

Surgical Intervention in Upper Lid Haemangioma To Prevent Amblyopia: Two Cases Report

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Background : Non-operative treatment for Hemangioma involving deeper skin structures extending into the mucosa is usually unsuccessful, since usually no involution will develop. Early surgical planning is important to prevent amblyopia and for the child's self confidence of their cosmetic appearance later in life.

Methods : We present two cases of superior palpebra hemangioma who underwent early surgical treatment.

Discussion : The first case involves a large hemangioma of the left superior palpebra with severe vision restriction; although later examination was proven that the visual function was still preserved. The second case involves a smaller periocular lesion. Surgery was performed on both cases. In the larger hemangioma, only partial of tumor was removed by incision at the lid crease and intra-lesion injection of corticosteroids was administered to the remaining tumor.

Conclusion: Advantages of surgical procedure in early rapidly growing stage and in very large hemangioma of upper eyelid; firstly, provides a definitive early treatment; secondly, safe because bleeding was manageable, and third prevent occlusion-related amblyopia. Besides these benefits surgery promises added esthetic outcome.

Keywords: Hemangioma of the eyelids, surgical excision

Latar Belakang: Terapi konservatif untuk kasus hemangioma yang melibatkan struktur kulit yang lebih dalam dan meluas ke mukosa umumnya kurang berhasil karena involusi tidak terjadi. Perencanaan untuk melakukan terapi pembedahan awal untuk mencegah ambliopia dan untuk meningkatkan kepercayaan diri penderita mengenai penampilan mereka untuk jangka panjang.

Methods : Dalam jurnal ini akan dibahas dua kasus hemangioma pada palpebra superior. Yang menjalani terapi pembedahan awal.

Diskusi : Pada kasus pertama didapatkan hemangioma ukuran besar yang melibatkan palpebra superior dextra dengan restriksi penglihatan. Dalam pemeriksaan lanjutan didapatkan bahwa fungsi penglihatan pasien masih dapat dipertahankan. Terapi pembedahan dilakukan pada kedua kasus. Pada kasus hemangioma dengan ukuran yang lebih besar dilakukan eksisi parsial dari tumor pada lipatan palpebra dan injeksi kortikosteroid intralesi pada sisanya.

Ringkasan : Keuntungan dari terapi pembedahan saat fase pertumbuhan pesat tumor dan pada hemangioma di palpebra superior, adalah memberikan terapi definitif lebih awal, lalu lebih aman karena perdarahan dapat lebih dikontrol, dan mencegah terjadinya ambliopia akibat oklusi. Selain keuntungan tersebut, tindakan pembedahan juga memberikan hasil estetik yang lebih baik.

Kata kunci: Hemangioma of the eyelids, surgical excision

Hemangiomas are benign tumors consisting of masses of capillaries vessels made up by abnormal growth of vascular endothelial cells. ^{1,2} Hemangioma is caused by an increase in angiogenesis factors accompanied by a decrease in cell suppressing factors. ¹ The lesion has a low rate of blood flow and is non-pulsatile. Clinical manifestation is

described as a growing red papule that was not present at birth but becomes larger and thicker over the first few months of life. ^{2,3} The tumor occurred during infancy and childhood, affecting approximately 10% of infants by the age of 1 year. ³ With improper management, the tumor may cause amblyopia and therefore affects the child's vision in the future.

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Facial hemangiomas tend to be solitary lesions and occur spontaneously, with a female predilection of nearly 5 to 1.2. In 80% cases occur as a single lesion.¹

Over the past few decades, the natural course of hemangioma and the appropriate therapy were still debatable. Mulliken and Glowacky (1982) describe there is a rapid progression of the tumor followed by involution phase which will regress completely by the age of 4-8 years old, and they distinctively define the difference between hemangioma and malformations of vascular in character and clinical appearance.^{3,4} According to (Keihn et al), facial hemangioma aggressive therapy is crucial, particularly if it is life threatening, such as invasion to the eyes and nose cavity.⁵

Surgical therapy is an option when vision threatening complications such as amblyopia is present. Characteristics of hemangioma near the mucosa were proven to be rapidly proliferative for 12-14 months even up to 2 years, and involution rarely occurred.¹

It has been reported by Jenny L et. al in a single center experience involving 18 patients that children older than 6 months with anatomically suitable lesions were occasionally considered for surgical excision of the hemangioma as a first line of treatment.⁶ Small preseptal lesions located away from the lid margin can generally be excised simply with minimal scars and may be removed for cosmetic reasons. Very large hemangiomas can be partially excised. Partial excision should be considered in situations where the partial removal will result in reducing the threat to visual impairment. None of the patients that underwent surgery suffered from persistent ptosis or ocular muscle dysfunction that requires further surgery. None had any rebound growth. Furthermore, no patient suffered deterioration visual function post-operatively.⁶

Most practitioners may have an assumption that hemangioma operations are associated with risk of major intra operative bleeding and a high level of technical difficulties. On the contrary to this assumption, hemangioma cases are commonly presented without any significant hemorrhage when compare to AVM

cases. The tumor acts as a tissue expander, therefore adequate tissue is nearly always available for advancement as needed to close the excision site. According to (Batniji R.K et al), When hemangioma has an extensive superficial component and the entire lesion cannot be excised without violating aesthetic line or making an unduly large incision, $\pm 10\%$ of hemangioma is left behind and allowed to undergo involution.⁵

The surgical technique proposed by Batniji involves incision along the eyelid crease or trans-conjunctival approach, followed by sharp dissection around the extent of the tumor and en bloc removal.⁵ However, the remaining hemangioma should be treated with intralesion steroid or pulsed-dye therapy if available. Surgical excision has been supported by other authors both as primary treatment as well as after failure of medical treatment.^{4,5}

In this report we present two cases of children with hemangioma of the palpebra that underwent early surgical excision during proliferating phase to prevent the development of amblyopia and to ensure a better outcome in cosmetic appearance.

CASE 1

A 6—months old male child, with hemangioma affecting the left palpebra resulting in ptosis was brought to us. Anamnesis was conducted and from the history of the patients illness the chief complaint was two months ago small red papules were identified in the left eyebrow. The lesion has overgrowth in size over the past 1.5 months. The parents seek medical advice in Lampung general hospital. There the patient was diagnosed with palpebral hemangioma. The patient then referred to the division of Plastic and Reconstructive surgery in Cipto Mangunkusumo hospital. Decision for early surgery intervention was made after thorough history taking of present illness, physical and visual field examination of the affected eye. In the mean time regular stimulation of the affected eye by flash light was conducted every four hours to preserve visual function.

Since intra operative bleeding was minimal, anesthesia should also be minimal.



Figure 1. Pre operative , the large mass of tumor causes ptosis affecting the left eyelid.

Figure 2. Conservative therapy with routine flash light to preserve vision.

Figure 3. Incision was made in the lid crease and excised up to the level of the levator muscle. Part of the deeper infiltrating hemangioma was left untouched.

Figure 4. The remaining lesion (extra orbital) is treated with intralesional injection.

Figure 5. The defect was closed with FTSG taken from bilateral retroauricular.

Figure 6. Three month post surgery, mild ptosis was still observed, the scar is still edema and the remaining cutaneous lesion that was injected with intra lesion corticosteroid beginning to show signs of involution (flattened, pale spots, decrease in size).

We performed excision of the hemangioma until the eyelid crease below the brow. The depth of the incision was up to the levator apparatus. Immediate closure of the defect with Full Thickness Skin Grafts from bilateral retroauricular region was performed. The remaining dermal hemangioma was treated with intralesion steroid injection. Later staged resection is often necessary, sometimes necessitating correction of palpebral length, ciliary margin, and adjustment of the levator apparatus is needed.

CASE 2

A 3-months old female child was diagnosed with smaller lesion of hemangioma palpebra of the right eye. She was brought by her parents because they fear that the tumor will grow bigger and might affect their child's vision in the future. They also requested to be excised and immediate reconstruction hoping that the child may have a good esthetic result. She had undergone excision of the tumor, the tarsal and conjunctival was left unexcised. The defect was reconstructed with bi-pedicled advancement flap.



Figures 7. Pre-operative: The lesion was preseptal and the size is about half of the palpebra.

Figures 8. Hemangioma was excised up until the subcutaneous tissue. The incision design was lid crease, and the defect was closed with bipedicle advancement flap.

Figures 9. 1 month post op, the wound heals without any scarring, although swelling was still visible.

Figures 10. 3 month post op the wound was almost mature, although there is still a little swelling left. However the result was satisfactory.

Various perspective for treatment of periorbital hemangioma

Several decades ago treatment for hemangioma over the eyelid using radiotherapy and sclerosing agent has been given up due to its long term sequelae and causing significant scarring.

Previous study by M.L. Kulkarni suggested systemic steroids for treatment of hemangioma which was present right from birth and occluding the right palpebral fissure or to treat the complications of Kasabach-Merritt Syndrome. The child was given a high dose of 1-2mg/kg/day for a period of a month and followed with tapering off.⁷

High local concentration of intralesional corticosteroids is another treatment option for rapidly enlarging hemangioma, with a mix of long and short acting steroid (i.e. 40 mg/ml of triamcinolone and 6 mg/ml of betamethasone mixed in a syringe). But this needs repeated

injections and carries significant complications (central retinal artery occlusion, full thickness eyelid necrosis, linear subcutaneous atrophy and localized lipoatrophy and pain).⁷

On the contrary Jordan M. Graff et al reported disadvantage of oral administration of prednisone which causing a child with hemangioma palpebra becomes irritable. There were no significant reductions in size of the lesion after the initiation of therapy using intralesion injection, topical and systemic steroid.² Pulsed dye laser is recent therapeutic tools useful for early superficial lesions and treatment option for ulcerated hemangioma.⁵

Surgical principles for peri-orbital hemangioma

Once a decision to treat a hemangioma with surgery is made, the exact technique to be utilized must also be determined. The most commonly used technique for small lesions is



very straightforward and involves removing the abnormal vascular tissue with a lenticular, or lens-shaped excision, that results in a linear scar. Recently, some surgeons have been advocating the use of an elliptical, circular, or irregular incision shapes, followed by a purse-string-type closure. This technique does result in a scar having radial (star shaped) ridges that can take several weeks to flatten. However, the overall result is a shorter scar that can be followed up by removal, using the lenticular excision technique.⁸

For upper lid hemangiomas, scars should ideally run in the lid crease and below or above eyebrow. This gives the most cosmetically acceptable result and allows good access to the intraorbital component of hemangiomas along the levator apparatus.^{5,6}

In a study conducted by Jenny L et.al is stated that management of surgical excision is generally reserved for lesion that are either sight threatening and cosmetically disfiguring in school aged children.⁶ Children older than 6 month with anatomically suitable lesions were occasionally considered for surgical excision of the hemangioma as a first line treatment. In their study 15 patients had hemangiomas affecting the upper lid to prevent amblyopia worsening. Ten of these patients had prior oral steroids. Few complications had been reported one patient had hematoma at two days post operative, another had intradermal cyst of the scar. Mild ptosis was seen in two patients, which resolved spontaneously within 6 months of surgery. No patient suffered persistent ptosis.⁶ A major review of the treatment options for infantile periocular hemangioma suggested that there was very little evidence to show that surgery was detrimental even during the hypertrophic phase.

Several authors have advocated the use of surgery for selected hemangioma. Plager and Snyder stated that the best cases for surgery are discrete lesions that tend to shell out cleanly, rather than diffuse, infiltrative lesions with indistinct borders.⁹

DISCUSSION

In our first case, a 6 month old boy has developed a large lesion resulting in ptosis. The

tumor was excised up to the levator muscles. Lid crease incision was preferred in this case. There's usually a pseudo capsule around the hemangioma that is a-vascular and aids excision. Parts that could not be excised was left in place. Lesion above the eye lid crease was treated with corticosteroid intra-lesion injection (Flamicort®) (case 1 fig C). The purpose of this procedure is to be able to observe directly the duration of accelerated involution phase (case 1 fig F). In addition, the use of donor tissue is minimized, which will decrease the morbidity of the tissue that is utilized to close the defect. Intra-operative bleeding was minimal and caused no technical difficulties in the excision of the lesion. Any hemorrhage encountered was effectively controlled using fine-tipped bipolar forceps. After removal of the tumor, the defect was primary closed using FTSG donor from bilateral retroauricular. Seven days after surgery the patient was able to open his eyes. Although FTSG was one hundred percent take, there was a still sign of inflammation. However, the most important aspect and successful value was the absent of restriction of vision. As predicted post-operatively mild ptosis still persist. Three month post operative, FTSG was mature and the result was very satisfying.

In the second case, hemangioma was excised also until the lid crease and the defect was closed with bilateral local advancement flap. Three months post-operatively, the wound healing was satisfactory, although light edema was still present.

Skillful and an experienced surgeon is another aspect to be considered to determine better result and cosmetic outcome. Consultation with a plastic surgeon is indicated for symptomatic involuting or proliferating lesions that are unresponsive to medical therapy and for which surgical excision is being contemplated.¹⁰ Local anesthetic infiltration of 1% lidocaine with 1:200,000 adrenaline and 1500 iu of hyaluronidase support both hemostasis and the dissection considerably. When levator aponeurosis is involved in the hemangioma, it can be carefully repaired using 6-0 prolene vertical mattress suture.

Early surgical intervention should be considered in a multidisciplinary team approach as a primary treatment option with selected, isolated hemangiomas, without a significant cutaneous component. Surgery is a safe, effective treatment for selected lesions, provides a definitive early treatment, and prevents astigmatism and occlusion-related amblyopia.¹¹ Besides these benefits surgery promises added esthetic outcome. These benefit, esthetic and function, can be seen directly after surgery and follow up after surgery.

Conservative therapy suggested to patients with large hemangioma of the superior palpebra is to apply light stimulus to the affected eye every four hour to prevent damage to the development of the eye nerve. By accepting that hemangiomas do not necessarily need to be completely excised, even infiltrative hemangiomas may be considered for surgical excision.

Our experience suggests that excision of these hemangiomas presents no particular technical difficulty. The affected skin is excised completely where possible. Partial excision of the affected skin can be undertaken if total excision would result in a cosmetically compromised wound closure. Extensive hemangiomas may be excised partially to protect sight with the aim of minimizing the morbidity of surgery and achieving the best final cosmetic outcome.

To sum up, Therefore, proper surgery can overcome problems such as: preventing the development of amblyopia, calming overly anxious parents, and the surgeon need not to fear surgical intervention in large tumor since the results were definitely advantageous.

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