In order to normalise the ESR and stabilise the arterial lesions, these patients were given daily doses of steroids higher than the standard for GCA (>1 mg/kg). Most of them showed significant improvements in claudication and pulses (60%). Although the pulses were not recovered in cases with total vascular thrombosis, amputation of the involved limb was avoided. The mean response time was 2–4 weeks.

Finally, this case emphasises the need to exclude GCA when ischaemia of the limbs occurs within a toxic inflammatory picture. It also emphasises the importance of excluding this type of complications in a patient with a known GCA, before attributing them to polymyalgia rheumatica.

Self-mutilation and behavioural disorder

S C Pradhan, K S Anand, A Prasad

A 26-year-old man presented with history of disinterest in performing household chores, hearing voices conspiring against himself, and poor personal hygiene. In addition, his relatives were extremely worried over the fact that the patient often pinched and scratched his nose, to the extent that he had torn off a portion of the left ala of the nose, exposing his nasal septum. The above complaints were of 13 years duration.

There was no history of head injury, epilepsy, substance abuse, prolonged elation or depression of mood or any other neurotic disorders. There was no past or family history of similar behaviour. His birth and initial developmental milestones were normal, and general physical examination was within normal limits. No abnormalities were found during systemic examinations including a detailed neurological examination. A complete haemogram, blood sugar, serum uric acid level, liver function tests and serum electrolytes were within normal limits. Electroencephalogram and computed tomographic scan did not reveal any abnormalities. Examination of the nose revealed chronic inflammatory changes of the left nasal septum with a portion of the left ala of the nose missing (figure). Detailed mental status examination revealed referential ideas, persecutory delusions of victimisation, and third-person auditory hallucinations, ie, voices discussing and threatening him.

Learning points

- intermittent claudication may be the first symptom of a giant cell (temporal) arteritis
- giant cell arteritis must be excluded in a patient with intermittent claudication associated with a toxic syndrome and/or elevated ESR which cannot be attributed to other causes

Final diagnosis

Giant cell arteritis.

Keywords: intermittent claudication; giant cell arteritis

Questions

1. What is the diagnosis?
2. What is the aetiopathology of the condition shown?
3. What are the differential diagnoses?
Answers

QUESTION 1
Chronic schizophrenia with repetitive self-mutilation.

QUESTION 2
The aetiopathology is shown in box 1.

<table>
<thead>
<tr>
<th>Chronic schizophrenia with repetitive self-mutilation: aetiopathology</th>
</tr>
</thead>
<tbody>
<tr>
<td>Psychological</td>
</tr>
<tr>
<td>● childhood sexual and physical abuse</td>
</tr>
<tr>
<td>● gratification of an unconscious need for self-punishment and establishment of self-control</td>
</tr>
<tr>
<td>● abnormal illness behaviour</td>
</tr>
</tbody>
</table>

Box 1

QUESTION 3
The differential diagnoses are shown in box 2.

<table>
<thead>
<tr>
<th>Differential diagnoses</th>
</tr>
</thead>
<tbody>
<tr>
<td>● Lesch Nyan syndrome</td>
</tr>
<tr>
<td>● Delange syndrome</td>
</tr>
<tr>
<td>● obsessive-compulsive disorder</td>
</tr>
<tr>
<td>● personality disorders, ie, borderline, histrionic, antisocial</td>
</tr>
</tbody>
</table>

Box 2

Discussion

Self-injurious behaviour may be a part of a syndrome or occur in isolation, ie, repetitive self-mutilation. Lesch Nyan syndrome is an X-linked recessive condition characterised by mental retardation, hyperuricaemia, choreiform movements, spasticity and self-injurious behaviour. In Delange syndrome, only mental retardation is associated with self-injurious behaviour. Obsessive compulsive disorder (OCD) with self-injurious behaviour, on the other hand, represents an ego alien state. However, unlike repetitive self-mutilation and personality disorders, the repetitive behaviour in OCD is designed to prevent or produce a specific circumstance.

Repetitive self-mutilation is described under the rubric of Impulse Control Disorder in DSM-IV. This disorder typically starts in adolescence, is associated with severe psychosocial dysfunction, and appears to be quite a homogenous entity compared to self-injurious behaviour without borderline personality disorder, which is often associated with a variety of psychopathological features. It is hypothesised that poor affect regulation is the underlying psychopathological mechanism of the disorder.

In the present case, the repetitive self-mutilation was due to the (predisposing) schizophrenic disorder. This combination is termed the Van Gogh syndrome, after Vincent Van Gogh (1853–1890) who, during one of his psychotic episodes, cut off a portion of his ear and sent it to a prostitute whom he had just visited. The act usually involves cutting, burning, interference with wound-healing, etc. The interesting feature in our case is the presence of insight of the patient into the act, ie, scratching and interference with wound healing, which he attributes to his prevalent mood state. This may be explained by the long duration and fluctuating nature of the illness.

Treatment modalities depend upon the nature of the disease and the associated features. The presence of psychotic states necessitates the use of antipsychotics or electroconvulsive therapy. Repetitive self-mutilation per se may be treated with serotonergic agents such as fluoxetine, or clomipramine and narcotic antagonists like naltrexone. Depending on the case, other treatment approaches may include psychodynamic psychotherapy, behavioural therapy, and self-help groups such as Self-mutilation Anonymous and SAFE (Self Abuse Finally Ends).

Final diagnosis

Chronic paranoid schizophrenia with repetitive self-mutilation (Van Gogh syndrome).

Keywords: self-injury; paranoid schizophrenia

Self-mutilation and behavioural disorder.

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