but not of the palms or soles. Rubbing or stroking of the lesions induces urtication (Darier's sign). Association with systemic mastocytosis is rare, and serum tryptase levels are normal. They rarely bleed and typically resolve at puberty. Extracutaneous mastocytomas (Fig 1) are rare and of unknown prognosis. Perivascular accumulation of scroll-poor, tryptase- and chymase-positive mast cells with spindle shape or spherical forms are characteristic (Figs 2-6). The differential diagnosis includes malignant melanoma, histiocytoma, eosinophilic granuloma, Kaposi sarcoma, and basal cell carcinoma.<sup>1-3</sup> Specific stains for tryptase, chymase, and chloroacetate esterase can lead to the diagnosis because poorly granulated mast cells stained with Giemsa or toluidine blue stains can be mistaken for eosinophils or histiocytes.

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**FIG 1.** Computed tomographic scan of the skull of a 13-year-old boy who lost consciousness after being hit in the head by a soccer ball. He had a history of a waxing and waning tender nodule in his left occipital area for more than 1 year. A computed tomographic scan of the head revealed a well-circumscribed lytic lesion in the left occipital region with sclerotic margins (arrow). A 2.5 × 2.0 × 1.0-cm red-purple soft tissue mass eroding into the bone and firmly attached to the dura but sparing the brain was excised. Vigorous bleeding occurred during attempts to resect the entire mass, and a bone defect of 4 cm in diameter was closed by using a skin graft.

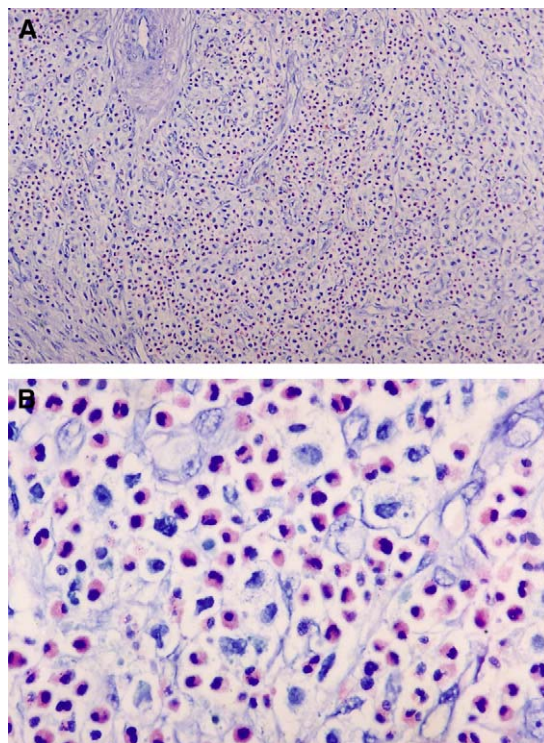


**FIG 2.** Diff-Quick staining of a section through the lesion under low (40×, A) and high (400×, B) magnification. The lesion consisted of a fibrous stroma containing cells of uniform size and pale pink-red cytoplasm. Although originally thought to represent eosinophils and a possible eosinophilic granuloma, the lack of nuclear segmentation suggested that this was not correct.

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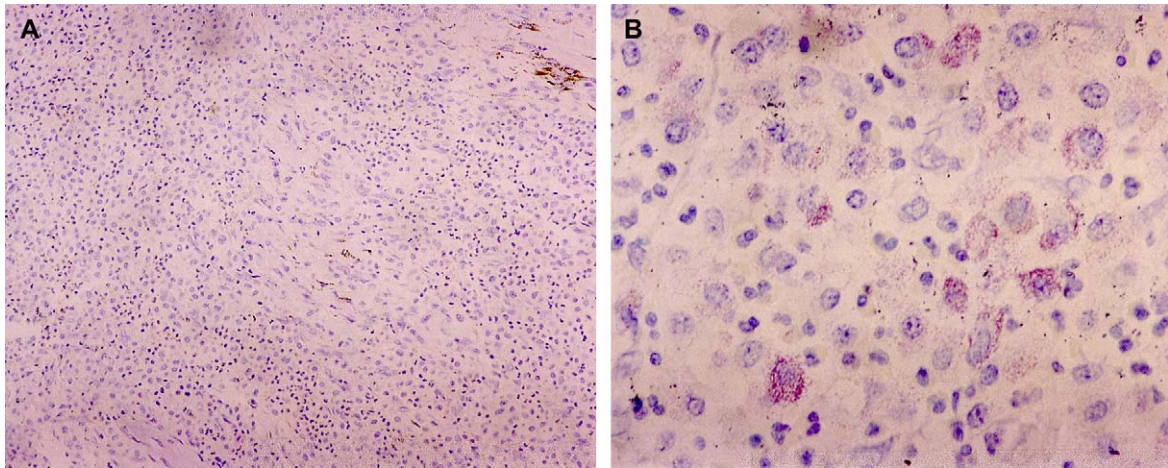


**FIG 3.** Toluidine blue staining of the formalin-fixed and decalcified specimen at low (40 $\times$ , **A**) and high (400 $\times$ , **B**) magnification. Many cells have no metachromatic granules, and some have few granules. Toluidine blue staining is not typical for eosinophilic granuloma. There is no evidence of mitotic figures.

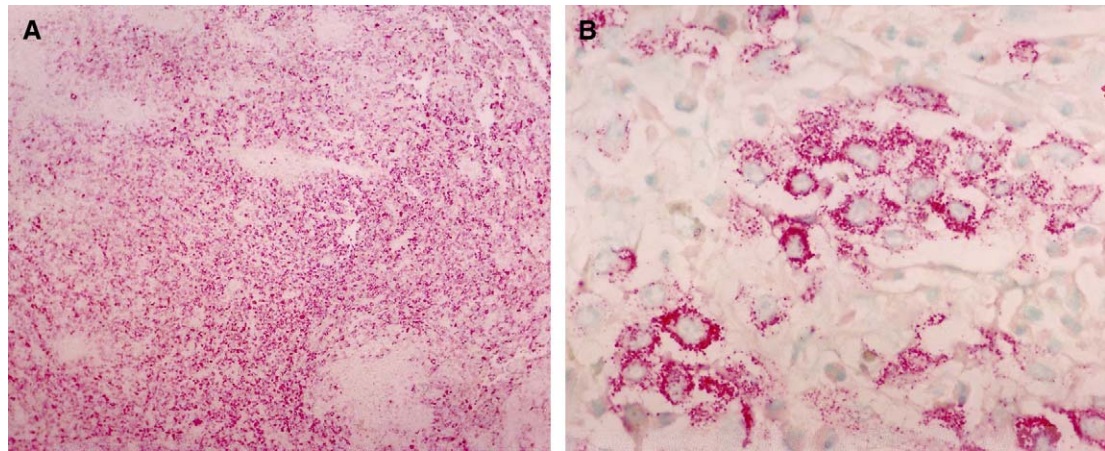


**FIG 4.** Chloroacetate esterase staining of the formalin-fixed and decalcified specimen at low (40 $\times$ , **A**) and high (400 $\times$ , **B**) magnification, showing staining of more than 80% of mononuclear cells with varying degrees of granularity. The discreet cytoplasmic granules and mononuclear morphology are both consistent with a mast cell origin.

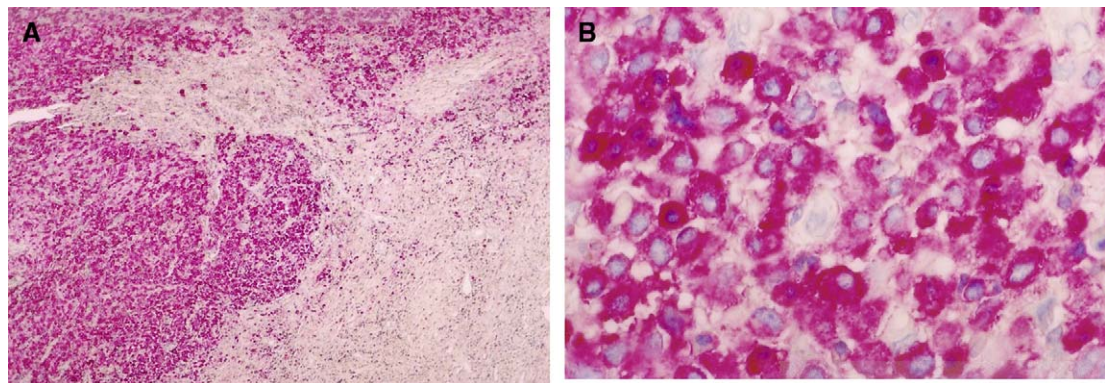


**FIG 5.** Tryptase staining of the formalin-fixed and decalcified specimen at low (40 $\times$ , **A**) and high (400 $\times$ , **B**) magnification. Uniform intense staining of the cell population at the borders of the mass resection (**A**), indicating the presence of tryptase and confirming the mast cell origin, is shown. The majority of the cells show intense staining of the cytoplasmic granules with a mononuclear morphology (**B**). Stained cells have varying degrees of granularity and aggregate in clusters and sheets.

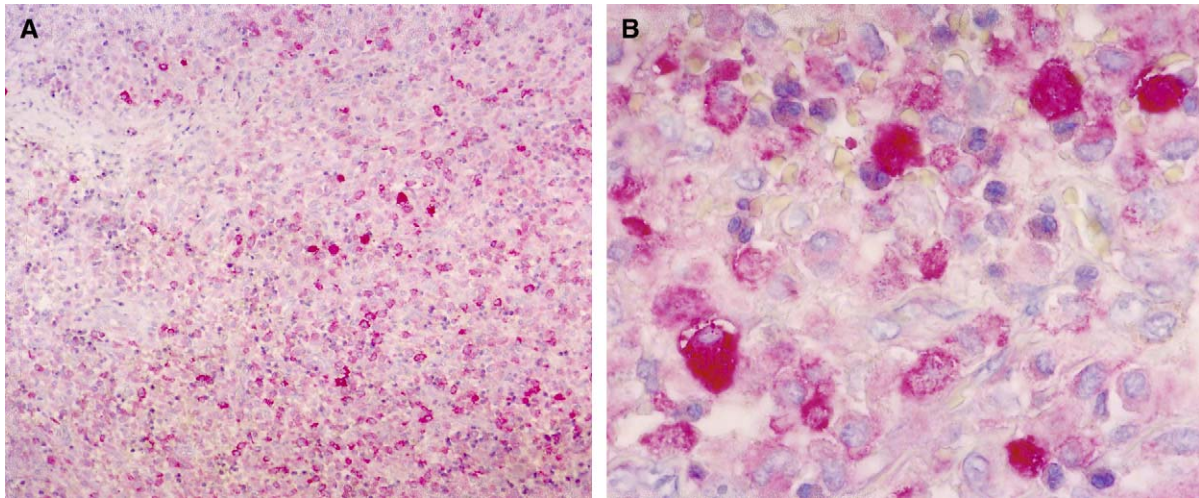


**FIG 6.** Chymase staining of the formalin-fixed and decalcified specimen at low (40 $\times$ , **A**) and high (400 $\times$ , **B**) magnification. The lesion contains fewer chymase-positive cells than tryptase-positive cells, indicating that it consists of a mixture of tryptase-positive/chymase-positive and tryptase-positive/chymase-negative mast cells, as frequently seen in cutaneous mastocytosis and mastocytomas.

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