Necrotising Fasciitis of the Head and Neck: A case series from the West of Scotland

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Abstract

Introduction- Necrotising fasciitis is a rapidly progressive disease with high morbidity and mortality. Head and neck necrotising fasciitis is a rare clinical entity but may be admitted under an ENT specialist team. An understanding of the presenting features and the need for prompt management is imperative. Case Series-We present a series of patients admitted to hospitals across Greater Glasgow, United Kingdom under Otoloryngology and Maxillofacial surgical teams with head and neck necrotising fasciitis over an 8-year time period. Discussion-8 patients were identified in this case series. We highlight the clinical features, patient demographics and operative courses of this patient group. We highlight the need to recognise peri-ocular necrotising fasciitis, especially in the paediatric population which has not been discussed in the otolaryngology literature previously.

Main Document

Key Points

- Head and Neck necrotising fasciitis has a high morbidity and mortality which requires prompt intervention, surgical debridement and potential soft tissue reconstruction
- Periorbital cellulitis is a common presentation and is frequently admitted under ENT specialist teams especially in the paediatric population
- Peri-ocular necrotising fasciitis is a very rare clinical entity but can be easily missed if a high index of suspicion is not maintained
- Knowledge of the underlying pathophysiological mechanisms is vitally important when assessing patients with possible head and neck necrotising fasciitis
- To our knowledge, peri-ocular necrotising fasciitis has not been discussed in the otolaryngology literature previously

Introduction

Necrotising fasciitis (NF) is a life-threatening infection which can affect the skin, subcutaneous tissue, superficial and deep fascia with muscular extension[1]. Predisposing factors for developing NF include chronic immunocompromised states, prolonged corticosteroid use, diabetes mellitus and intra-venous drug use[1].

NF originating in and affecting the head and neck region is rare[1]. NF has been documented in patients with dental infections, traumatic neck wounds and deep space neck infections[1]. Peri-ocular NF is very rare, with a rate of 0.24 cases per million per annum[2]. Mortality rates of NF can exceed 50%, with peri-ocular NF mortality ranging from 3-10%[2,3].

The pathophysiological mechanism behind NF includes the seeding and proliferation of a bacterial pathogen in the subcutaneous tissue and fascial layers. This pathogen triggers the release of inflammatory mediators, including toxins and cytokines[2]. This inflammatory cascade results in microthrombi formation heralding

ischaemic necrosis of tissue. Severe pain, erythema, bullae formation, and surgical emphysema with systemic sepsis are hallmark features which should raise suspicion.

Four types of necrotising fasciitis have been described in the medical literature, with types 1 and 2 the most prevalent[3]. Type 1 NF is a polymicrobial infection usually consisting of mixed anaerobes and aerobes acting in a synergistic fashion[3]. Type 1 can account for up to 80% of all necrotising fasciitis cases[3]. Type 2 usually has a monomicrobial aetiology, with Group A beta-haemolytic*Streptococcus* (GAS) as the most prevalent pathogen[3]. Type 2 NF accounts for 20-30% of all cases of necrotising fasciitis, and can present as aggressive and rapidly progressing. GAS can induce a large inflammatory response by reducing phagocytic function and interferon secretion. Type 2 NF is more likely to produce bacteraemia with streptococcal toxic shock syndrome[4]. Type 3 NF is mono-microbial and usually gram negative in origin with Type 4 NF occurring in immunocompromised patients and is usually fungal in nature[4].

Management of necrotising fasciitis requires prompt recognition, intravenous anti-microbials and urgent surgical debridement.

Methods

Medical records at NHS Greater Glasgow and Clyde provided a database of all patients admitted to hospitals from 2013 onwards who were coded as "necrotising fasciitis". Data collection of the case series was undertaking from September to November 2021. Retrospective analysis of clinical records of included patients with head and neck necrotising fasciitis were used to determine site of infection, operative procedures, radiological and biochemical results.

Ethical Considerations- No ethics committee requirements were needed for this article

Reporting Guideline- PROCESS reporting guideline was used

Results

In total, 92 patients were admitted to hospitals across Greater Glasgow & Clyde with necrotising fasciitis from 2013-2021. Eight patients (10.4%) with head and neck NF were identified. The mean age of patients was 41 (median-43, range 1-81) years, see Table 1. Five out of 8 patients (63%) had features consistent with peri-orbital cellulitis on admission.

The average length of stay in hospital was 19.5 days (range-1-42 days). Patients, on average underwent 4 surgical procedures during their primary admission period (range 1-6). The average time to go to theatre after presentation to hospital was 9.75 hours (range 3-23 hours). All patients underwent surgical debridement and all were admitted to the intensive care unit post-operatively.

Average Laboratory Risk Indicator for Necrotizing Fasciitis (LRINEC) score on admission was 7. All patients had a raised C-reactive protein (CRP) on admission, with an average of 219 mg/L. Seven out of 8 patients had a leucocytosis on admission with an average value of 27.8 x10ˆ9/L.

Six (75%) patients required subsequent reconstructive surgery. Three patients underwent soft tissue reconstruction to reconstruct peri-orbital defects, see figure 1 and 2. One patient underwent enucleation of the left eye with subsequent orbital implant insertion.

Six (75%) patients had evidence of isolated pan-sensitive*Streptococcus pyogenes* in both pus and tissue samples. One patient had isolated mixed anaerobes, whilst another patient isolated*Streptococcus constellatus* . Blood cultures grew no organism in 7 of 8 samples. Six of 8 (75%) histological specimens showed evidence of necrotising fasciitis.

Two patients had no predisposing factors/illnesses. One patient had an intercurrent *Varicella Zoster* infection. Two patients were active smokers (>10 cigarettes/day) and were taking regular inhaled corticosteroids. One patient had a history of previous alcohol dependence. Two patients had a diagnosis of psoriasis and had multiple courses of topical mild and potent corticosteroids. One patient had type 2 diabetes mellitus and was

taking oral hypoglycaemic medication. One patient had been taking a long term non-steroidal inflammatory drug, namely diclofenac.

Discussion

Head and Neck NF has a high morbidity, as highlighted by our results which correlates with previous literature. NF of the face and neck in particular has a reported mortality rate of up to 25%, with 65% of cases having evidence of mediastinal extension[5]. Type 2 diabetic patients have a mortality rate of 30.3% when presenting with head and neck NF[6].

NF of the head and neck is divided into two subgroups. Firstly, the eyelids/scalp and secondly, face/neck. Minor trauma is the usual cause of the first subgroup with odontogenic infections and pharyngeal/tonsillar infections the usual causative factors of the second subgroup[5].

The presentation of head and neck NF is variable but 5/8 patients identified in this series and admitted under the ENT team presented with features consistent with peri-orbital cellulitis. One patient had evidence of parapharyngeal abscess formation with another patient having evidence of peri-tonsillar abscess. One patient developed NF as a result of a *Varicella Zoster* lesion which may have provided a route of entry for bacterial spread.

Peri-orbital or peri-ocular necrotising fasciitis is a rare entity but can have longstanding functional, cosmetic and psychological consequences as highlighted by one of the cases where the patient required enucleation of the left eye and subsequent reconstructive surgery.

NF can be very difficult to distinguish from severe cellulitis clinically. Misdiagnosis is common, especially amongst children[7]. Diagnosis of peri-ocular NF can be even more challenging due to the difficulty differentiating pre-septal from post-septal orbital cellulitis particularly in the paediatric population. Necrosis of the eyelid can become apparent quickly due to a reduction in subcutaneous fat[6]. Bilateral peri-ocular NF can occur as infection spreads across the nasal bridge, and is limited posteriorly by the highly vascularised orbicularis oculi muscle as demonstrated in 2 of the cases studied[7]. Left untreated, progression to blindness, intra-cranial disorders and death have been reported[8].

Immediate aggressive debridement of necrotic tissue down to viable tissue is important as NF spreads along avascular tissues planes, with limited antibiotic penetration. Anti-microbial therapy can have little effect on the cytotoxins which mediate the pathological process[3]. Re-exploration procedures are important to ensure any nidus of infection has been removed and to facilitate subsequent reconstructive surgery.

A systematic review by Lazzeri et al showed 28% of peri-ocular NF had no cause identified, 22% of cases were caused by penetrating injuries and 17% caused by local blunt trauma[8]. Localised eyelid swelling, pain and non-specific erythema were the most common presenting features[8].

Rajak *et al.* looked at 29 cases of peri-ocular NF with an age range of 21-95 years[2]. Fourteen percent of patients suffered unilateral visual loss, with 41% of patients requiring soft tissue reconstruction of the eyelid[2].

Fifty percent of patients with peri-ocular NF had subsequent morbidity, with a reduction in visual acuity noted over a 2-year period in the United Kingdom[9].This was consolidated by Amrith*et al.* in 2003 who looked at 95 patients, and reported that morbidity was related to loss of vision[10]. Visual loss in these patients was caused by orbital spread of the infection, central retinal artery occlusion and corneal perforation[10].

Gas formation on cross sectional imaging is non-specific and usually seen in type 1 necrotising fasciitis. This emphasises that imaging should not delay surgical intervention if there is clinical concern. If imaging is undertaken, MRI is most sensitive and specific, with a 100% sensitivity, compared to CT scanning with an 80% sensitivity.

Mortality of peri-ocular NF increases with late diagnosis and delay in treatment. This can be accounted for by the rarity of the condition and a lack of awareness[10].

The necessity for the ENT surgeon to have a high index of suspicion is imperative, particularly with elevated inflammatory markers and a history of rapidly progressing erythema/ecchymosis. The need to recognise this clinical entity is vitally important, especially amongst the paediatric population.

Conclusion

This study has highlighted the need to recognise head and neck NF especially the peri-ocular variant. Peri-ocular NF is likely to be admitted under the ENT team especially in the paediatric population. Extension of peri-orbital erythema to the contralateral peri-orbital side should raise the possibility of peri-ocular NF.

The need to act quickly and decisively is extremely important to reduce the burden of surgical interventions, the need for subsequent reconstructive surgery and the long-term psychosocial impacts to patients. This is the first published literature looking at peri-ocular NF in the UK from an otolaryngology view point.

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