CASE REPORT

Successful management of bilateral periorbital necrotising fasciitis with ocular involvement

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SUMMARY

A 4-year-old girl had suffered from multiple pustules and severely swollen eyelids for a week which led to difficulties in opening her eyes. The pustules burst 2 days prior to admission, causing blood and pus to come out. As a result, the evaluation of eye movement, visual acuity and intraocular pressure was hindered. Eyelids were spasming, hyperaemic, warm and of soft consistency on palpation. Crust and necrotic tissues were also observed. The patient had a history of fever with cough and nasal congestion. CT scan revealed soft tissue thickening on bilateral anterolateral periorbital region. She also underwent debridement in both eyelids, followed by partial periosteal graft on the left eye due to corneal perforation. During follow-up after 1.5 months, she also underwent excision of the periosteal graft and synechiolysis, followed by cataract extraction of the left eye.

BACKGROUND

Necrotising fasciitis is a rare severe infection involving subcutaneous tissue and superficial fascia with secondary necrosis of the overlying skin. It is characterised by rapidly progressing necrotising infection which worsens over hours. The most common causative organism is group A β -haemolytic *Streptococcus*.¹ Necrotising fasciitis commonly occurs on the extremities, abdomen and perineum, but rarely found on the head and neck. However, periorbital region is the most common site to be affected. It is usually associated with preceding trauma or surgical intervention involving periorbital area.^{1–3} Rapidly progressing nature of necrotising fasciitis leads to poor prognosis which contributes to the high morbidity and mortality rate of 8%–15% and 13%–30% rate of vision loss.⁴ Consequently, prompt diagnosis and treatment of necrotising fasciitis are crucial.^{3–5} Broad spectrum antibiotics, surgical exploration and debridement are the main treatments for necrotising fasciitis.^{6,7}

This case report presents a rare case of bilateral periorbital necrotising fasciitis with ocular involvement. Multidisciplinary approach, proper antimicrobial and surgical treatment, as well as continuous follow-up and rehabilitation of visual function are required to achieve successful management of the disease.

CASE PRESENTATION

A 4-year-old girl had suffered from swollen eyelids with multiple pustules for a week. As the pustules

grew in size, the patient experienced difficulties in opening her eyes. Two days prior to admission, the pustules burst, causing blood and pus to come out. She had also suffered from fever with cough and nasal congestion within the previous 10 days. There was no history of teeth infection, dysuria, history of trauma, prolonged fever or prolonged cough. Due to the severe oedema, any evaluation of eye movement, visual acuity and intraocular pressure was hindered. Both eyelids were spasming, hyperaemic, warm and of soft consistency on palpation. There were also prominent crust and necrotic tissues forming. As a result, other parts of the eyes were difficult to be evaluated (figure 1). However, there was no occurrence of lymph node enlargement.

INVESTIGATIONS

Blood laboratory examinations indicated leucocytosis ($16.4 \times 10^9/L$), high C reactive protein (12 mg/dL), high procalcitonin (0.11 ng/mL), normal range of CD4 and CD8, negative HIV and non-reactive Hepatitis B surface antigen (HbsAg). There were no abnormalities in terms of liver function, renal function, electrolyte, glucose level, total IgE, IgG, IgM and IgA. Analysis on the urine sample showed cloudy, yellow-coloured urine with leucocyte sedimentation higher than normal (4–6/high-power field) and also traces of leucocyte esterase.

Orbital CT scan with and without contrast revealed soft tissue thickening on the anterolateral periorbital region bilaterally until the cutis area, with indistinct and irregular edges (figure 2). The thickening reached the anterior part of the intraocular muscle, pushing the left bulbus oculi anteroinferolaterally. There was also haziness on the sinuses, suggesting the involvement of ethmoid, bilateral sphenoid and left maxillary sinuses. The patient was therefore diagnosed with bilateral necrotising fasciitis of the eyelid. She was then hospitalised and consulted further to the paediatrics department and also to the ear, nose and throat (ENT) department.

DIFFERENTIAL DIAGNOSIS

Differential diagnoses of periorbital necrotising fasciitis would include any infectious or inflammatory processes in the periorbital region. Preseptal cellulitis and suspected orbital cellulitis may be considered as the differential diagnoses in this case.

TREATMENT

The patient was diagnosed with bilateral necrotising fasciitis of the eyelid, suspicious septicaemia

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Figure 1 Clinical appearance of the patient at initial examination: (A) severe oedema and spasm of both eyelids with crust and necrotic tissue, (B) right eye, (C) left eye.

and urinary tract infection by the paediatrics department. Intravenous ampicillin sulbactam 4×950 mg and oral ibuprofen 3×200 mg were prescribed. Meanwhile, the ENT department diagnosed the patient with upper respiratory tract infection with a differential diagnosis of sinusitis which necessitated for immediate nasoendoscopy examination.

The patient underwent debridement on both eyes. Laboratory examinations showed that *Klebsiella pneumoniae* was the micro-organism found on the swabs of the palpebra wound culture and urine culture examination. However, the result of blood culture test was negative. The necrotic tissues were eventually removed from the palpebra of both eyes. Necrotomy of the left superior palpebra caused a partial thickness defect of size 22×12×4 mm. Conjunctiva and ciliary injections were administered on the left eye; the cornea was hazy, especially on the inferior part with iris prolapse sized 2×3 mm. The anterior chamber was flat, while the other parts were difficult to be evaluated.

The patient also underwent partial periosteal graft on the inferior part of the left cornea to close the wound (figure 3). The ENT department also conducted examination under anaesthesia which revealed upper respiratory infection without any sign of sinusitis or sinus involvement. The patient was given amoxicillin clavulanate 500 mg three times per day and ibuprofen 3×200 mg/day orally, as well as gentamicin sulfate eye ointment three times per day on the left eye and levofloxacin eye-drops six times per day on both eyes.

OUTCOME AND FOLLOW-UP

The infection and inflammation on both eyes were successfully managed by a combination of multidisciplinary approach, proper antibiotics and surgical treatment. Follow-up examination 1.5 months after the surgery showed good eye movements in all

Figure 2 CT Scan orbita showed soft tissue thickening, pushing left bulbus oculi anteroinferolaterally: (A) sagittal view, (B) coronal view, (C) axial view.

Figure 3 Intraoperative pictures (A) postdebridement of both eyes, (B) right eye and (C) left eye, corneal perforation with epithelised iris prolapse on the inferior part, (D) left eye, post partial periosteal graft on the inferior part, (E) left eye, partial thickness superior palpebra defect.

directions, normal intraocular pressure, diminishing oedema and hyperpigmentation on the palpebra of both eyes (figure 4). Measurements on the right eye were within normal limit while there was cicatrix of superior palpebra without defect on the left eye; however, there was no sign of inflammation or infection in the intraocular region. The periosteal graft was healing properly, although posterior synechia and complicated cataract were observed on the left eye. Ultrasonography examination revealed normal anterior and posterior segments of the eyes. However, the patient was not cooperative during the assessment of the visual acuity.

The patient then underwent excision of the periosteal graft and synechiolysis of the left eye. Afterwards, cataract extraction and insertion of intraocular lens on the left eye were performed. Periodical follow-up examinations were highly suggested in order to evaluate the condition of the patient, especially to assess the visual acuity and to determine further plans regarding the possibility of amblyopia of the left eye.

DISCUSSION

Periorbital necrotising fasciitis is a severe and uncommon infection.^{3 4} The eyelids tend to be resistant to the infection because it has anatomical barriers that prevent the spreading of inflammation from the orbit.^{8 9} Majority of the cases are found to be unilateral and occurring in adults.^{10–12} Bilateral involvement of the eyelid is very rare which may be explained by a poor resistance to the infection spreading in the subcutaneous tissue over

Figure 4 (A) Clinical appearance of the patient (1.5 months after surgery). (B) Right eye, within normal limit. (C) Left eye, periosteal graft tissue on the inferior part of cornea with complicated cataract. (D) Left eye, minimal cicatrix on the superior palpebral region. (E) Left eye, post cataract extraction procedure (with air bubble).

the nose.¹² Clinical manifestations are crucial in the diagnosis of necrotising fasciitis.^{4,5} It is characterised by the acute onset of a painful erythematous rash with oedema on the palpebra. Necrotic changes due to progressive thrombosis of dermal and subdermal perforating vessels occur more rapidly.^{1,6} Ocular involvement in periorbital necrotising fasciitis is rarely found, with manifestation such as keratitis, uveitis or chorioretinitis.^{4,7}

The management of periorbital necrotising fasciitis includes early diagnosis and immediate aggressive multidisciplinary treatments.⁵⁻⁷ Early diagnosis, extensive debridement and prompt antibiotic treatment contribute significantly in reducing morbidity and mortality.^{7,13,14} MRI and CT scan can detect the extent and identify any soft-tissue oedema which infiltrates the fascial planes hours before the appearance of cutaneous signs.¹⁵ Aggressive debridement is crucial in removing the necrotic and infected tissues which act as the nidus for the infection because it decreases bacterial load, reduces production of toxins and enzymes, and also permits bacterial dissection through connective tissue. Traces of infection are commonly found along the avascular tissue planes where access to antibiotics is limited. Thus, early surgical debridement is very important in managing necrotising fasciitis.^{1,5} Palpebral defect commonly occurs following aggressive debridement procedure. Reconstruction on the defect can only be done when the infection has been completely resolved, and the wound has been cleaned. The technique involved in the reconstruction depends on the extent of the defect.¹²

Surgical treatment of necrotising fasciitis should be combined with intravenous antimicrobial therapy.^{1,5,6} The systemic broad-spectrum antibiotics used were proven to be effective against β -haemolytic *Streptococcus* which is the most prominent organism involved in necrotising fasciitis. After the culture and resistance tests' results are obtained, clinicians should adjust the proper antibiotic dose for the treatment.^{8,12} In this case, the patient was a 4-year-old girl with bilateral periorbital necrotising fasciitis which is extremely rare. There were pustules and infection on both eyelids since 1 week before admission. In addition, she also suffered from upper respiratory tract infection which manifested as cough and nasal congestion, followed by fever, for 10 days before admission. Urinary tract infection was also observed. These factors may contribute to the development of necrotising fasciitis.

It is essential that the diagnosis of periorbital necrotising fasciitis is made primarily based on clinical signs and symptoms, including severe oedema of the eyelids with pustules and quick formation of necrotic tissues. Corneal perforation of the left eye was found intraoperatively, although it is rarely found in necrotising fasciitis. History of fever and upper respiratory tract infection further justified the diagnosis. Additional examinations such as ancillary tests will help clinicians to make a more accurate diagnosis.

Intravenous ampicillin-sulbactam was immediately given to the patient at the initial visit. This broad-spectrum antibiotic is effective against certain gram-positive bacteria, gram-negative bacteria and anaerobes. Further examination of the necrotic tissue revealed that it was sensitive to ampicillin-sulbactam; hence, the antibiotic was deemed appropriate to be used for treatment. Considering the extensiveness of tissues that were infected, the patient underwent aggressive surgical debridement immediately. Partial periosteal graft of the left eye was also done to treat corneal perforation of the left eye. The necrotic tissues were examined by the pathology anatomy department which revealed *Klebsiella pneumoniae* as the cause of infection. The choice of reconstructive procedure depends mainly on the

extent of the defect. To cover the periorbital skin defect, split or full-thickness skin grafts are usually done because they tend to provide more satisfactory outcome.¹⁵ Reconstruction was scheduled to close the left palpebra defect caused by surgical debridement; however, it was eventually not executed because the defect healed spontaneously due to scarring process over time.

Corneal perforation, posterior synechia and complicated cataract are examples of the orbital involvements that may occur in periorbital necrotising fasciitis. They are the results of infection on the deeper structure which also causes inflammation process intraocularly.¹² Although keratoplasty is the ideal treatment for corneal perforation, partial periosteal graft procedure was done instead because of the unavailability of corneal donor. The patient also underwent synechiolysis and cataract extraction procedure later on to overcome the pupil seclusion and cataract on the left eye.

There is a lack of literature that suggests *Klebsiella pneumoniae* to be the organism responsible for periorbital necrotising fasciitis. The most common causative organism of this disease is β -haemolytic *Streptococcus* (mainly *Streptococcus pyogenes*), occasionally in combination with *Staphylococcus aureus*.^{11,12} Other causative agents that are rarely found include *Staphylococcus aureus* on its own, *Pseudomonas aeruginosa*, *Moraxella catarrhalis* and *Cryptococcus neoformans* (mainly on immunosuppressive patients).¹ Necrotising fasciitis caused by *Klebsiella pneumoniae* is strongly associated with diabetes mellitus and liver abscess; these symptoms were however absent in this patient.

Prognosis of necrotising fasciitis is poor due to the rapid progress of infection. The morbidity and mortality rates of this disease are high. Several factors which are significantly associated with the increased mortality rate include advanced age, presence of comorbidities and surgical delay more than 24 hours from admission time.^{1,7,12} Early recognition of the signs and symptoms was done in this case, while aggressive intravenous antibiotic was also administered early. Besides, there were no comorbidities on this patient which led to better prognosis. Although necrotising fasciitis is a rapidly progressing infection, extensive debridement was performed and aggressive antibiotics were given to the patient which prevented its fulminant course with a fatal outcome. The ophthalmology, paediatrics, pathology anatomy and radiology departments were involved in the multidisciplinary approach to provide comprehensive management of the disease.

Learning points

- ▶ Prompt diagnosis and treatment with systemic antibiotics and debridement of necrotic tissues are crucial in treating rarely occurring bilateral necrotising fasciitis due to its rapidly progressing nature.
- ▶ Successful management requires multidisciplinary approach, proper antimicrobial and surgical treatment.
- ▶ Management of bilateral necrotising fasciitis crucially depends on the severity of each eye.
- ▶ In cases with complication on other tissues such as corneal perforation, emergency periosteal graft can be done temporarily while waiting for corneal donor to prevent tissues from prolapsing tectonically, as well as to prevent infection from spreading further into the intraocular tissue.

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REFERENCES

- 1 Lazzeri D, Lazzeri S, Figus M, *et al.* Periorbital necrotising fasciitis. *Br J Ophthalmol* 2010;94:1577–85.
- 2 Puri P, Innes JR. Necrotising fasciitis of the orbit: early recognition and treatment a key to success. *Eur J Ophthalmol* 2001;11:180–2.
- 3 Costet-Fighiera C, Lagier J, Bastiani-Griffet F, *et al.* [Necrotizing fasciitis of the eyelids and orbit: a life-threatening ophthalmological emergency]. *J Fr Ophthalmol* 2002;25:375–8.
- 4 Flavahan PW, Cauchi P, Gregory ME, *et al.* Incidence of periorbital necrotising fasciitis in the UK population: a BOSU study. *Br J Ophthalmol* 2014;98:1177–80.
- 5 Elner VM, Demirci H, Nerad JA, *et al.* Periocular necrotizing fasciitis with visual loss pathogenesis and treatment. *Ophthalmology* 2006;113:2338–45.
- 6 Jensen SL, Amato JE, Hartstein ME, *et al.* Bilateral periorbital necrotizing fasciitis. *Arch Dermatol* 2004;140:664–6.
- 7 Amrith S, Hosdurga Pai V, Ling WW. Periorbital necrotizing fasciitis -- a review. *Acta Ophthalmol* 2013;91:596–603.
- 8 Luksich JA, Holds JB, Hartstein ME. Conservative management of necrotizing fasciitis of the eyelids. *Ophthalmology* 2002;109:2118–22.
- 9 Poitelea C, Wearne MJ. Periocular necrotising fasciitis--a case report. *Orbit* 2005;24:215–7.
- 10 Bilyk JR. Periocular infection. *Curr Opin Ophthalmol* 2007;18:414–23.
- 11 Shayegani A, MacFarlane D, Kazim M, *et al.* Streptococcal gangrene of the eyelids and orbit. *Am J Ophthalmol* 1995;120:784–92.
- 12 Stone L, Codère F, Ma SA. Streptococcal lid necrosis in previously healthy children. *Can J Ophthalmol* 1991;26:386–90.
- 13 Elwood ET, Sommerville DN, Murray JD, *et al.* Periorbital necrotizing fasciitis. *Plast Reconstr Surg* 2007;120:107e–11.
- 14 Hu V, Turner S, Robinson F. Non-progressive periorbital necrotising fasciitis. *Orbit* 2008;27:59–62.
- 15 Young SM, Leng SL. Fulminant Streptococcal Infections and Necrotizing Fasciitis. In: Mukherjee B, Yuen H, eds. *Emergencies of the Orbit and Adnexa*. India: Springer, 2016:315–25.

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