

Role of intralesional bleomycin sclerotherapy as the sole or adjunct treatment of superficial ocular adnexal lymphatic malformations

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Abstract

Objective To report the role of intralesional bleomycin as sole or adjunct therapy in the management of superficial ocular adnexal lymphatic malformations.

Design Retrospective study.

Participants Three patients/three eyes.

Methods Retrospective chart analysis of patients receiving intralesional bleomycin sclerotherapy as sole or adjunctive treatment for superficial ocular adnexal lymphatic malformations at a single tertiary level eye care institution.

Results Degree of clinical regression of the lesions (graded by percentage) and resolution of symptoms and signs associated with the lesions.

Conclusions Use of intralesional bleomycin sclerotherapy appears to be an effective sole or adjunct treatment in the management of superficial ocular adnexal lymphatic malformations.

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Design and methods

The study was a retrospective chart analysis and adhered to the tenets of Helsinki. Informed consent was obtained in all cases. Three cases were included. Contrast CT imaging was done to delineate extension of the lesions and a mixed macro/micro cystic pattern was noted. Histopathological tissue diagnosis was confirmed for all cases. All received two or more injections of intralesional bleomycin depending upon assessment of clinical response. One case underwent surgical debulking of the conjunctival component of the LM in addition to intralesional bleomycin injection. Bleomycin (1 mg/ml) mixed with 2% lignocaine in a ratio of 4:1 was used.⁵ Percutaneous injection was performed through a 26G needle under direct visualization using bony anatomical landmarks as guidance. Total dose ranged between 2–4 mg per treatment sitting. Repeat injections were given 4 weeks or more apart depending on the clinical response. The total cumulative dose did not exceed 10 mg for any patient. This was significantly less than the reported cumulative systemic dose (450 mg) necessary for a risk of pulmonary fibrosis.⁶ No other reported adverse effects of bleomycin were seen.⁶ Clinical response was graded as very good (>80% resolution), good (60–80% resolution), moderate (40–60% resolution), and poor (<40% resolution).

Brief report of cases

Case 1

A 2-year-old-male child presented with active bleeding from the left eye for 2 weeks. A glabellar and left upper eyelid swelling with

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Introduction

Lymphatic malformations (LMs) are benign, hamartomatous lesions that often involve the ocular adnexae.¹ Multiple treatment modalities are described for these lesions.^{2,3} Use of various intralesional sclerosing agents have been reported.⁴ We report the role of bleomycin sclerotherapy as the sole or adjunctive treatment in three cases of superficial ocular adnexal LMs.

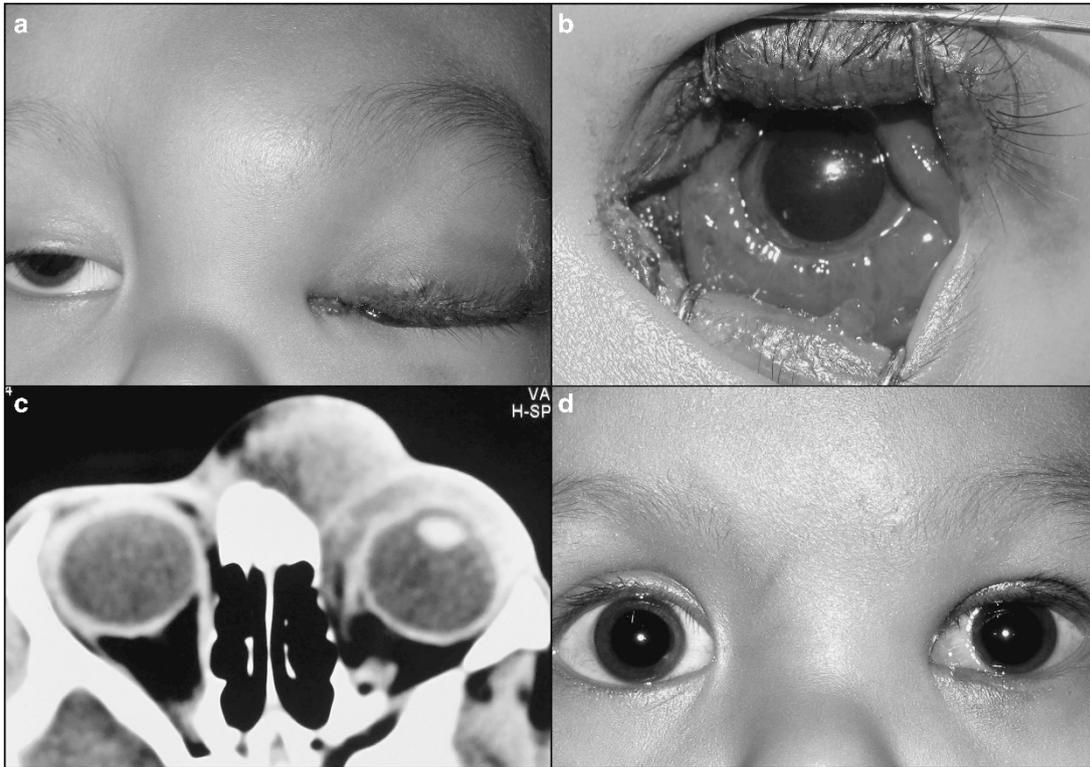


Figure 1 Case 1 at presentation (a) with gross upper lid swelling and a large glabellar swelling. Diffuse upper lid margin involvement with extensive conjunctival involvement and hemorrhage seen (b). CT scan shows extent of involvement (c). Appearance at last follow-up shows an excellent response (d).

gross chemosis and subconjunctival hemorrhage was seen (Figures 1a and b). CT showed an enhancing heterogeneous soft tissue mass (Figure 1c). He received three injections of bleomycin (cumulative dose 10 mg). At 11 months of follow-up, very good regression (>80%) without any recurrence was evident (Figure 1d).

Case 2

A 28-year-old female presented with active bleeding from the right eye of about one month duration. The eyelids were grossly swollen with presence of a multilobulated conjunctival mass (present since birth) (Figures 2a and b). CT showed an enhancing soft tissue mass (Figure 2c). She received two injections of bleomycin (cumulative dose 10 mg). The conjunctival component was surgically debulked along with cauterization of the base of the lesion in the lower conjunctival fornix. No grafts were used for ocular surface reconstruction. This was done simultaneously with the first sitting of intralesional bleomycin. Clinical regression was very good (>80%) without recurrence at 18 months of follow-up (Figure 2d).

Case 3

An 18-year-old female presented with recurrent episodes of swelling and active bleeding from her left eye over several days. Left eyelid and periocular swelling was seen with diffuse chemosis and subconjunctival hemorrhage (Figures 3a and b). CT showed a diffuse enhancing soft tissue mass involving the preseptal region and the medial anterior orbit (Figure 3c). She received two injections of bleomycin (cumulative dose 8 mg). At 13 months of follow-up, clinical regression was judged to be moderate (40–60%) (Figure 3d).

Discussion

Ocular adnexal LMs pose a therapeutic challenge due to their infiltrating and arborizing nature.^{2,3} Surgical management in the ocular adnexal region is fraught with danger of collateral damage to functionally critical tissues. Intralesional sclerotherapy offers a potentially safer alternative for treatment.⁵ Sclerotherapy for orbital lymphangiomas with multiple agents has been reported in ophthalmic literature^{2,7–9} but reports on the use of bleomycin are comparatively sparse.^{5,10}

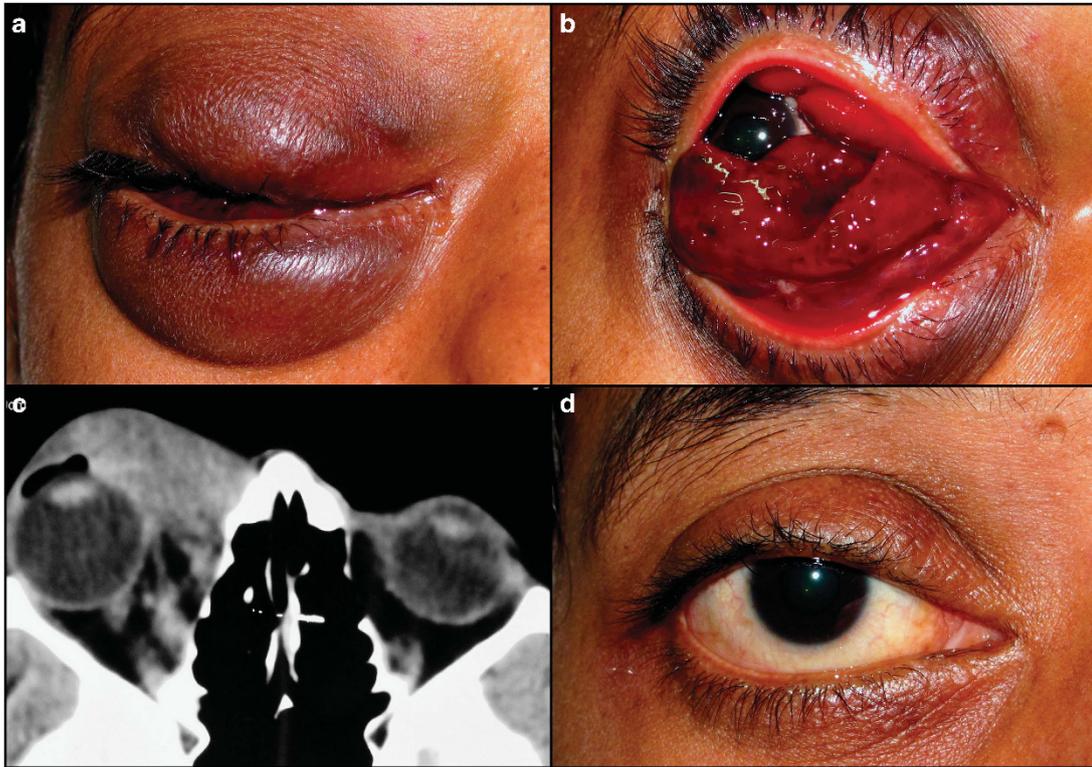


Figure 2 Case 2 at presentation shows gross eyelid swelling with ecchymosis (a) and a congested, bleeding conjunctival component (b). CT scan shows extent of involvement (c) and an excellent response is seen at last follow-up (d).

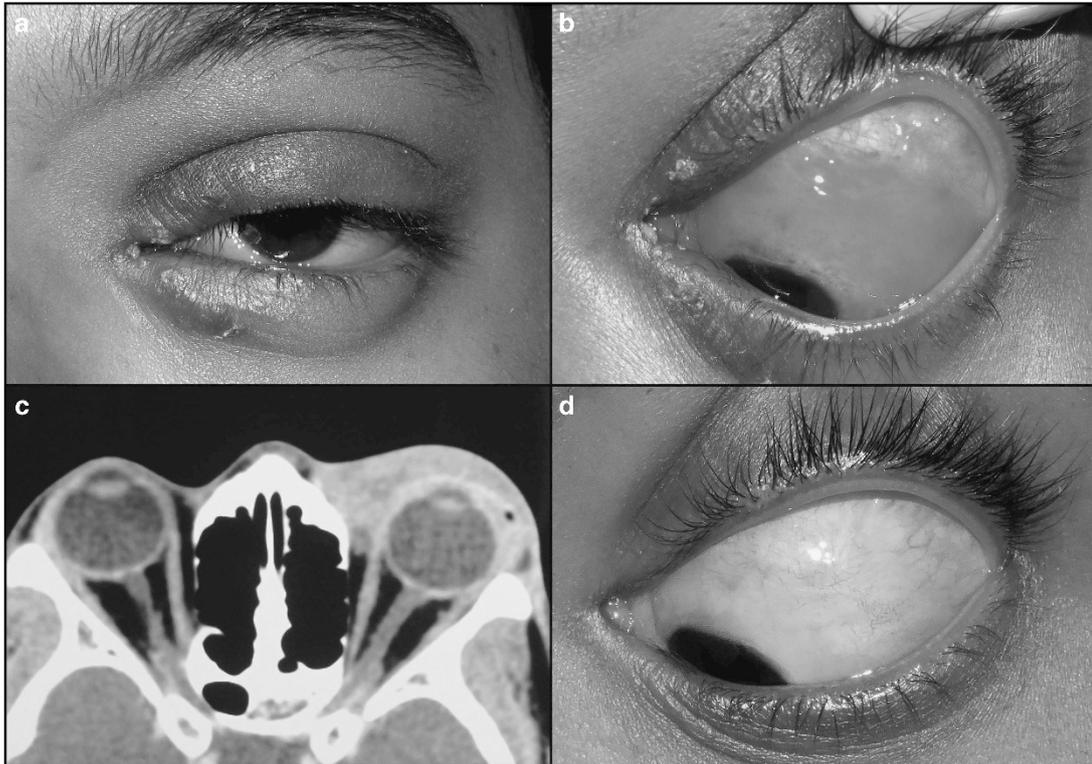


Figure 3 Case 3 at presentation shows eyelid swelling and ecchymosis most marked medially (a) with extensive conjunctival involvement (b). CT scan shows extent of lesion (c). At last follow-up, moderately good response to treatment is evident with regression of the conjunctival component as well as a noticeable decrease of the medial lid swelling (d).

Bleomycin causes endothelial cell detachment with inflammation resulting in fibrosis and involution.⁴ The authors chose bleomycin due to its easy availability, suitable adverse effect profile at the doses used for sclerotherapy and reported better efficacy.¹¹ The orbital and eyelid components appeared to respond better to sclerotherapy than the conjunctival component. Subconjunctival injection of bleomycin was not attempted for these cases. Although the use of bleomycin has been reported for subconjunctival venous malformations,¹² the authors are not aware of any report of subconjunctival use for LMs as of now.

The nature of these cases is slightly different than the classical deep orbital lymphangiomas for which bleomycin and pingyangmycin sclerotherapy have been reported.^{5,10} These are a distinct subset that primarily had an infiltrating eyelid and conjunctival component with only an anterior orbital component. The anterior location made these amenable to direct percutaneous sclerosant injection without the use of imaging guidance. The conjunctival component needed additional surgical debulking in one case. Bleomycin acted as an effective sole agent or adjunct for management of these cases.

Intralesional bleomycin sclerotherapy appears to be a very useful sole or adjunct treatment of superficial ocular adnexal LMs. Therapy can be safely performed without the aid of imaging guidance in selected cases. Good outcomes with minimal side effects or long-term morbidity are achievable.

Summary

What was known before

- Bleomycin is an effective intralesional sclerosant for lymphatic malformations
- Bleomycin has a good safety profile and minimal adverse effects when used as a sclerosant

What this study adds

- Bleomycin is a very useful sole or adjunct treatment in superficial ocular adnexal lymphatic malformations
- Imaging guidance may not be essential for intralesional injection in superficial ocular adnexal lesions

Conflict of interest

The authors declare no conflict of interest.

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