

# Craniocervical necrotising fasciitis—an interesting case with review of the literature

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## Abstract

Craniocervical necrotising fasciitis (CCNF) is an aggressive and potentially fatal infection associated with high morbidity and mortality if early intervention is not implemented. Patients are often unwell at presentation, the clinical picture is often unclear thus presenting with diagnostic difficulty.

We report a case of CCNF presenting at an advanced stage and discuss the condition including its management and associated complications. © 2010 The British Association of Oral and Maxillofacial Surgeons. All rights reserved.

**Keywords:** Craniocervical; Necrotising; Fasciitis; Facial; Swelling; Infection

## Clinical relevance

Patients may initially present to their dental speciality as the onset of craniocervical necrotising fasciitis (CCNF) is commonly associated with odontogenic infections.<sup>1</sup> CCNF also involves the head and neck region and early subtle signs may therefore be identified by the maxillofacial team.

It is important to be aware of this rare and potentially life threatening condition and to include it in the differential diagnosis of unusual dental infections that may present. Early diagnosis and rapid referral to a maxillofacial department may be associated with a more favourable outcome, highlighting the importance of the requirement of oral and maxillofacial surgery in every unit.

## Introduction

Necrotising fasciitis (NF) is a rapidly progressing soft tissue bacterial infection resulting in extensive necrosis and gan-

grene of the superficial fascia and subcutaneous tissues with sparing of muscle and skin.<sup>2,3</sup> It is an aggressive infection with a rare occurrence but review of the literature suggests a worldwide increase in its incidence,<sup>4</sup> with a predilection for the lower extremities, abdomen and perineum following trauma or surgery.<sup>1,4,5</sup> NF can affect patients of all ages with no partiality to sex or race.<sup>5</sup>

NF confined to the head and neck region (craniocervical necrotising fasciitis)<sup>1</sup> is extremely uncommon with only 67 cases reported between 1945 and 1990.<sup>4</sup> Causative agents include odontogenic infections,<sup>1,3,5,6</sup> minor trauma, burns, surgery, insect bites, peritonsillar abscess and parotid infections.<sup>3–5</sup> This condition can commonly be caused by a dental source of infection<sup>1,3</sup> and can mimic a dental abscess in its early stage. It is important to be aware of this condition and to be able to include this in the differential diagnosis of atypical dental infections.

The aggressive and potentially fatal outcome of this condition is associated with extensive tissue destruction and systemic involvement.<sup>3</sup> Mortality rates were previously quoted as a range from 50% to 73%,<sup>1,7</sup> but recent reports suggest increased survival, possibly due to increased awareness amongst clinicians, with rates from 0% to 10% (without mediastinal extension).<sup>1,3,8</sup>

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Fig. 1. Diffuse swelling extending from lower lip to the upper mediastinum associated with erythema and skin vesiculation.

We discuss a case of CCNF which presented at an advanced stage and ultimately became fatal. We discuss the severity and rapid spreading nature of this condition and the importance of early diagnosis so that immediate management can be initiated.

## Case report

### History

In December 2006, a 73-year-old female attended the accident and emergency department with a two-day history of mouth ulcers, fever and chills. She noticed overnight development of right-sided jaw swelling extending into the neck and upper chest. There were no complaints of dental pain or a history of trauma or recent surgery.

Her medical history revealed ischaemic heart disease with previous heart surgery, chronic renal failure, rheumatoid arthritis, gout and osteoporosis.

### Examination

Examination revealed 92% oxygen saturation on air, BP 90/70, HR 130/min reg, RR 26/min and Temp of 37.2 °C. A diffuse swelling extending from the lower lip to the upper mediastinum associated with erythema, tenderness and skin vesiculation (Fig. 1) was noted. Intraoral examination revealed edentulous arches with no associated mucosal tenderness, swelling or ulceration. Moderate trismus was noted.

Chest examination was unremarkable.



Fig. 2. Biopsy site of skin and underlying subcutaneous tissue revealing underlying necrotic tissue.

### Investigations

Of relevance, haematological studies revealed a raised CRP (83), HB 10.9, and a reduced WCC (4.3). Arterial blood gases were consistent with moderate metabolic acidosis. Chest radiograph revealed no acute pathology. An ultrasound scan of the neck was performed at a later stage, which confirmed the presence of subcutaneous oedema.

### Management and outcome

A provisional diagnosis of septic shock was made; the underlying cause at this stage was unclear.

Immediate resuscitation measures were commenced, followed by intravenous antibiotics (benzylpenicillin and metronidazole) and steroids. Subsequent transfer to a high dependency unit for continuing management was made.

Rapid spread of ecchymosis and vesiculation of the skin of the upper chest was noted. The patient's progression in HDU deteriorated within a few hours; developing severe metabolic acidosis, airway distress and reduced level of consciousness resulting in rapid sequence intubation to secure her airway. During this procedure pharyngeal oedema and blistering were noted. A biopsy of the skin and mucosa of the chin and lower lip was taken at this time which revealed underlying necrotic tissue (Fig. 2). A clinical diagnosis of CCNF was made.

Blood cultures revealed group A  $\beta$ -haemolytic Streptococci.

The patient returned to HDU for continued medical support but died the following morning. Her demise was due to multi-organ failure secondary to septicaemia caused by necrotising streptococcal infection of subcutaneous tissues of the face and neck.

### Discussion

Necrotising fasciitis is still a rare disease, especially in the head and neck region.<sup>4,5,7</sup> Suggestions in the literature of

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