



MEDICAL PROBLEMS DENTAL STUDENTS COULD IDENTIFY PART 3: LUPUS

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You can't judge a book by its cover: first recorded in 1867, this proverb has since remained in common currency and is applicable to many aspects of life in the 21st century. In the context of clinical dentistry, it serves as a plea to look beyond the mouth. In the former part of this series, I emphasised the importance of history taking as a cornerstone of holistic patient care but this should compassionately acknowledge potential gaps of what is fundamentally: a self-reported account. Examination offers the chance to piece together any missing shapes of the jigsaw. Orofacial lesions are often just the cover to a book, in some cases these may be untold stories that dental care workers hold the responsibility of enlightening to their patient. In other instances they are age-old novels that bear a considerable burden for the patient, and though we may be able to regenerate the cover, we must appreciate the broader picture. Systemic lupus erythematosus is a potentially fatal, autoimmune disease, affecting multiple organ systems of the body. More than 40% of patients with this condition will present orofacial manifestations, which constitute one of eleven diagnostic criteria for the disease.^{1,2} Subsequently, dental care workers are positioned at the forefront of an intricate multidisciplinary care team for patients with the disease.

Systemic lupus erythematosus Background and epidemiology

Lupus erythematosus is a chronic, autoimmune disorder causing widespread inflammation to connective tissues.³ The most common type, discoid lupus erythematosus (DLE), resembles a mucocutaneous condition

affecting small areas of skin. Systemic lupus erythematosus (SLE), is a more severe multi-system disease with far-reaching impacts. SLE is characterised by episodic exacerbations and remissions of symptoms, which vary in severity. Sufferers will experience mild forms of the disease for a number of years, aided by corticosteroid and immunosuppressant therapeutics, which are shown to improve life expectancy. Unfortunately, the fate of the SLE footprint on the body is rarely positive. Organ systems progressively decline and damage to organs pave life-threatening outcomes for patients with the disease. This commentary will centre attention to SLE, although recommended reading relating to DLE are mentioned in the further reading section.

SLE is estimated to affect 28 per 100,000 adults.⁴ Extrapolated across the population of England and Wales, this equates to a cohort of 10,500. This distribution is significantly skewed towards women, particularly those in their childbearing years.⁵

In the former parts of this series, suggestions were offered to allow dental care workers to screen for coeliac disease, owing to the unique oral manifestations,⁶ and osteoporosis through orthopantomography.^{7,8} Unlike these diseases, which are typically disguised of any notable clinical presentation, SLE patients may present a plethora of systemic conditions.

Aetiology and pathophysiology

The precise aetiology of SLE is poorly understood. There is a consensus agreement that the disease is governed by an extremely complex multifactorial aetiology and orchestrated by a range of genetic, hormonal

and environmental influences.⁹ As a hallmark of autoimmune disorders, predisposed individuals possess susceptibility genes, which, when activated by an environmental stimulus, allow a dysregulation of an otherwise harmonious system and contribute towards its clinical expression.¹⁰ Perturbations within the immune system include: the loss of immune tolerance; increased antigenic load; excessive T cell help; impaired B cell suppression (thus polyclonal B cell activation and hyperactivity); and the production of pathogenic autoantibodies (anti-dsDNA).¹⁰ The latter being distinctively peculiar to the disease.³ The immunopathology of SLE is an expansive area and will not be discussed further in this clinically-orientated commentary; readers may wish to refer to the papers recommended in the further reading section for more information.

Risk factors

- Female > male
- Most common between the ages 15 and 44
- Black, Asian or minority ethnic people
- Familial history of SLE.

Clinical features

SLE can be characterised by a range of clinical features. These vary considerably in severity, where some manifestations are deemed fatal. From the perspective of a dental care worker, documenting these features are necessary to complete a thorough medical history, though attention should be paid to progressive exacerbations of features presenting in the head, neck or mouth. Often the classic butterfly rash pattern is attributed as a hallmark lesion of the disease, however

such rashes are relatively uncommon and not specific to lupus erythematosus.³ Given the extensive effects across multiple organ systems, the acronym SOAPBRAIN MD,¹¹ serves as a practical *aide-mémoire* to classify the disease:

S - serositis
O - oral ulcers
A - arthritis
P - photosensitivity
B - blood disorder
R - renal disease
A - antinuclear antibodies
I - immunological disorder
N - neurological disorder
M - malar rash
D - discoid rash

Individuals who possess four or more of these eleven criteria are considered SLE patients. This classification criteria, endorsed by the American College of Rheumatology, exemplifies the need of a multidisciplinary approach to SLE identification.² Oral ulcers can arise as a result of various underlying conditions, therefore correctly assigning their aetiology is essential.

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Oral manifestations

More than 40% of individuals with SLE present oral lesions,¹ which are reported to significantly influence patients’ oral health-related quality of life (OHRQoL).¹² Despite their prevalence amongst SLE patients, oral manifestations are frequently overshadowed by other clinical features, and remain under-diagnosed.¹ Amongst these orofacial manifestations, mucocutaneous lesions are most commonly documented.¹ The classical presentation is a white plaque, consisting of a central erythematous area surrounded by keratotic striae, resembling and often confused with lichen planus.¹³ The morphology of the described lesions can vary, adding to the diagnostic challenges, and depend on the disease type. Oral

ulcers may be accompanied by lichenoid lesions, leukoplakic plaques, petechiae, raised keratotic plaques are purpura.¹ Importantly, oral ulcers, which fail to respond to initial therapeutic interventions risk malignant transformation into squamous cell carcinoma.¹⁴ Clinicians may notice xerostomia or widespread carious lesions, through subsequent Sjögren syndrome, observed in approximately 30% of SLE patients.¹⁵

COVID-19

At the time of writing, the coronavirus disease 2019 (COVID-19) pandemic has claimed the lives of over 400,000 people across 188 countries and territories.¹⁶ When a vaccine is unavailable, strict public health interventions are enforced to limit transmission, hence the widespread adoption of physical distancing and quarantine measures. Whilst curfews help slow the rate of transmission, they do not manage the disease activity. Therefore, scientists are entertaining the prospect of repurposing drugs in an attempt to lessen disease severity, arrest disease progression and reduce mortality: a *teaching old drugs new tricks* approach. The antimalarial drug

hydroxychloroquine quickly emerged as a contender and accelerated in popularity following President Trump’s endorsement of the drug - despite unproven efficacy.¹⁷ Numerous clinical trials were quickly established to explore the effect of hydroxychloroquine for patients with COVID-19,

in turn creating a shortage for patients who relied on the drug for autoimmune conditions such as lupus and rheumatoid arthritis.¹⁷ The impact of withdrawing hydroxychloroquine for a fortnight can exacerbate flares and heighten disease activity in otherwise stable SLE patients.¹⁸ This highlights the importance of an evidence-based approach, which we increasingly recognise in the practice of dentistry.¹⁹

Considerations

It is vital to institute oral management at an early stage, to improve OHRQoL and prevent the indication for future invasive procedures.¹² Surgery may exacerbate symptoms and are preferred when lupus activity subsides.³ Regular oral health care is recommended,

given the weight placed on oral ulcers as a diagnostic criterion.² At each appointment, a contemporaneous record of medications is imperative.

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