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Inflamed Non-Limbal Scleral Dermoid Masquerading as Nodular Scleritis

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Abstract

Background: Dermoid cysts are one of the most common orbital lesions in childhood. They typically present as a palpable subcutaneous mass in the superotemporal region of the orbit along the frontozygomatic suture. When involving the eye, ocular dermoids typically present as visible lesions at the limbus (the border of the cornea). There are no documented cases of non-limbal ocular dermoids in literature.

Clinical Case Report: A five-year-old male presented with focal injection of the left nasal conjunctiva sparing the limbus that progressed to a non-mobile scleral nodule with overlying 3+ injection, tenderness to palpation, and minimal blanching with phenylephrine suspicious for nodular scleritis versus abscess. The inflammation improved, but the nodule persisted despite treatment with topical antibiotics, steroids, and oral NSAIDs. Superonasal subconjunctival excisional biopsy revealed an 8mm mass with a visible hair protruding from lesion, with pathological analysis revealing findings of acutely inflamed dermoid cyst.

Discussion: This is a unique case of ocular dermoid. Typical ocular dermoids present as painless bumps on the corneal limbus or lesions of the orbit that may cause inflammation if ruptured in the setting of trauma. In this case, the inflammation may have occurred secondary to leakage of oil or keratin from the cyst, irritation from the protruding hair, or reaching a threshold size to incite disruption of surrounding tissue.

Conclusion: To our knowledge, this is the first documented case of a non-limbal ocular dermoid masquerading as nodular scleritis.

Background

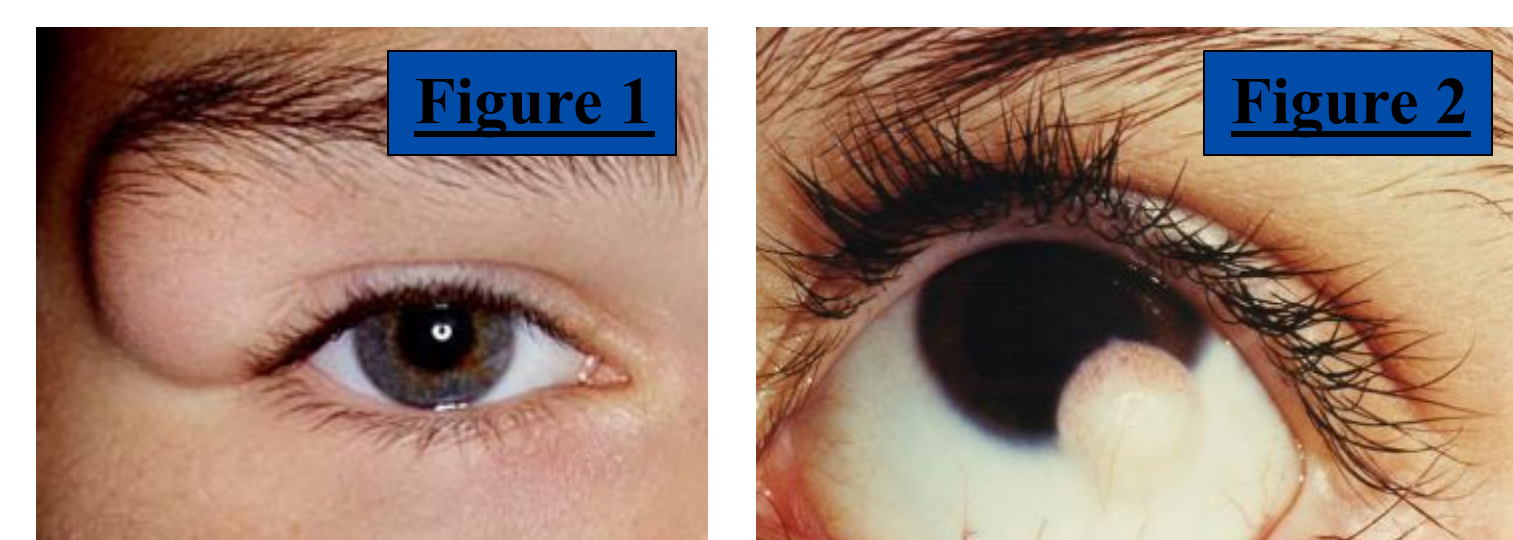


Figure 1: Typical appearance of orbital dermoid
Figure 2: Typical appearance of limbal dermoid

- Dermoid cysts are congenital choristomas consisting of mature tissues including skin, hair follicles, sebaceous glands
- Dermoid cysts are the most common cystic orbital lesion in childhood, and comprise 40% of all orbital lesions in childhood¹
- They most commonly present as a painless palpable mass in the superotemporal region of the orbit (**Figure 1**) from aberrant ectodermal tissue sequestered in the frontozygomatic suture during closure in embryologic development²
- Ocular dermoids typically present at the limbus as epibulbar cysts (**Figure 2**)

Report of a Case

- A five-year-old male with a past medical history of an episode of anemia, thrombocytopenia, and neutropenia of unknown etiology prior to age one initially presented with redness in the left eye. Visual acuity was 20/30 right eye, 20/40 left eye. Examination revealed focal injection of the left nasal conjunctiva suspicious for episcleritis (inflammation of the episclera) versus phlyctenule (nodular inflammation that results from a hypersensitivity reaction to a foreign antigen) and the patient was started on prednisolone acetate QID in the left eye
- Examination three days later revealed stable visual acuity and progression of the lesion on slit lamp examination with the development of a non-mobile scleral nodule with overlying 3+ injection, tenderness to palpation, and minimal blanching with phenylephrine suspicious for nodular scleritis versus abscess (**Figure 4**)

- There was no intralesional fluid visualized on anterior segment ocular coherence tomography, a non-invasive imaging technique that employs near-infrared light (**Figure 3**), making abscess less likely
- There was no posterior extension on dilated exam and B-scan ultrasound, making malignancy less likely
- Evaluation of basic, inflammatory, and autoimmune labs was unremarkable

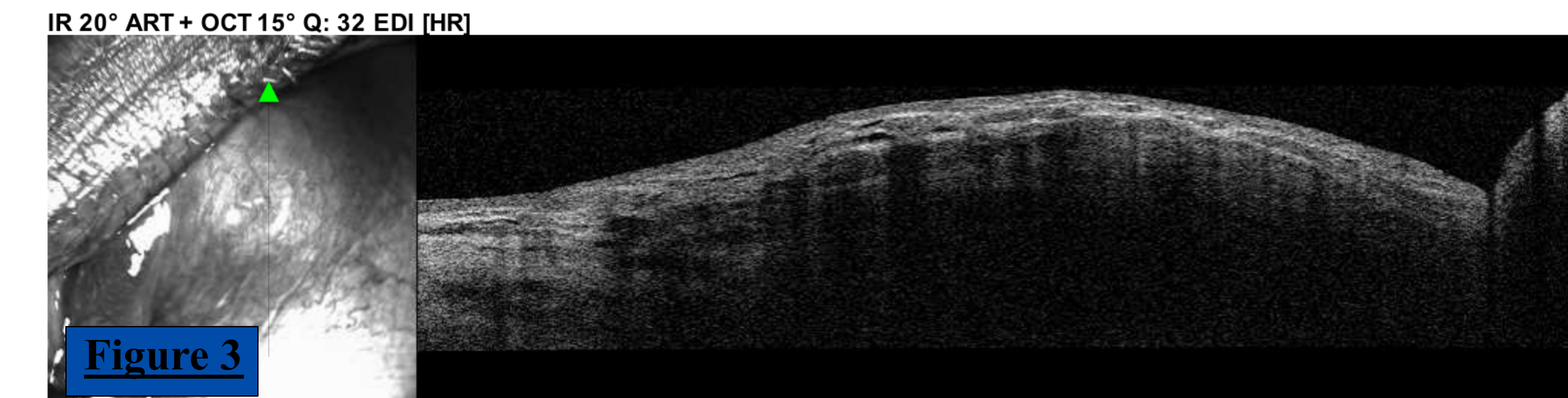


Figure 3: No intra-lesional fluid visualized on OCT

- Two weeks later, there was moderate improvement in injection with use of prednisolone acetate QID, moxifloxacin TID, and ibuprofen 10mg/kg TID but persistent nodular lesion (**Figure 5**)



Figure 4: Pre-treatment: scleral nodule with 3+ overlying injection, 4 days after onset

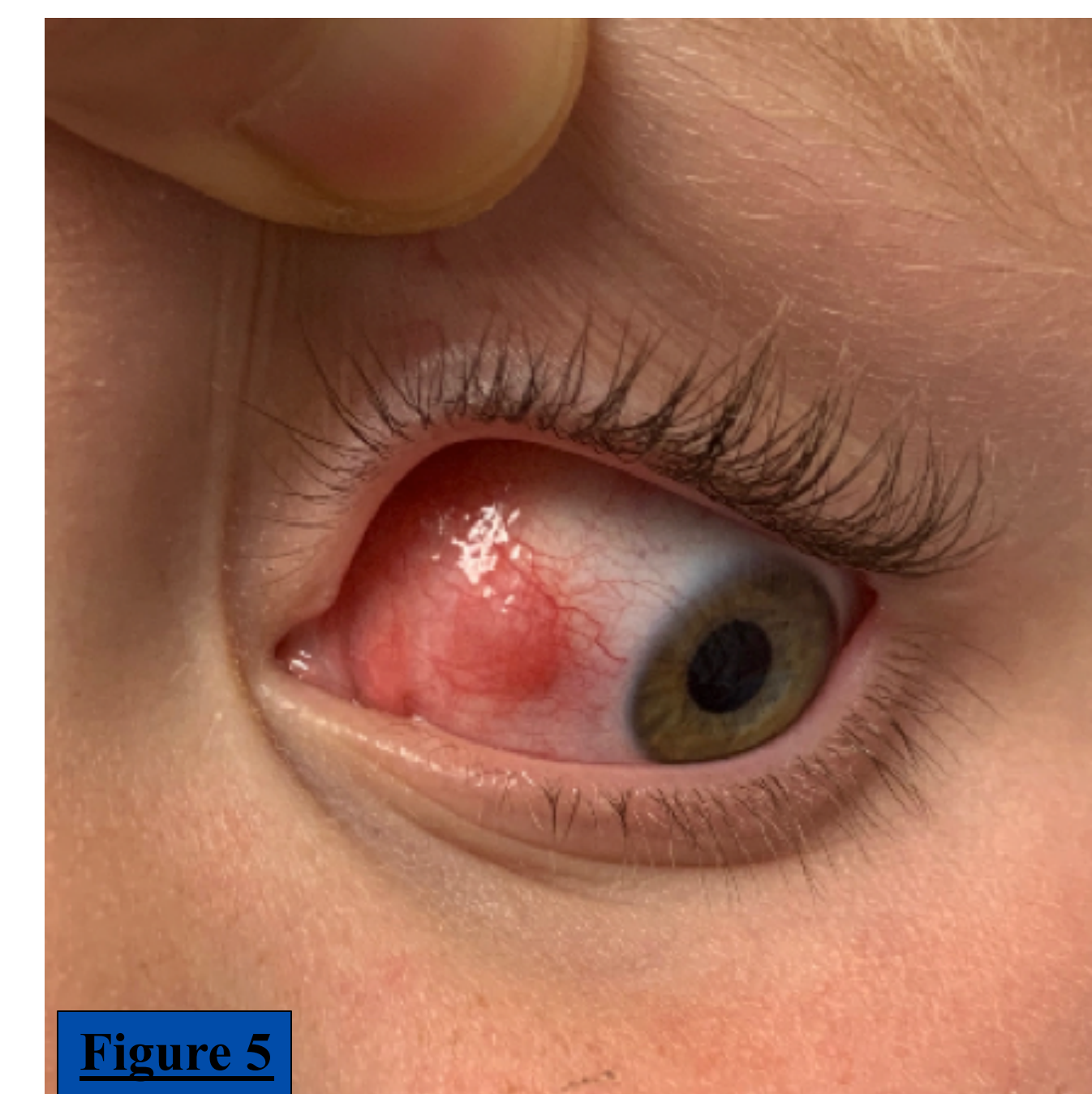


Figure 5: Post-treatment: persistent scleral nodule with improved 1+ overlying injection, 17 days after onset

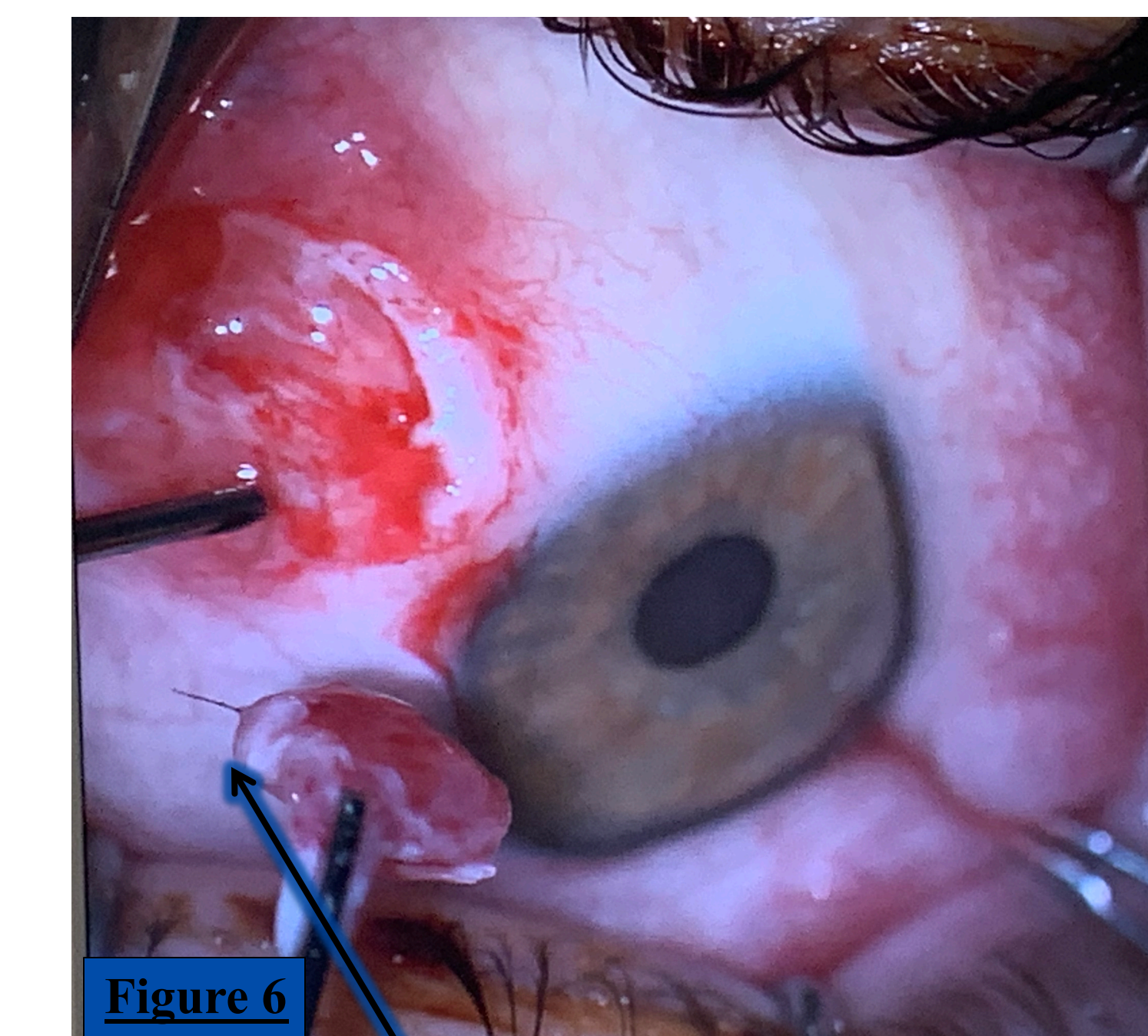


Figure 6: Intraoperative photo demonstrating 8mm excised subconjunctival lesion with visible hair protruding from lesion

- Superonasal subconjunctival excisional biopsy approximately 2 weeks after initial presentation revealed an 8mm mass with a visible hair protruding from lesion (**Figure 6**)
- Pathological analysis revealed hair shafts with focal keratin debris, granulation tissue, and extensive acute and chronic inflammation consistent with acutely inflamed dermoid cyst (**Figures 7-9**)
- The patient was healing well without complication at 1-month follow up visit



Figure 7: Lesion lined with keratinized stratified squamous epithelium

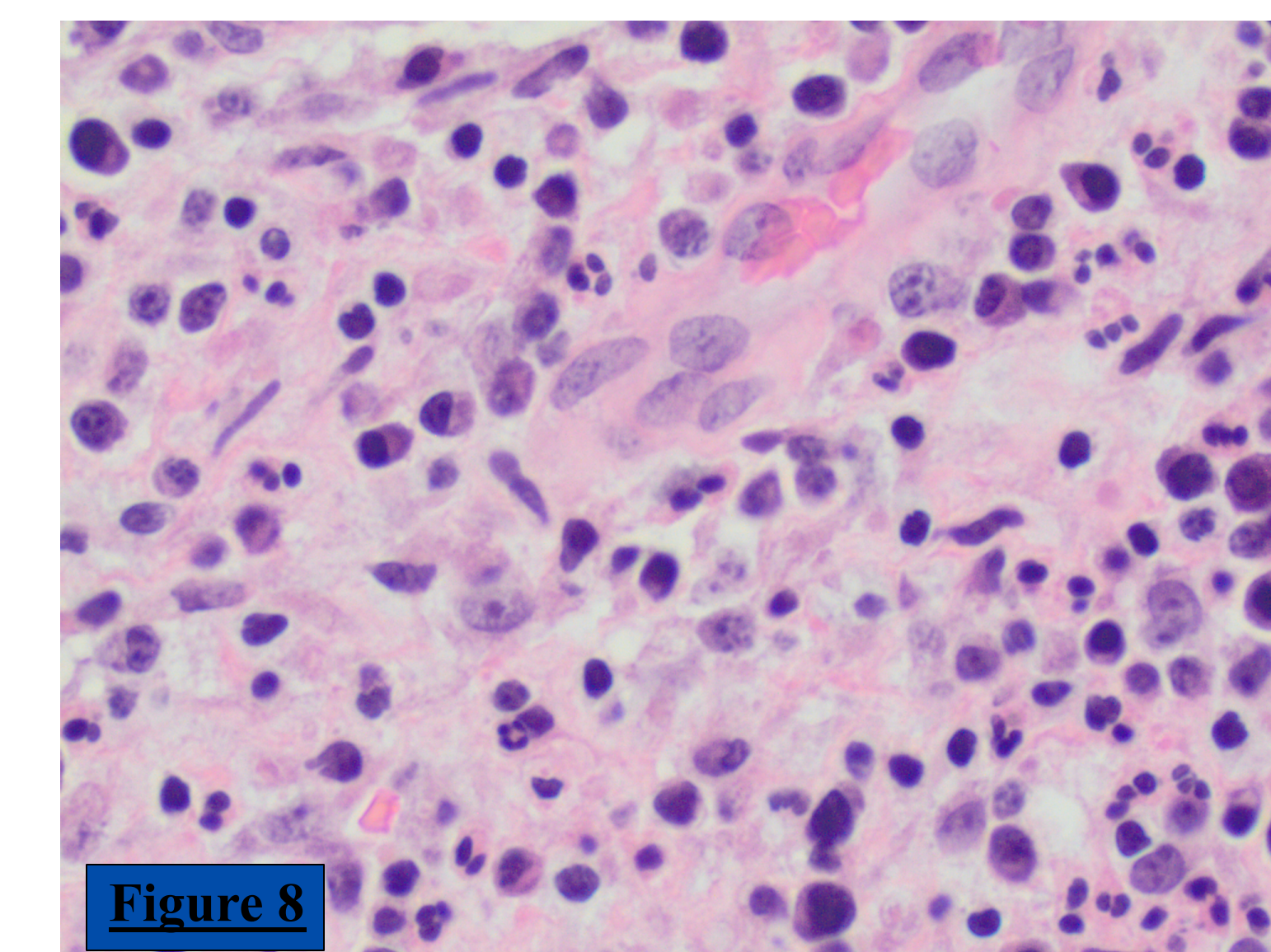


Figure 8: Evidence of acute and chronic inflammation with lymphocytic infiltration, rare neutrophils, and fibroblasts

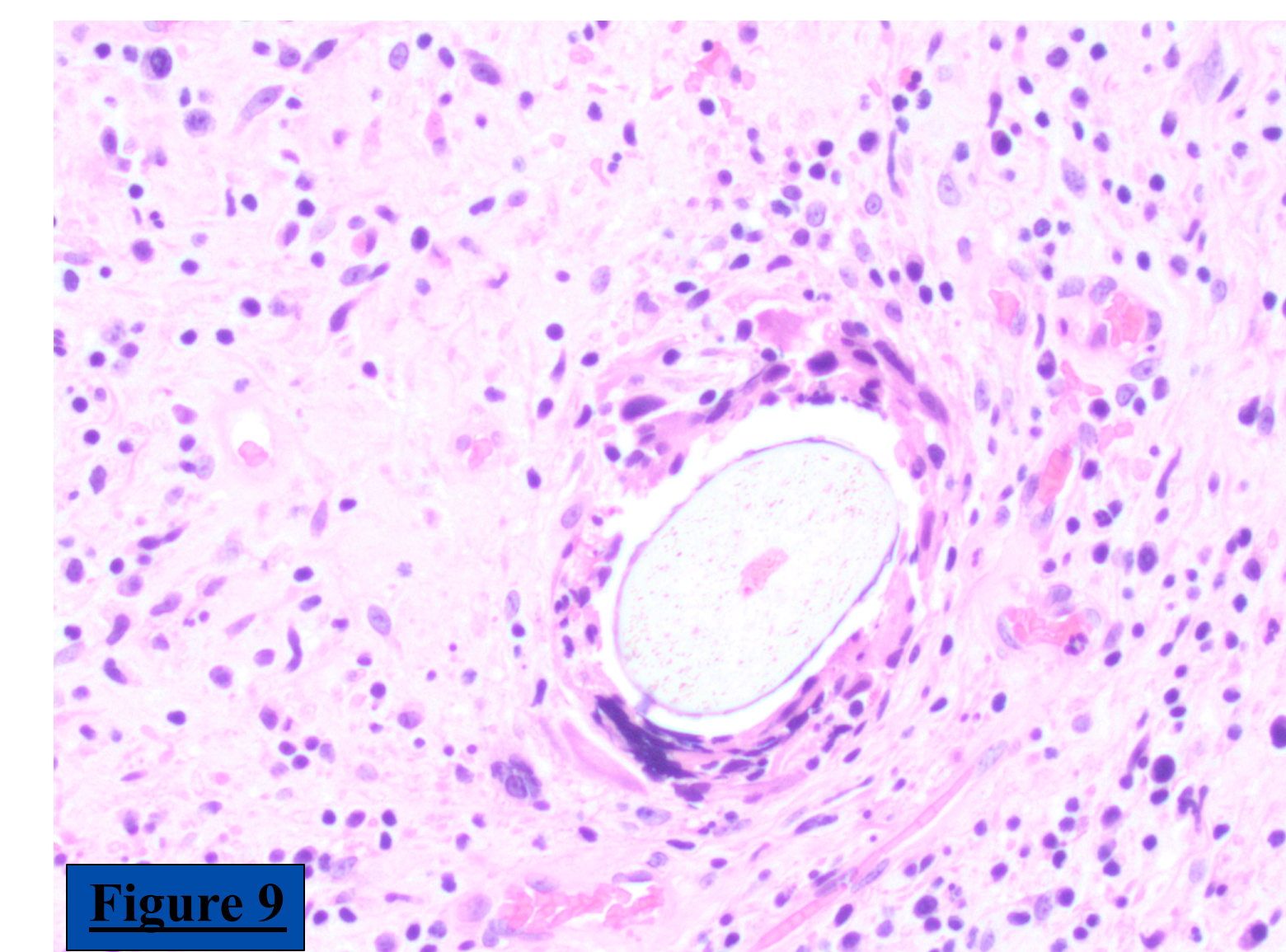


Figure 9: Hair shaft surrounded by inflammatory cells

Discussion

This is a unique case of an ocular dermoid cyst masquerading as nodular scleritis and located in the subconjunctiva, sparing the limbus. Most cases of dermoid cysts present as painless palpable masses in suture lines, and may rarely present with inflammation. In prior cases, inflammation can occur after frank rupture of the cyst due to trauma or inadvertently during surgical excision, however this patient had no prior history of trauma or ocular surgery. The inflammation may have occurred secondary to leakage of oil or keratin from the cyst, irritation from the protruding hair, or reaching a threshold size to incite disruption of surrounding tissue.

Conclusions

To our knowledge, this is the first documented case of a non-limbal ocular dermoid masquerading as nodular scleritis according to a pubmed search of the terms “Ophthalmology dermoid,” “Ophthalmology dermoid rare,” “Ophthalmology dermoid unusual,” “Ophthalmology dermoid masquerade.”

References

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