

A girl with a lid mass

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Question

A 9-year-old girl presented with a painless, slow growing left upper lid mass for 9 months. There was no history of trauma. Her past medical health was unremarkable. Examination disclosed a 3- x 2-mm firm non-tender orange-red mass with a smooth surface (**Figure 1**). Excisional biopsy was performed and the mass was submitted for histopathological evaluation (**Figures 2 and 3**).

1. What does the histopathology show?
2. What is the diagnosis?

(Answers and discussion on page 38)



Figure 1. Non-tender smooth mass.

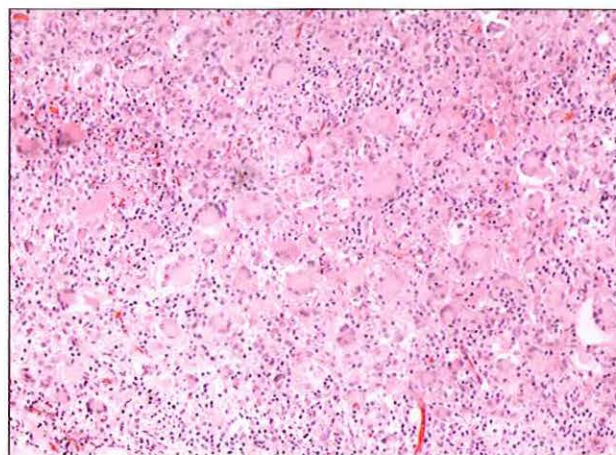


Figure 2. Histopathology of smooth mass. (hematoxylin and eosin; original magnification, x 20).

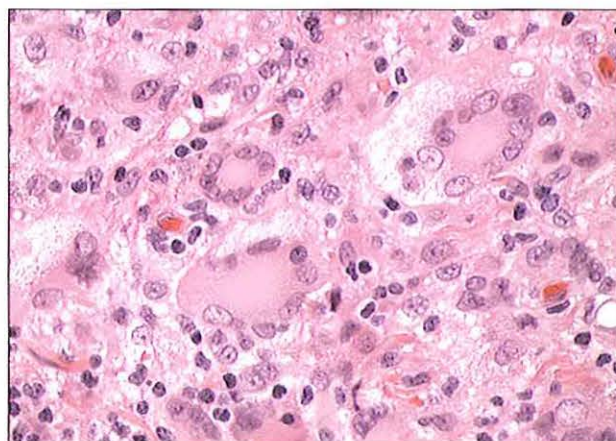


Figure 3. Histopathology of smooth mass. (hematoxylin and eosin; original magnification, x 40).



Answer

1. An extensive infiltration of histiocytes (large macrophages with abundant cytoplasm and vacuoles), Touton giant cells (multinucleated giant cells with a peripheral ring of nuclei surrounding the central foamy cytoplasm), and lymphocytes.
2. Juvenile xanthogranuloma.

(Questions on page 22)

Discussion

Juvenile xanthogranuloma (JXG) is a benign self-limiting cutaneous disorder that mostly occurs in infancy and early childhood.¹ The cutaneous lesions usually present as firm orange-tan papulonodules and are commonly found over the face, neck, and upper trunk.² Histopathology shows a mixture of foamy epithelioid histiocytes, with some Touton multinucleated giant cells and lymphocytes in a prominent vascular network.^{2,3} The exact etiology of JXG is still unknown. It has been postulated that JXG represents an inflammatory response to an unidentified stimulus resulting in accumulation of activated macrophages.⁴ JXG can be associated with neurofibromatosis type 1 and Niemann-Pick disease.^{5,6}

References

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JXG can, rarely, have extracutaneous involvement. The most common extracutaneous lesion can be found in the iris.³ Iris JXG may present as congenital or acquired heterochromia iridis, uveitis, or spontaneous hyphema, which may result in secondary glaucoma, corneal staining, amblyopia, or even blindness.^{7,8} Other ocular sites of involvement include the conjunctiva, cornea, episclera, retina, choroid, optic nerve, and orbit.⁹⁻¹¹ Extracutaneous lesions may rarely be found in the lungs, pericardium, testes, bones, and central nervous system.^{4,12}

Although cutaneous JXG usually involutes spontaneously over 1 to 5 years, excisional biopsy should be considered for definitive diagnosis or cosmesis. However, spontaneous regression is less frequent for ocular lesions. Since iris lesions may lead to serious ophthalmic sequelae, patients with cutaneous JXG should be screened for any ocular involvement, particularly iris lesions. Iris lesions or other intraocular lesions are initially treated with topical steroids. Systemic steroids should be considered for severe cases. Low-dose radiotherapy may be a treatment option for diffuse uveal involvement, or cases that are refractory to steroid treatment. Intralesional steroid injection has been used for orbital lesions. Rare cases of severe systemic JXG may require combination chemotherapy.¹³