

‘Black and white and every wrong colour’: The medical history of Jane Austen and the possibility of systemic lupus erythematosus

Lupus

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Abstract

Jane Austen died 200 years ago at the age of 41 and authors have attributed her premature death to a wide variety of causes, which include Addison’s disease and lymphoma.

We have reviewed all of her available letters and extricated relevant medical information which reveal rheumatism, facial skin lesions, fever and marked fluctuation of these symptoms. The severity of these symptoms increased, leading to her death within a year.

This range of clinical features fulfils the most recent classification criteria for systemic lupus erythematosus.

Keywords

Systemic lupus erythematosus, Jane Austen, medical history

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Introduction

Jane Austen died over 200 years ago but the cause of her death still remains unknown. We have undertaken a comprehensive review of her letters, considered the previously postulated diagnoses, and with reference to the medical advances since that time have arrived at a further possibility.

It is scrutiny of both Jane’s letters and those of other members of the family that reveal the important clinical features which lead to a potential diagnosis. The letters were censored and some destroyed by her sister Cassandra who was desperate to preserve the privacy of her sister. We are fortunate, however, that the remaining letters have enough clues to sustain a diagnosis. The letters were compiled by James Edward Austen-Leigh,¹ William and Richard Austen-Leigh² and R.W.Chapman.³ Deirdre Le Faye has written the most comprehensive and scholarly critique of the letters, first published in 2011 but extending to four editions.^{4,5} Her contribution to this paper is greatly appreciated.

Her early life

Jane was born in 1775 about 4 weeks late but appears to have had a healthy and active childhood. She was

sent with her sister and cousin to board in Southampton at the age of seven and whilst there developed typhus. Infected sailors returning from Gibraltar apparently carried the infection. Her young adulthood was normal, healthy and sociable and she attended dances and visited her brothers in London and Godmersham. She suffered with minor ailments including an attack of conjunctivitis following a cold at the age of 23.⁶ (Figure 1) In 1813 she developed severe headaches with pain in her face, serious enough for her to ‘rest a cushion against her cheek’ which is suggestive of trigeminal neuralgia in the absence of any other neurological symptoms.⁷

In general there was no undue concern about her health until 1816.

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Figure 1. Jane Austen by Cassandra Austen. Pencil and watercolour circa 1810.
Source: By kind permission of the National Portrait Gallery.

Jane's medical history extracted from her letters

In the spring of 1816 Jane experienced some lack of energy which led her to visit Cheltenham for some relaxation. This was not successful, and on her return to Chawton she visited the Fowles at Kintbury, who had the impression that her health was failing.

At the end of August 1816, Jane had an attack of her illness with pain in her back.

On 8th September 1816 she writes: 'Thank you my back has given me scarcely any pain for many days.

I have an idea that agitation does it as much harm as fatigue.'

In December 1816 she found walking to dinner at Wyards was 'beyond her strength' but in January things began to improve.

On January 23rd 1817 she writes: 'I feel myself stronger than I was half a year ago' and on 27th was able to embark upon *Sanditon* which included Mr Parker's exclamation that 'sea air and sea bathing was antibilious and antirheumatic'.

On February 20th 1817. 'I am almost entirely cured of my rheumatism, just a little pain in my knee, now

and then, to make me remember what it was and keep on flannel.’

By **March 13th 1817** she is able to write, ‘I am got tolerably well again, quite equal to walking about and enjoying the air.’

But on **March 25th 1817** there is a marked change of tone. ‘I certainly have not been well for many weeks, and about a week ago was very poorly. I have had a good deal of fever at times and indifferent nights, but am considerably better now and recovering my looks a little which have been bad enough – black and white and every wrong colour. I must not depend upon being ever very blooming again’. She was too tired to continue writing *Sanditon*.

March 26th 1817. ‘A good deal of wind does not suit me as I still have a tendency to rheumatism’.

April 1817. Her condition deteriorated further with fevers and a bilious attack, and she was confined to bed from April 13th.

On **May 22nd 1817** she describes herself as being severely ill with ‘feverish nights, weakness and languor’ but with a clear head and no pain – this discharge was upon her for above a week and prompted Mr Curtis to refer her to Mr Lyford, a surgeon at Winchester.

On **May 27th 1817** a mild improvement occurred so she was up from 9 am to 10 pm and could walk from room to room. She also noticed that her face had ‘not yet recovered its proper beauty’.

In **June and July 1817** she experienced gradual deterioration with weak pulse, progressing to almost continuous sleep until she finally died on July 20th aged just 41 years. She had faintness and oppression and her face finally ‘gave one the sense of a beautiful statue’.

Discussion

Although Jane felt unwell in the spring of 1816 her specific symptoms began at the end of August and she died almost exactly 11 months later. Her illness was characterised by pain in her back and knee, an unusual intermittent skin rash, a waxing and waning course with episodes of severe illness and fever, interspersed with periods of feeling normal, finally culminating in a state of progressive debilitation.

The lack of information about her health meant that it took 147 years before Sir Zachary Cope made a diagnosis of Addison’s disease on the basis of progressive decline, fevers and the ‘pathognomonic skin appearance’.⁸ Addison in 1855⁹ described patients with asthenia, nausea and vomiting and a peculiar discoloration of the skin: these changes resulted in increased tanning of the whole body and were permanent. Jane’s rash affected her face and was multicoloured and transient.

In the 19th century Addison’s disease was frequently due to tuberculosis and both adrenal glands had to be involved. Tuberculosis accounted for at least 20% of deaths in the 17th, 18th and 19th centuries including those of Keats, Chopin and the Brontës. Jane had no chest or orthopaedic problems to suggest TB, and both her doctors, Curtis and Lyford, would have been familiar with the diagnosis.

In response to Sir Zachary Cope’s paper Dr Bevan suggested Hodgkins disease in a letter in the *BMJ* describing a patient, with a similar medical history to Jane’s, who was found to have a lymphadenoma.¹⁰ Claire Tomalin in her excellent biography favoured ‘a lymphoma like Hodgkins’¹¹ and this was strongly supported by Upfal.¹² There were no specific features to favour Hodgkins or B cell lymphoma and there is no mention of enlarged lymph glands. In addition, patients with lymphomas do not suffer from either arthritis or skin lesions. Other authors have suggested TB,¹³ carcinoma of the stomach,¹⁴ and post-infectious diseases¹⁵ but without positive evidence.

The lack of evidence for Addison’s disease or lymphoma has inspired us to search for pivotal clinical clues, which might enable us to suggest a plausible alternative diagnosis.

Rheumatism was a major feature of Jane’s history with backache reported in both August and September 1816. In February 1817 she is ‘almost entirely cured of her rheumatism, just a little pain in my knee’. At the end of March she again mentions rheumatism, so has now complained of this for eight months, suggesting an acute arthritis of multiple joints. Jane was familiar with rheumatism as her brother Charles suffered from it, and Colonel Brandon was afflicted as Elinor said in *Sense and Sensibility* ‘Did you not hear him complain of his rheumatism, and is that not the commonest infirmity of declining life?’

Another pivotal clue is the facial skin rash described in March as ‘black and white and every wrong colour’ which occurred concurrently with a fever and then resolved, at least in part, and probably settled by the time of her death. Jane’s description of these lesions is most specific and can only be interpreted as either pigmentation with areas of pallor, or with haemorrhage and bruising. An inflammatory process is likely in view of the resolution. The facial changes appeared in March at a time when the sun was emerging in Chawton and this might indicate some photosensitivity, as lesions are not described elsewhere. We have emphasised the fluctuating nature of her disease. She had symptoms in August 1816, but was better in September, only to deteriorate again in December, ‘unwilling to walk far’. She was better in February

‘completely cured of rheumatism’ but in March she deteriorated and this deterioration persisted until her death.

There were no significant events in her past history that seem relevant to her final illness as she was happy, healthy and energetic. In April and May 1817 she experienced a severe exacerbation of her illness which lasted a few weeks and she saw Mr Lyford in Winchester.

We have described the critical features of her final year which are: rheumatism, facial skin lesions, exacerbations and remissions, with fever and fatigue, progressing to an early death in a young female.

We have evaluated diseases producing an acute polyarthritis in younger people, and are left with three possibilities:¹⁶

- 1). spondyloarthritis, 2). rheumatoid arthritis, and
- 3). systemic lupus erythematosus (SLE).

Spondyloarthritis is an umbrella term which includes ankylosing spondylitis and extra-articular features such as psoriasis, uveitis, and bowel disease. Men are often affected and mortality is in the normal range.

Rheumatoid arthritis involves multiple joints, particularly the hands with synovial involvement and nodules. These two conditions do not show pathognomic skin lesions, and they rarely have a limited life expectancy.

SLE was first described 34 years after Jane’s death¹⁷ and is a multisystem autoimmune disease with antibodies that produce immune complexes which activate inflammatory pathways. There is a marked female preponderance of 9:1 with age of onset from late teens, and death often occurring in the 30s or 40s.¹⁸

An international consortium has recently developed a comprehensive new classification for SLE,¹⁹ instigated by the European League against Arthritis and the American College of Rheumatologists, with the aim of improving the sensitivity (96.1%) and specificity (93.4%) of the diagnosis of lupus. They establish clinical and haematological criteria (domains) based on a weighted points system. If we apply this system to Jane Austen, inevitably including only clinical criteria, she achieves the necessary 10 points to confirm the diagnosis. Indeed she earns 12 points, with joint problems (6 points), skin problems (4 points) and fevers (2 points).

Joint problems (musculoskeletal)

Joint involvement was defined as synovitis or tenderness in two or more joints. Joint pain and stiffness was present in 89% of patients and although any joint could be involved, the hand and the knee were more often affected in females.

Skin changes (mucocutaneous)

The skin changes are usually on the face due to light sensitivity, and the changes are characteristic and occur in 70% of cases. The subacute form is an annular or papulosquamous cutaneous eruption. In addition a

discoid form is characterised by an erythematous violaceous rash with secondary changes of scarring and dyspigmentation. Acute cutaneous lupus has a characteristic butterfly rash over the nose and cheeks (malar rash). Resolution is a feature.

Fever and fatigue (constitutional)

Fever is now recognised as a specific criterion for the diagnosis of SLE together with fatigue. In the cohort of 339 patients (93% female) recruited for the classification study, fever occurred in 54%, and fatigue in 89% in the first year of disease.

In addition, the fluctuating form of Jane’s illness is consistent with current medical experience. Flares and remissions are a prominent feature of SLE and a study of 460 cases showed that flares represented an exacerbation of symptoms or an altered immune status and could assist in treatment.²⁰ In another study of 109 patients flares were predictive of outcome.²¹

Early death is frequent in SLE in contrast to other causes of acute arthritis. Mortality from SLE in the UK (1999–2012)²² recorded 2740 cases with 227 deaths which is 67% higher than controls: 75% were females with a mean disease duration of 3.8 years. This is despite modern treatment. The lethal nature of the disease is emphasised by a paper in April 2018 ‘*Lupus is quietly killing young women*’.²³ Pathology of several patients under our care showed vasculitis, with inflammation and infarcts in the heart, kidneys and brain.²⁴

Conclusion

‘Sickness is a dangerous indulgence at my time of life’ wrote Jane Austen in March 1817 and indeed she was quite right. However, the extraction of medical information from her remaining letters has enabled us to make certain deductions, and as Hippocrates stated ‘Deductions are to be made only from facts’. The facts reveal a history of prominent joint and skin problems, fever and fatigue occurring in a fluctuating disease. These facts closely accord with the latest classification criteria for SLE from the leading physicians in this field. Thus for 200 years this disease has retained the clinical pattern that has been so devastating for young females.

We would therefore like to postulate that the demise of this distinguished author was due to systemic lupus erythematosus.

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