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BRIEF REPORT

SSc and the 2013 diagnostic criteria: the case of Paul Klee's manual pathology and dysgraphia

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Abstract The German-Swiss modernist painter Paul Klee (1879–1940) suffered in the final years of his life from a severe illness, diagnosed in 1936 as scleroderma, later renamed SSc. New classification criteria for this disease issued in 2013 now allow for a diagnosis to be confirmed. Important for this process, however, is the question of whether or not Klee's hands were affected by his illness. The morphology of the artist's hands and evidence of dysgraphic changes in his handwriting are reviewed as indications of his manual pathology. Despite his illness, Klee triumphed over his infirmity, simplifying his painting and drawing styles and substantially increasing his artistic output from 1936 until his death in 1940.

Keywords Diagnosis · Dysgraphia · Micrographia · SSc

Introduction

The twentieth century artist Paul Klee, a leader in the modernist movement, has been a figure of interest in the history of medicine since 1940, when he died at age 61 from scleroderma, later renamed systemic sclerosis (SSc). This condition is a devastating auto-immune connective tissue disease, a microvasculopathy with fibroblastic dysfunction, leading to extracellular matrix deposition, malignant in its progression and in the suffering that it causes its victims.

The extensive literature dedicated to Klee's life, his art and his illness includes much subjective information about these matters in his own writings and in accounts by his family and personal acquaintances [1, 2]. The present review, however, will draw primarily on the writings of medical authors.

From Klee's correspondence, we learn that as a child he suffered from diphtheria and in early adulthood, he suffered from recurrent upper respiratory infections. He reported arthritis in his right shoulder in 1930 and arthritic pain in his hands thereafter. Most important medically, however, was a mysterious illness which first afflicted him in 1935, aged 56.

An extreme fatigue kept him bedridden for a year, and he was also troubled by dyspnoea due to bilateral broncho-pneumonia, pleurisy and myocarditis. An excoriating rash covered his body, which was diagnosed as 'measles' by some, and by others as 'scarlet fever', or an allergic reaction to medication, or the effect of poisoning from paints and solvents, or perhaps telangiectasies. Klee developed disturbing intestinal hypermotility, and also dysphagia, forcing him to consume fluids and pureed foods only [3].

The diagnosis of scleroderma was proposed in 1936, and from that time onward, a progressive systemic disease developed until his death in June 1940 [3–9]. The rash reappeared in 1939, but in a lesser form, making it unlikely to be of infectious origin, neither viral nor bacterial, but rather an expression of the SSc.

Diagnosis of Paul Klee's disease according to ACR/EULAR criteria

In view of developments in the diagnostic procedures for SSc, new criteria for diagnosis based on points accumulated in

The authors dedicate this contribution to the memory of Elaine Goren who suffered greatly from SSc, but maintained her spirit until the end.

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various scoring options were presented in 2013 by ACR/EULAR (the American College of Rheumatology and the European League against Rheumatism) [10]. According to these criteria, the presence of 'skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints' is sufficient in itself to score the necessary nine points for diagnosis.

It has been suggested that Klee's hands were not affected by his disease, but this opinion is based on the later recollections of non-medical observers and on the claim that 'up until his death, the artist continued to be able to write and produce intricate drawings and could paint without problems' [11]. Nevertheless, there is evidence, to be discussed below, which is indicative of Klee's manual pathology. This evidence will support the diagnosis of SSc on the basis of the first ACR/EULAR criterion. Alternatively, the required nine points for the diagnosis of SSc can also be obtained in Klee's case from the combination of sclerodactily (four points), Raynaud's phenomenon (three points) and interstitial lung disease (two points) [10].

Klee's manual pathology

Morphology of the hands

Le Roy compared photos of Klee's hands in 1924 with those in 1938, noting an increased tautness of the skin. Such tautness is evidence of skin thickening, as it is caused by the development of excess subcutaneous connective tissue. In a similar way, Le Roy also called attention to 'the shiny, taut skin of the face' in a photo of 1939, presenting this evidence as a 'visual verification of the diagnosis' [5]. Photographs of Klee's hands toward the end of his life show swollen dorsum of the hand, tight skin, with no visible veins or tendons, enlarged knuckles, swollen fingers with the appearance of

Fig. 1 **a** Klee's left dominant hand with swollen MP and PIP joints and flexed distal phalanxes, October 1938 (image received from Klee Zentrum, Bern). **b** Klee's hands, December 1939. Note sclerodactily with flexed distal phalanxes (image received from Klee Zentrum, Bern)

pulp diminution and slightly bent distal phalanxes (Fig. 1). Reliance on only one or a few photographs for a proposed diagnosis would of course be 'highly questionable' [3], but when, as in Klee's case, a large number of photographs all show similar morphological changes, then the evidence they present must be given weight.

Even before the onset of his other symptoms, in early 1933, Klee wrote that he experienced significant discomfort in his fingers when the temperature fell below freezing, while on the contrary, he never felt pain in his fingers when the weather was very hot. This appears to be a subjective description of Raynaud's phenomenon [9] which, with its impairment of circulation and eventual contracture of the fingers, would produce morphological changes in the hands.

No colour photos are available to indicate a change in circulation on the basis of paleness or cyanosis. It was reported, however, that by November 1937, the artist needed to use longer and wider brushes for his painting because of 'contractures of the fingers' [9]. Klee was able to 'paint without problems' [11] in his final years only because of this adaptation of his working methods to accommodate his disability. The change in the tools which Klee used in his work, together with the simplification of both his painting and his drawing styles, allowed him to increase his output of artistic works significantly in the last years of his life despite his manual impairment.

The spirit of SSc patients is often acknowledged in the medical literature. It is an 'indomitable spirit' [3, 12] that allows these unfortunate patients to rebound from a period of inactivity with an outburst of energy. They have to 'Endure!' (as Klee entitled one of his 1940 drawings) in order to overcome their infirmity [13]. In Klee's case, this endurance enabled him to produce over 1250 art works in 1939, the highest output he achieved during any year of his career [14].

Graphological changes

A review of Klee's handwriting leads to interesting observations. Earlier, in 1920, higher than usual letters are found in his correspondence, neatly arranged in straight lines, which would suggest his artistic precision. His handwriting is also well organised in cataloguing his work in 1923 and 1925 (Fig. 2).

During the third decade of the century, when SSc was diagnosed, a decline in calligraphic neatness, or dysgraphia, becomes evident. In parallel with this change, a progressive micrographia develops, with the written letters becoming smaller (thus minimising finger movement), irregular in size and arranged in a non-aligned order. A further deterioration in his writing is observed in 1939, more pronounced in early 1940, and finally, in May 11,1940, 1 month prior to

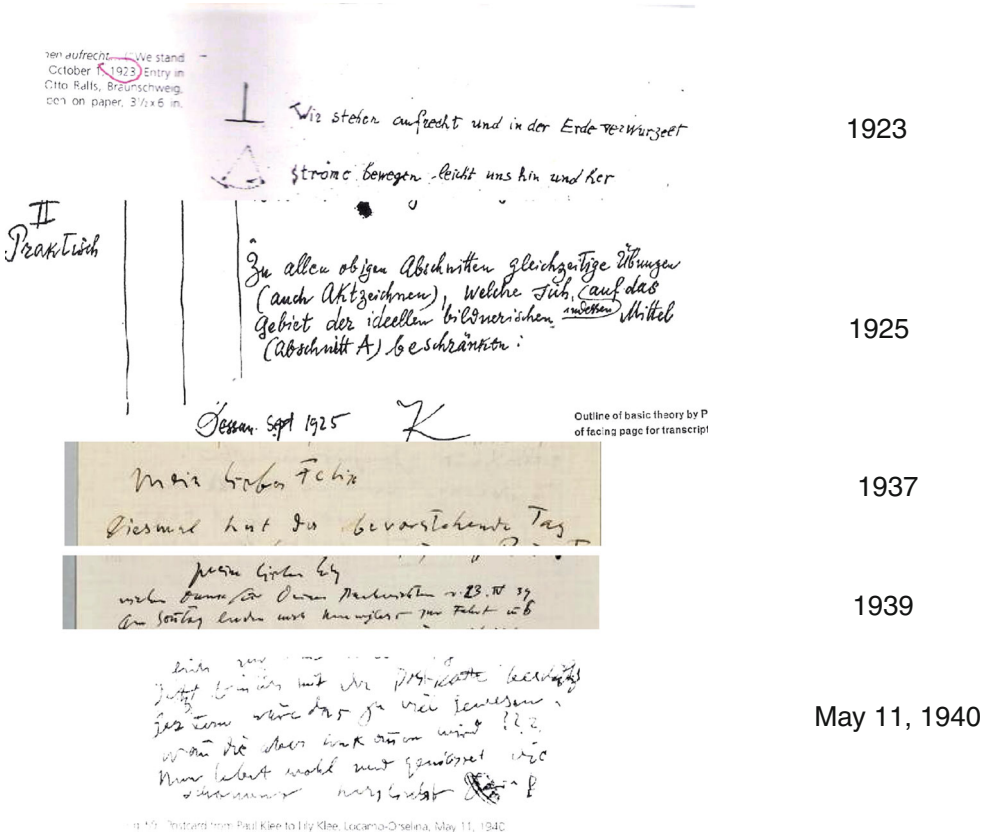

Fig. 2 The progressive deterioration of Klee's handwriting, from 1923 until 1940, a few days before his death

his death on 29 June, his handwriting is significantly distorted (Fig. 2).

Such a deterioration in handwriting may occur for several reasons other than manual pathology. Klee may have felt a need for haste as he became more aware of the fatal nature of his disease and the limited time remaining to him [14], or he may have experienced sporadic periods of excessive fatigue [3], or he could have been psychologically depressed because of his situation and therefore uninterested in what he was writing.

In the case of Klee's late drawings, however, where haste was certainly involved, the result was increased simplicity and clarity rather than the unsteadiness shown in his handwriting [14]. The simplicity of these compositions would have enabled Klee to draw with broad sweeps of his arm rather than precise movements of the fingers, something which is not possible when writing, thus suggesting that it was the condition of his fingers which accounted for his dysgraphia.

The consideration of excessive fatigue as a possible causal factor must take into account Klee's prodigious output of artistic works in his final years. In 1938, he produced 490 pictures, 'more than he had ever made before in a single year', and in 1939 'the extraordinary number of 1,253' [14]. Bouts of extreme lassitude could only have been episodic, followed by a recovery of energy. One does in fact find Klee's handwriting becoming worse on some occasions and then becoming better again, which is consistent with sporadic fatigue sometimes contributing to his dysgraphia. But the overall trend in his last years is one of progressive deterioration of his handwriting.

Finally, the hypothesis of psychological depression is contradicted by all reports of Klee's mental state from those who knew him in his final years, both family and close friends [1, 2]. The conclusion, then, must be that the deterioration of Klee's handwriting was largely if not wholly caused by his manual impairment.

Conclusions

The new ACE/EULAR diagnostic scoring system allows a retrospective diagnosis to be made of Paul Klee's disease as SSc. Progressive deterioration in calligraphy and the development of micrographia support the diagnosis of SSc affecting the hands and significantly reducing the dexterity of the painter.

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Conflict of interest The authors declare no conflict of interest.

References

1. Grohmann W (1954) Paul Klee. Lund Humphries, London
2. Klee F (1964) Recollections. In: Klee F (ed) The diaries of Paul Klee, 1898–1918, tr. P. B. Schneider, R. Y. Zachery, and M. Knight. University of California Press, Berkeley, p 418
3. Suter H (2010) Paul Klee and his illness. Karger, Basel
4. Varga J (2004) Illness and art: the legacy of Paul Klee. Curr Opin Rheumatol 16:714–717
5. Le Roy EC, Silver RM (1996) Paul Klee and scleroderma. Bull Rheum Dis 45(6):4–6
6. Keller C, Dotz W, Varga J (1999) Scleroderma of Paul Klee, part I. Dermatopathology 5(1):19–22
7. Dotz W, Varga J, Keller C (1999) Scleroderma of Paul Klee, part II. Dermatopathology 5(1):23–25
8. Cahoon L (2008) Paul Klee, a tragic metamorphosis. Science 322(5902):669
9. Ostendorf B, Maiburg B, Schneider M (2004) Sklerodermie und Paul Klee. Z Rheumatol 63(4):318–325
10. van den Hoogen F et al (2013) 2013 classification criteria for SSc. Arthritis Rheum 65(11):2737–2747
11. Suter H (2013) Case report on the illness of Paul Klee (1879–1940). Case Rep Dermatol 6:108–113
12. Cole TB (2013) Hot-blooded girl. JAMA 309(15):1562
13. Wolf G (1999) Endure!: how Paul Klee's illness influenced his art. Lancet 353:1516–1518
14. Franciscono M (1991) Paul Klee: his work and thought. University of Chicago Press, Chicago