CASE REPORT

Appearances are deceptive...

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daniel.franzen@usz.ch**SUMMARY**

A 31-year-old man was referred with sudden onset of a painful periorbital angio-oedema. Assuming a first episode of allergic or acquired angio-oedema, the patient was treated with a plasma-derived C1 esterase inhibitor, tranexamic acid and fresh-frozen plasma, after the initial treatment with corticosteroids and histamine blockers was unsuccessful. As blood cultures grew group A streptococci, the diagnosis of periorbital necrotising fasciitis was made, and treatment was subsequently changed to clindamycin and ceftriaxone. Necrotising fasciitis involving the periorbital area is a rare and potentially lethal differential diagnosis of acute angio-oedema, in particular if accompanied by severe pain and/or fever. Knowledge on the appearance of this disorder cannot be overemphasised, as only early recognition and immediate initiation of high-dose penicillin and clindamycin combined with tissue debridement help to decrease the mortality and morbidity.

BACKGROUND

Our patient presented with an impressive swelling of his both orbital regions, which was erroneously taken for allergic/non-histaminergic angio-oedema. After the blood cultures turned out to be positive with the growth of β haemolytic streptococci, the diagnosis and treatment was changed to necrotising periorbital fasciitis. Our photographs give a good impression into this presentation. As the differential diagnosis is crucial, and the right diagnosis must be made visually and timely, we feel that this case report and the photographs should be spread to a broad readership caring for emergency patients.

CASE PRESENTATION

A 31-year-old man was referred with a sudden onset of rapidly progressive and painful facial oedema beginning a few hours earlier in connection with fever, chills, nausea and vomiting. He denied itching or exanthema. There was no history of allergy or (hereditary) angio-oedema. Other than immunosuppressive drugs (ciclosporine, mycophenolate mofetil and prednisone following renal transplantation because of kidney aplasia), he had bisoprolol and calcitriol, but no ACE-inhibitor.

INVESTIGATIONS

Clinical examination revealed severe angio-oedema of both periorbital regions of firm consistency, which caused pain upon palpation (figure 1). The swelling of his eyelids precluded proper eye opening. The tongue and throat were unremarkable. His vital signs and his breath sound were completely normal. Laboratory results were significant for raised inflammatory markers and acute-on-chronic

Figure 1 Assumed allergic or acquired angio-oedema immediately after hospital admission (about 12 h after initiation of facial swelling).

Figure 2 Periorbital fasciitis, which was erroneously taken for allergic or acquired angio-oedema, 24 h after the initiation of facial swelling, and 12 h after antiallergic and C1-esterase inhibitor treatment.

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renal failure (leucocytes 14.1 g/l with the eosinophil count within the normal range, C reactive protein 81 mg/l, glomerular filtration rate 23 ml/min).

TREATMENT

Assuming a first episode of acquired angio-oedema, the patient was treated with a plasma-derived C1 esterase inhibitor, tranexamic acid and fresh-frozen plasma, after the initial treatment with corticosteroids and histamine blockers were unsuccessful. Simultaneously, an antibiotic medication with piperacillin/tazobactam for suspected infection of unknown site was instituted. Since then, the angio-oedema was not progressive, and the pain was slightly improving (figure 2). However, the initial diagnosis was wrong!

OUTCOME AND FOLLOW-UP

Two days after, his C reactive protein value began to rise significantly to 408 mg/l, while the leucocytes count was falling.

Figure 3 (A) Contrast-enhanced, axial T1-weighted MRI of the head revealing diffuse subcutaneous oedema of both periorbital regions and contrast media enhancement in muscular fascia. (B) Corresponding axial T2-weighted MRI of the head.

Figure 4 (A) Periorbital fasciitis 5 days after presentation. Necrotic tissue starts to delineate. (B) After debridement of necrotic tissue including skin, orbicularis muscle, fat and fascia (septum). Levator muscles, tarsus and conjunctiva could be preserved.

Blood cultures grew group A streptococci. In the MRI of his head, the diagnosis of periorbital necrotising fasciitis was confirmed (figure 3A,B). His treatment was subsequently changed to clindamycin and ceftriaxone, and the patient recovered slowly. Thanks to his general well-being, delineation of necrosis could be awaited (figure 4A) before final surgical debridement of his eye lids including skin, orbicularis muscle and fascia (orbital septum) (figure 4B). Full necrosis excision is crucial to succeed recovery. Defects were covered in a second surgery using a free skin graft. He could be discharged from hospital after 37 days.

DISCUSSION

Periorbital necrotising fasciitis (PNF) is a rare and potentially lethal differential diagnosis of acute angio-oedema, in particular if accompanied by severe pain and/or fever. Here, we present a case of PNF in a young man, which was erroneously taken for allergic or acquired angio-oedema. As a similar, but by far less severe, case was published recently in a 45-year-old, previously healthy woman,¹ this differential diagnosis seems to gain utmost significance in view of an adequate and timely treatment. However, the incidence of PNF is rare recognising a total of 94 patients in 61 reports.² In half of the cases, the organism responsible for PNF is group A β haemolytic streptococcus. The reported overall death rate is 8.5–14.4%.^{2–3} But, potential adverse outcomes range from severe disfigurement and permanent loss of vision followed by soft-tissue defects. Knowledge on the appearance of this disorder cannot be overemphasised, as only early recognition and immediate initiation of high-dose penicillin and clindamycin combined with tissue debridement help to decrease the mortality and morbidity.^{1–3}

Learning points

- ▶ Periorbital necrotising fasciitis (PNF) is a rare and potentially lethal differential diagnosis of acute angio-oedema.
- ▶ In most of the cases, the organism responsible for PNF is group A β haemolytic streptococcus.
- ▶ Early recognition and immediate initiation of high-dose penicillin and clindamycin combined with tissue debridement is crucial.

Contributors DF, RB and KC cared for the patient. DF and RB wrote the report and reviewed the literature, KC revised the manuscript.

Competing interests None.

Patient consent Obtained.

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