

Original communication

# Pseudosclerodermatous triad of perniosis, pulp atrophy and ‘parrot-beaked’ clawing of the nails – a newly recognized syndrome of chronic crack-cocaine use

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Received 20 October 2005; accepted 19 January 2006

Available online 17 April 2006

## Abstract

The number of people dependent on crack-cocaine in the UK has increased substantially in recent years. Some crack-cocaine users develop coarsening changes in the appearance of their hands after prolonged use of the drug.

These changes have most often been recognized in females and include:

- (i) Perniosis with cold, numb hands, sometimes with pernioitic hyperkeratosis over the knuckles.
- (ii) Finger pulp atrophy of the distal part of the pulps of some digits, especially the thumbs and index fingers.
- (iii) Claw-like curvature of the nails. As the distal pulp is lost, it can no longer splint the nail straight and so the nail curves, claw-like, and reminiscent of a parrot’s beak as it clings to the new contour. As the pulp atrophy progresses, the nail eventually also becomes smaller.

This triad may be due to ischemia consequent upon peripheral vasoconstriction induced by crack-cocaine. Early changes may resolve with abstinence. In the patients described the syndrome does not appear to be related to intravenous drug usage. It may occur without concomitant use of heroin, whether smoked or via the intravenous route. The syndrome does not occur in all crack-cocaine users. It is hypothesized that those with a vasoreactive circulation (i.e., those with vasomotor instability/perniosis) are more susceptible to this reaction pattern.

The syndrome consisting of the triad of perniosis, pulp atrophy and parrot-beaked clawing of the nails should alert the clinician to the possibility of prolonged crack-cocaine misuse.

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## 1. Introduction

In recent years, there has been a dramatic increase in the use of crack-cocaine in the UK. A large number of individuals addicted to illicit drugs are assessed in police custody by forensic physicians. There has been a threefold increase of the numbers of crack-cocaine misusers assessed in police custody in the last decade or so.<sup>1</sup>

Cocaine is a highly addictive central nervous system stimulant, extracted and refined from the leaves of the

coca plant (*Erythroxylon coca*). ‘Crack’ is the street name given to cocaine that has been processed from cocaine hydrochloride. It produces a crackling sound when smoked. It is made by adding baking soda to aqueous cocaine hydrochloride and heating it until the freebase cocaine precipitates into small pellets. The mixture is cooled then the resultant ‘rocks’ are smoked in a crack pipe.<sup>2</sup> Some crack used will binge continuously for periods of up to 200 h.<sup>3</sup> Many of those dependent on crack-cocaine may also be dependent on heroin. Many will inject one or both drugs. Specific features – burns and blackened hyperkeratotic lesions – on hands or digits

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associated with the use of hot glass cocaine pipes have led to the term ‘crack hands’.<sup>4</sup>

Regular examination of crack-cocaine dependent individuals detained in police custody has recently resulted in the observation of a phenomenon of changes in appearance of the hands – generally female hands – noted by examining doctors, police and the individuals themselves. The changes, in broad terms – appear to show a coarsening of the features of the hands.

These features have been documented prospectively in a number of cases and consideration given to the possible mechanism and explanation for such injuries.

## 2. Method

A number of persons seen in the course of general assessment in police custody gave their consent to examination and photographic documentation of their hands to further explore this phenomenon.

## 3. Case reports

### 3.1. Case 1

Figs. 1a, 1b and 1c shows the hands of a 30-year old female who had been misusing drugs since the age of 19. She did not inject and had not done so previously. She was currently smoking large amounts of crack and heroin. She had noticed that her hands were constantly cold. The figures show the palmar and dorsal surfaces of the hands. Perniosis, bolstering of the proximal nail folds and loss of the cuticles of the proximal nail folds is seen. Fig. 1c shows the ‘parrot’s beak’ clawing appearance of one thumbnail. Fig. 1d shows a lateral view of the head of a parrot is illustrated to show the ‘parrot’s beak’.

### 3.2. Case 2

Fig. 2 shows the hands of a 30-year old female who did not inject and had not previously injected. She was cur-

rently smoking large amounts of crack and heroin. She also had noticed constantly cold hands. Loss of the cuticles of the proximal nail folds is noted in Fig. 2.

### 3.3. Case 3

Fig. 3 shows the hands of a 30-year old female who had been misusing drugs for by smoking and injection for 14 years. She used both heroin and crack-cocaine daily. She had noted that her hands became numb in cold conditions. Fig. 3 shows the atrophy of the nails of the thumbs and index fingers, especially of the left hand.

### 3.4. Case 4

Fig. 4 shows the hands of a 24-year old female who had smoked heroin and crack for 3 years. She had never injected. She had noted colour changes in her hands – particularly over the knuckles associated with coldness and numbness.

### 3.5. Case 5

Figs. 5a and 5b shows the hands and thumbs of a 40-year old female who had smoked crack for 7 years she had never injected or used other drugs. She had noticed cold hands and ‘parrot’s-beaked’ clawing (see Fig. 5c) of some nails, somewhat reminiscent of clubbing. This curvature is consequent upon atrophy of the pulps of the affected digits, especially the thumbs.

### 3.6. Case 6

Fig. 6 shows the hands and thumbs of a 38-year old female who had smoked crack for 15 years. She had never injected. She had noticed that her hands became numb in the cold. Perniosis and amateur tattoos are also noted.



Fig. 1a. Dorsal view of hands of case 1.

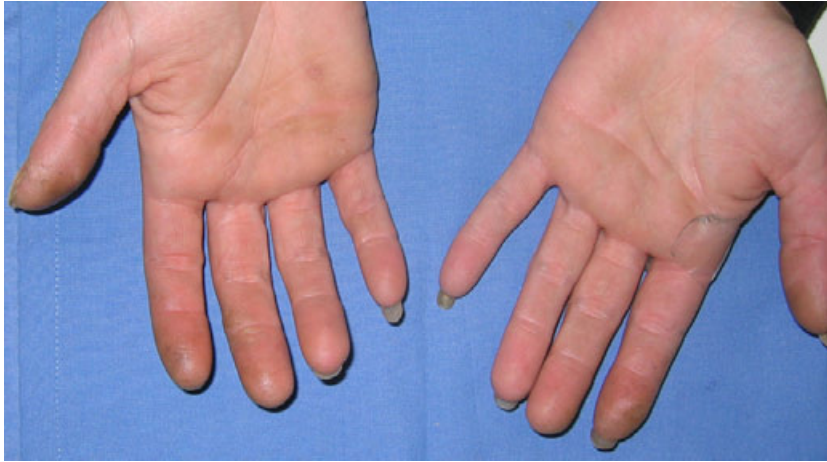


Fig. 1b. Palmar view of hands of case 1.



Fig. 1c. Thumbnail of case 1 – note the 'parrot's beak clawing' of the nail.



Fig. 1d. Lateral view of parrot's head showing the beak profile.

### 3.7. Case 7

Fig. 7a shows the hands of a 30-year old female who had smoked drugs for 15 years. She had never injected. Currently she was using crack and heroin. She had noticed that her fingernails had changed shape – becoming smaller and more rounded. This is a consequence of atrophy of the tips

of the pulps of the affected digits. Fig. 7b illustrates 'parrot's beak' appearance of the left thumb.

### 3.8. Case 8

Figs. 8a and 8b shows the hands of a 35-year old female who had injected heroin and crack in both arms since the age of 21 years. She had noticed that her hands had altered colour were sensitive to cold and appeared coarser in recent years. Perniotic hyperkeratosis (compatible with late chilblains) is noted over the knuckles of the left index and middle fingers. The cuticles of the proximal nail folds are lost.

## 4. Discussion

We believe that a newly recognized syndrome characterized at its extreme by a pseudosclerodermatous triad, of perniosis, pulp atrophy and parrot-beaked clawing of nails has been described. These appearances are most marked in females, and their presence should alert examining clinicians to the possibility of habitual crack-cocaine uses. The more widespread use of crack has led to the observations noted above. Our observations relate solely to the crack-dependent drug user who may be using almost continuously – sometimes for up to 200 h at a time – and do not apply to the recreational or occasional user of either crack- or powder-cocaine.

The assessment and interpretation of findings in drug misusers is not straightforward and the possibility of follow-up is limited because of the often chaotic lifestyle. The validity and accuracy of reported use of drugs of dependence should be treated with caution<sup>3,4</sup> but with appropriate history taking can be used appropriately. Female crack users are most reliable in admitting to use.<sup>5</sup>

Specific features – burns and blackened hyperkeratotic lesions – on hands or digits associated with the use of crack have been reported with reference to the use of hot glass cocaine pipes.<sup>6,7</sup> Reports from users dependent on crack-





Fig. 2. Dorsal surface of hands of case 2.



Fig. 3. Dorsal surface of hands of case 3.



Fig. 4. Dorsal surface of hands of case 4.

cocaine or crack-cocaine and heroin, irrespective of whether they are intravenous users confirmed and observed pattern of 'coarsening' of the hand features. These changes were noted most predominantly by females. We are not able to explain this apparent gender predominance which is in contrast with cocaine-related heart disease in which male gender predominates. Perniosis, a vasoreactive

phenomenon, describes a blueish cold circulation which is intolerant of extremes of temperature. The types of changes seen may be associated with wear and tear, such as manual work, and the perniotic changes are compatible with the perniotic changes that may be associated with riding a motorcycle without adequate gloves, where the wind chill factor may cause knuckle chilblains.



Fig. 5a. Dorsal view of hands of case 5.

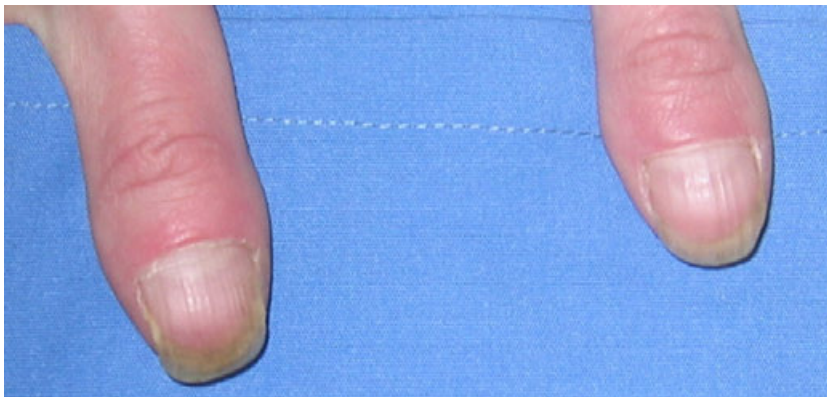


Fig. 5b. Close-up view of thumbs of case 5.



Fig. 5c. Lateral views of thumbs of case 5.

Vasoreactivity (i.e., vasomotor instability) is common, especially in women.<sup>8</sup> Vasoreactivity lies within the range of normal physiology. It describes those with a circulation that is more responsive than average, especially to temperature change. Individuals who are vasoreactors often have a pernicious circulation, especially at the hands and feet (acroperniosis). At rest, a pernicious circulation is characterized by a cold (often clammy) purplish periphery. In perniosis, cold can cause vasoconstriction severe enough to

induce temporary distal ischaemia (e.g., over the knuckles). On rewarming, vasodilatation occurs and the blood returns. If there had been enough cold damage to cause a thermal injury, then now follows reactive erythema, inflammation and oedema (i.e., chilblains). Taken far enough, these thermal injuries may cause microscopic foci of necrosis. If repeated, the affected tissue will gradually atrophy. Cocaine acting as a vasoconstrictor may be expected to aggravate the vasoconstriction that underlies perniosis. The digital cutaneous circulation has also been shown to be highly sensitive to the vasoconstrictive effects of intravenous cocaine.<sup>11</sup> Such cocaine-induced vasoconstriction persists beyond the more general hypertensive response.<sup>12</sup> It is recognized that certain individuals are at higher risk for cardiac complications, and that haemodynamic response patterns to cocaine differ amongst individuals.<sup>13</sup> Those individuals referred to in our practice and in the cases described above may use crack almost continuously in binges of several days at a time. Thus, the peripheral vasoconstrictive effects of crack-cocaine can be prolonged for hours or days. This prolonged vasoconstriction would be able to cause prolonged relative hypoxia at the periphery. We realise this is the likely reason for the syndrome. The pulp atrophy and secondary curvature and hyperplasia of





Fig. 6. Dorsal surface of hands of case 6.



Fig. 7a. Dorsal surface of hands of case 7.



Fig. 7b. Close up of lateral view of left thumb of case 7.

finger nails is reminiscent of that seen in scleroderma. However, the punctate necrotic lesions and calcinosis that characterize scleroderma are absent in the hands we describe in this patient group. These mechanisms and findings should be differentiated from acute peripheral arterial thrombosis associated with cocaine use (either powder-cocaine intranasally inhaled, or smoked crack-cocaine) which presents as distinct episodes of ischaemia<sup>9</sup> and also from acute digital gangrene that is a consequence of arterial occlusion by

impurities and can occur after accidental intraarterial injection. Cocaine (mode and amount unspecified) has also been associated with an angiographic picture of an apparent small vessel vasculitis where digital arteries were small with multiple occlusions and very limited flow.<sup>10</sup>

Diffusion capacity for carbon monoxide (an indirect measure of pulmonary circulation), decreases in those using long term cocaine either IV or by smoking.<sup>11</sup> Some of the changes appear reversible after abstinence. This reversibility may account for the apparent remission of some of the signs in the hands described by crack-cocaine users after prolonged abstinence. Perceived improvement occurred in some of our patients in whom absolute abstinence was enforced by being in prison.

We postulate therefore that, in susceptible vasoreactive persons, prolonged habitual use of crack-cocaine can result in visible digital changes (caused by multiple prolonged episodes of vasoconstriction with consequent hypoxia).

Further prospective research is required to document the incidence and prevalence of these changes (in terms of both symptoms and signs) and to relate them to length of time and mode of crack-cocaine use, and their relationship to other medical disorders. Hand X-rays would record any atrophy of the distal phalanges. Histological examination of these changes might assist in ascertaining the mechanism of change, but obtaining appropriate specimens poses ethical and practical problems. For those individuals



Fig. 8a. Dorsal view of hands of case 8.



Fig. 8b. More detailed image of dorsal surface of left hand of case 8.

who are under the care of medical practitioners, consideration should be given to performing an echocardiogram which may assist in detecting cocaine-related heart disease.

We conclude that the pseudosclerodermatous triad of perniosis, pulp atrophy and parrot-beaked clawing of hands and nails may be consequences of long term crack-cocaine usage.

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