

Cranio-cervical necrotizing fasciitis: case report and review of the literature

Fascite necrotizzante cranio-cervicale: descrizione di un caso clinico e revisione della letteratura

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Summary

Necrotising fasciitis is a rapidly progressive bacterial infection of the soft tissues and generally attacks the walls of the abdomen, the perineum, the limbs or, to a lesser degree, the cranio-cervical area. In the latter region, the infection involves the soft tissues of the neck, in a more or less extensive manner, and causes diffuse necrosis. Crepitation, areas with linear infiltration and others with fluctuation are detected on manual examination. Systemic symptoms such as fever, tachycardia, tachypnoea and signs of septic shock are always present, at least during the more advanced stages of the disease. Computed tomography may prove fundamental since it reveals an increase in the thickness and degree of impregnation of the cervical soft tissues, as well as the presence of liquid or gaseous infiltration in the thoracic areas, especially in cases of mediastinitis. Personal experience in a case is described which led to a review of the literature. The best approach in the management of this devastating condition is early diagnosis, adequate antibiotic treatment and radical surgical procedures, which may often need to be repeated several times.

Riassunto

La fascite necrotizzante è un'infezione batterica dei tessuti molli rapidamente progressiva che generalmente coinvolge la parete addominale, il perineo e le estremità e meno frequentemente il distretto cranio-facciale. In quest'ultimo caso l'infezione interessa più o meno estesamente i tessuti molli del collo provocandone una necrosi diffusa. Palpatoriamente può essere apprezzato crepito così come la presenza di zone lineari infiltrate ed anche aree di fluttuazione. Sempre presenti, almeno in fase più avanzata, sono i sintomi sistemici tra cui febbre, tachicardia, tachipnea e segni di shock settico. La TC risulta essere fondamentale potendo mettere in evidenza un incremento di spessore e di impregnazione dei tessuti molli cervicali ed anche la presenza di versamento liquido o gassoso a livello delle regioni toraciche soprattutto in caso di mediastinite. Riportiamo l'esperienza relativa ad un caso giunto alla nostra osservazione che ci ha indotto ad una revisione dei dati della letteratura. La diagnosi precoce, una copertura antibiotica adeguata e procedure chirurgiche aggressive, ripetute anche più volte, rappresentano la terapia ottimale per il trattamento di questa devastante affezione.

Introduction

Necrotising fasciitis (NF) is a devastating clinical condition, well known to physicians for over a century¹, and has been referred to by various names (streptococcus gangrene, Meleney's gangrene, gangrenous erysipelas, necrotising erysipelas)². From an epidemiological point of view, Cranio-Cervical Necrotising Fasciitis (CCNF) is a somewhat rare disease and there would appear to be less than 200 cases reported in the literature. In the majority, this serious condition originates from infections, in the cephalic area, particularly odontogenous³⁻⁵, even if there have been reports of other sources such as infections in the upper respiratory tract (pharyngeal-tonsillitis, epiglottiditis)⁶⁻⁹. Reports of cases secondary to iatrogenic lesions⁹⁻¹² are certainly rarer.

We describe our experience with a patient, referred to

our attention due to an apparently simple pharyngeal-tonsillitis, which rapidly developed into CCNF that spread into the mediastinal area giving rise to pleural empyema.

Case report

In February 2001, a 57-year-old male was referred to our Emergency Department with pharyngeal-tonsillitis; there were no important illnesses in his medical history and, until that moment, the patient had enjoyed good health. However, since his general health was poor he was hospitalised in the Infectious Diseases Department, where he received infusions of antibiotics (betalactamic and quinolinic preparations). Despite this treatment, the patient's conditions precipitated showing signs of cardio-respiratory disorder

ders and metabolic deficiency. Due to the patient's failing health, he was transferred, the next day, to the Intensive Care Unit (ICU) where, due to the acute septic condition (temperature 41°C), orotracheal intubation, after curarization, was performed; high doses of antibiotics (glycopeptides and aminoglycosides) together with adjunct therapy (cardiotonics, liquids, steroids, oxygen therapy) were administered. In spite of this treatment, the patient's general status gradually deteriorated and hepato-renal and cardio-respiratory conditions worsened.

Manual palpation of the neck revealed the presence of an unyielding area in the anterior and lateral cervical areas with signs of considerable subcutaneous phlogosis (hotness, hyperaemia). However, there was no crepitation or other signs of colliquation. An endoscopic examination revealed an infection in the pharynx and in the epiglottis (pronounced oedema and hyperaemia with diffuse secretion) while the glottis plane, as well as the remainder of the objective otorhinolaryngoiatric examination, were within normal limits. A CT scan of the neck revealed the presence of small pools of liquid/gaseous material in the bilateral latero-cervical areas (more pronounced on the left side) with considerable detachment of the muscle planes and no clear signs of abscess (Fig. 1). The liquid material had leaked into the mediastinum, resulting in expansion of the mediastinum due to the pool of liquid and gaseous material as well as considerable pleural effusion/empyema which were clearly visible on CT scan of the chest (Fig. 2). Due to the rapid deterioration of the various organs, resulting from the septic situation, the patient was submitted to left neck dissection together with anterior and posterior drainage of the mediastinum by cervicotomy. The procedure was performed making a large cervical flap (from the left mastoid to the right mediocervical area through the jugulum), which allowed good control of all the structures involved; the necrotic tissue was excised and the surrounding areas were removed until healthy bleeding tissue was reached. During surgery, all the vital structures of the neck were preserved, including the internal jugular vein and the accessory spinal nerve. After haemostasis, the flap was repositioned but the wound was not permanently stitched in order to allow for drainage to be applied. The next day, the patient once more underwent surgery for right neck dissection and for revision of the left one. Drainages were introduced, both in the mediastinum and in the neck, and lavages with antibiotic solutions (rifampicina) were performed several times a day. Histological examination of the surgical specimen demonstrated necrotic areas involving the connective tissue and the musculature, with signs of diffuse acute phlogosis. Bacteriological analysis of the specimen revealed a mixture of flora: *Streptococcus Viridans*, *Staphylococcus epidermidis* and the *Bacteroides Buccae anaerobe*.



Fig. 1. CT image of cervical area; areas of effusion of liquid mixed with gas are visible between muscle planes.

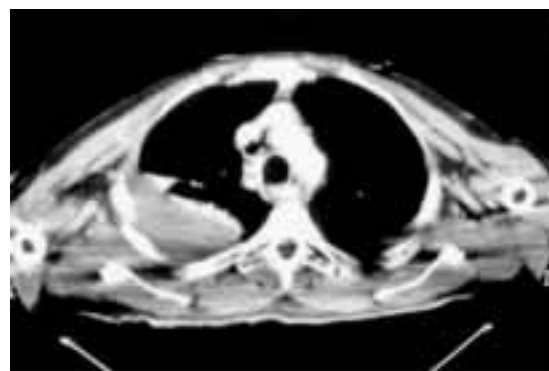


Fig. 2. CT image of thoracic area; mediastinum structures are expanded due to presence of liquid and gas. A pleural effusion is clearly visible on the right side.

The patient's condition remained critical even after these surgical procedures and he remained hospitalised in the ICU for ~80 days. During this period, the patient received antibiotic and support therapy; conventional tracheotomy permitted better management of catarrhal secretions and breathing, even if this obviously exposed the patient to further risks of contamination. The patient's general health gradually improved and he was discharged in good health and with no significant complications.

Discussion

Necrotising fasciitis is a rapidly progressive bacteri-

al infection of the soft tissues, generally involving the abdominal walls, perineum, limbs and, to a lesser degree, the cranio-cervical region^{13,14}. CCNF is usually a complication resulting from odontogenic infections³⁻⁵; cases secondary to infections in the upper respiratory tract (pharyngo-tonsillitis and epiglottiditis)⁶⁻⁹ are less frequent. Very occasionally, cases can be traced to cutaneous¹⁵, cervical¹⁶ and thyroid¹⁷ infections. The term Necrotising Fasciitis was first used by Wilson¹⁸ during the early Fifties, although the disease had already been known, for at least a century, under other names. In CCNF, the host organism is unable to circumscribe the infection and, consequently, abscesses do not develop. In fact, cervical fascia infection usually involves surrounding soft tissues more or less severely, leading to diffuse necrosis.

From a histological point of view, necrosis can be found in the connective tissue, with extension into the fascial planes and destruction of the cutaneous and subcutaneous structures. There are usually also areas of muscular necrosis, though not in a dominant manner.

Abuse of alcohol, heavy smoking, precarious social (and consequently hygienic) circumstances, as well as immunodepressive conditions, are additional factors responsible for the development of CCNF. The use of anti-inflammatory drugs (both FANS and steroids) has also been considered an important factor since these might be responsible for modifying the activity of certain cells involved in the immune processes (granulocytes)¹⁹. Albeit, this mechanism remains to be elucidated. It is worthwhile pointing out that these elements were absent in our patient and, furthermore, he had not been taking anti-inflammatory drugs or any other treatment. This aspect gives rise to several queries concerning the germ/host relationship, which, if modified, triggers this very severe disease.

If CCNF is not treated promptly, the condition quickly induces systemic toxicity that leads to multi-organ deficiency. The complications that can occur are: obstruction of the respiratory tract, occlusion of the large vessels in the neck (Lemierre's syndrome or septic thrombophlebitis of the internal jugular vein), mediastinitis, pneumonia, pericardial effusion with a risk of plugging, and also pleural effusion/empyema^{7,20}.

From an aetiological point of view, the infection is frequently polymicrobial; Gram+ cocci (*Staphylococcus*, *Streptococcus sanguis*, *mellerii*, etc) associated with Gram- bacteria (lactobacillus and diphtheroid) and anaerobic germs (*Bacteroides* sp., *Peptostreptococcus* spp, *Prevotella* and *Porphyromonas* spp, etc.) are often found. Even germs belonging to the Enterobacteriaceae (*Pseudomonas* spp, etc.) and Clostridium families have been found. The presence

of several bacteria may be responsible for a sort of 'synergism' that causes an increase in the virulence of the disease and, consequently, rapid necrosis of the tissues which is typical of CCNF⁴. In the case of our patient, the bacteriological analysis revealed a mixture of *Streptococcus Viridans*, *Staphylococcus epidermidis* and *Bacteroides Buccae anaerobe*.

Diagnosis of this devastating disease is prevalently clinical; the simultaneous presence of systemic symptoms of sepsis (fever, tachypnoea, tachycardia, signs of deficiency in several organs) and local signs such as a cervical inflammatory-type swelling, sometimes associated with pain, orientate diagnosis towards that of CCNF. The absence of pain may be justified by the fact that the infected areas can be anaesthetised due to damage to the nerve terminals. Manual palpation of the neck reveals both crepitation and streaks of linear infiltration, and even areas of fluctuation. When the disease is at an advanced stage, the prevalent signs are those of toxic shock. The decisive investigation is a CT scan of the neck, since this can reveal the presence of the liquid/gaseous material between the muscle planes. Increased thickness and enhanced impregnation of the perimuscular soft tissues and of the subcutaneous tissue are often observed. The CT images prove to be very important for detecting complications in the cervical vessels and, especially in cases of mediastinitis, the presence of effusion or empyema.

The mortality rate in this disease is related to various factors, the most important of which are undoubtedly delay in diagnosis and treatment, the general health conditions of the patient and the type of germs involved; this rate has been reported, in the literature, to be approximately 15-20%, although some authors estimate this to be higher when the mediastinic structures are involved^{3,20-24}.

Hence, the correct management of this disease is timely diagnosis followed by aggressive medical and surgical treatment. Surgery should be aimed at excising the necrotic tissue by performing a large cervical flap and, generally, a tracheotomy; it is advisable to suture the surgical wound loosely since adjustments are often needed in the area treated.

There is much debate concerning the type of antibiotic to be used, even if there is general agreement that treatment should be aimed at anaerobic and aerobic germs. Moreover, some Authors have suggested the addition of hyperbaric oxygen therapy (HOT)^{23,25}; HOT appears both to increase phagocyte activity of the neutrophils and to contribute to creating a hostile environment against the development of anaerobic germs. Moreover, hyperbaric treatment also appears to stimulate angiogenesis and the deposition of collagen.

The use of common fly maggots to devour the necrotic debris has also been suggested²⁶.

Conclusions

Cranio-cervical necrotising fasciitis is a rapidly progressive clinical condition, which, if not treated, can lead to septicaemia and multi-organ deficiency. Timely diagnosis and prompt, aggressive therapy can

reduce mortality and morbidity rates. Chest involvement is one of the relatively frequent complications and must be treated appropriately. Bearing in mind our experience in this case, the significance of risk factors should be further assessed, since these were not present in our patient.

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