CASE REPORT

Unilateral Lower Limb Hypertrophic Osteoarthropathy due to Aortic Graft Infection and Aortoenteric Fistula

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Introduction

Secondary hypertrophic osteoarthropathy (HOA) is a clinical and radiological syndrome consisting in periostal new bone formation, digital clubbing and synovitis. It occurs most commonly in association with intrathoracic malignant diseases1-3; it has also been described associated with chronic intraabdominal processes such as Crohn's disease, cirrhosis and hepatic neoplasms.4 The clinical manifestations of HOA include subperiostal cancellous bone formation at the level of diaphysis of tubular bones, arthritis with noninflammatory synovial fluid, and clubbing of fingers or toes.5 Characteristic clinical features are generally symmetrical, involving both upper and lower limb. Changes in the lower limb rarely occur in the absence of upper limb abnormalities,6 even rarer is the restriction of HOA to one isolated limb.10 The pathophysiology of HOA is unknown and the theories proposed remain speculative; HOA most likely represents a nonspecific response to noxious humoral7,8,11,12 or neural stimuli.1-3

Some authors have reported examples of this disorder confined to the bones of one or two limbs in association with the presence of infected Dacron grafts, some of them complicated with aortoenteric fistula.7-17 The purpose of this communication is to report a new case with similar clinical characteristics, stressing the diagnostic value of HOA in the early detection of graft infection.

Case Report

A 47-year-old man was first admitted with haematemesis, melena, hypotension and falling haematocrit. Emergency laparotomy revealed a small fibrotic abdominal aortic aneurysm in association with an aortic-duodenal fistula. An aortobifemoral Dacron bypass was performed and the small bowel communication was repaired with no signs of secondary infection. After 30 days the patient was discharged.

Eighteen months later he was readmitted because of night sweats, rigors, fever and pain in his right buttock and leg. Physical examination showed a diffuse swelling of the entire leg, an effusion of the right knee, tenderness over the tibia and midthigh as well as clubbing of the toe. A right buttock abscess was also present. A mild leukocytosis and an increased erythrocyte sedimentation rate were detected. X-rays of the right leg exhibited subperiostal new bone formation and cortical thickening of tibia and femur, findings also disclosed on tomographic roentgenograms and CT-scans (Figs. 1 and 2). Synovial fluid was non-inflammatory, clear in appearance and viscous; microbiological cultures were negative. Blood cultures were positive for Streptococcus agalactiae, as were cultures from buttock abscess. Thallium scintigrams showed a diffuse increased concentration of radionuclide confined to the right leg compatible with subperiostal new bone formation along with periarticular uptake and increased activity was also present in the buttock abscess (Fig. 3). On the basis of these clinical and paraclinical features a diagnosis of unilateral HOA was made. An infected aortic bypass graft was suspected, but could not be confirmed despite upper gastrointestinal tract roentgenographic series, abdominal and
pelvic CT-scans and arteriography. The patient was treated with penicillin G, imipenem and vancomycin, improving his clinical condition.

Six months later the patient was readmitted because of persistent fever, pain and swelling of his right leg, and recurrent abscesses in right thigh and foot. A gallium scan was performed with no sign of aortic graft infection. The abscesses were surgically drained,

Fig. 1. Tomographic roentgenogram showing a marked periostal new bone formation along right femur. Note the extension of the periostal reaction and its lamellar aspect.

Fig. 2. CT-scan of both femurs show periostal new bone formation on the right, consistent with HOA.

Fig. 3. Thallium scintigram. Diffuse increased concentration in the right femur and tibia rather characteristic of subperiostal reaction. Note the periarticular uptake emphasising the presence of knee joint synovitis along with accumulation of isotope in the right buttock abscess.
Table 1. Characteristics of patients with localized HOA and graft sepsis.

<table>
<thead>
<tr>
<th>Sex/Age</th>
<th>Graft</th>
<th>Interval</th>
<th>Localized HOA</th>
<th>Infection</th>
<th>Aortoenteric fistula</th>
</tr>
</thead>
<tbody>
<tr>
<td>King J.O.</td>
<td>Female 50 years</td>
<td>Dacron</td>
<td>24 months</td>
<td>Bilateral</td>
<td>Yes</td>
</tr>
<tr>
<td>Gibson T.</td>
<td>Male 52 years</td>
<td>Dacron</td>
<td>33 months</td>
<td>Unilateral</td>
<td>Yes</td>
</tr>
<tr>
<td>Stein H.B.</td>
<td>Male 61 years</td>
<td>Dacron</td>
<td>16 months</td>
<td>Bilateral</td>
<td>?</td>
</tr>
<tr>
<td>Rosenthal L.</td>
<td>Male 59 years</td>
<td>Dacron</td>
<td>48 months</td>
<td>Unilateral</td>
<td>?</td>
</tr>
<tr>
<td>Sorin S.B.</td>
<td>Male 59 years</td>
<td>Dacron</td>
<td>24 months</td>
<td>Bilateral</td>
<td>Yes</td>
</tr>
<tr>
<td>Walter R.D.</td>
<td>Female 57 years</td>
<td>Dacron</td>
<td>108 months</td>
<td>Unilateral</td>
<td>Yes</td>
</tr>
<tr>
<td>Dalinka M.K.</td>
<td>Female 52 years</td>
<td>Dacron</td>
<td>41 months</td>
<td>Bilateral</td>
<td>Yes</td>
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<tr>
<td>Voiriot P.</td>
<td>Male 73 years</td>
<td>Dacron</td>
<td>120 months</td>
<td>Unilateral</td>
<td>Yes</td>
</tr>
<tr>
<td>Stiles R.G.</td>
<td>Male 67 years</td>
<td>Dacron</td>
<td>84 months</td>
<td>Unilateral</td>
<td>Yes</td>
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<tr>
<td>Fauquert P.</td>
<td>Male 67 years</td>
<td>Dacron</td>
<td>12 months</td>
<td>Unilateral</td>
<td>Yes</td>
</tr>
<tr>
<td>Ho A.</td>
<td>Male 76 years</td>
<td>Dacron</td>
<td>156 months</td>
<td>Unilateral</td>
<td>Yes</td>
</tr>
<tr>
<td>Present Report</td>
<td>Male 48 years</td>
<td>Dacron</td>
<td>18 months</td>
<td>Unilateral</td>
<td>Yes</td>
</tr>
</tbody>
</table>

and cultures were positive for *Morganella morganini*, *Streptococcus faecalis* and *Pseudomonas aeruginosa*. The patient received ciprofloxacin and clindamycin but despite appropriate antimicrobial therapy, a new abscess developed in the right thigh, *Staphylococcus epidermidis* and *Streptococcus sp.* were isolated. Blood cultures were negative. A worsening of his septic condition was complicated by thrombosis of the right limb of the bifurcated graft and massive gastrointestinal bleeding. An urgent laparotomy was performed. Laparotomy revealed an infected aortobifemoral graft with retroperitoneal extension and erosion into the third portion of the duodenum.

Discussion

Prosthetic aortic graft infection continues to be the most challenging diagnostic and therapeutic complication of vascular reconstruction. The possibility of graft infection must be suspected in any patient with a vascular prosthesis in whom an otherwise unexplained illness develops.

In our patient unilateral lower extremity HOA predated the clinical suspicion of graft infection. We have found 11 case reports of this disorder confined to one or both lower limbs in association with infected aortic Dacron grafts. Ho et al. reported a case of unilateral HOA confined to the upper limb in association with an infected axillary-axillary Dacron graft after 156 months of surgery. The most relevant clinical features are summarised in Table 1.

Although the pathophysiology of HOA remains unclear, the most acceptable mechanism is the humoral pathway by which an endotoxin, vasoactive compound or some other substance is activated or released by bacteria adherent or adjacent to the graft. The local involvement might be explained if the humoral factor were metabolised in the lungs or peripheral tissues. Septic embolisation and peripheral abscesses were present in our patient and in four other cases in the reviewed literature. Bacteremia was demonstrated in seven cases, and bacterial vegetations and emboli could be detected by arteriography in two. Eur J Vasc Endovasc Surg Vol 12, November 1996
In our patient, the coincidence in cultures obtained initially from blood and peripheral abscesses, together with the localisation of infection distal to the graft, lead us to agree with this theory. Other factors such as patency, changes in regional blood flow and a specific response of the Dacron material to infection have been suggested.\textsuperscript{12} On the basis of radionuclide studies, Rosenthal et al.\textsuperscript{10} postulated that periosteal oedema, subperiosteal serous effusion and subsequent new bone formation were due to a relative increase in blood flow. Other authors\textsuperscript{9,13} conclude that the development of HOA is linked to the aortoenteric fistula. The absence of HOA in dogs with experimentally induced abdominal aortic-graft infection without fistula\textsuperscript{21} supports this possibility. On the other hand, the case in which the symptoms were confined to the upper limb\textsuperscript{17} emphasises the influence of the septic graft and the Dacron material itself. When there is infection, aortoenteric fistulas are often present and cultures usually reveal polymicrobial enteric flora.\textsuperscript{14} In our patient, HOA developed several months prior to isolation of enteric flora from peripheral septic lesions; furthermore gastrointestinal bleeding occurred in the late stage of the process.

The afferent neurogenic pathway might be mediated by autonomic nerves and this may account for HOA seen in pulmonary lesions. It is supported by relief of symptoms following pulmonary resection and transthoracic vagotomy, perhaps due to reduction of blood flow to the involved extremities.\textsuperscript{1,3} We found two reports of patients with HOA secondary to the insertion of Celestine tubes\textsuperscript{22,23}; the presence of the device in the vagally-innervated portion of the oesophagus initiated the sequence of events leading to HOA, and removal of this stimulus was the factor responsible for its resolution. It is also possible that a local neurogenic mechanism is initiated by the infected graft.\textsuperscript{11,12} Indeed the two theories are not mutually exclusive as the parasympathetic nervous system may be operant in the formation of the humoral factor or may be stimulated by the humoral factor to cause the HOA.\textsuperscript{9}

Regardless of the pathogenic mechanisms, in a patient in whom HOA develops, the process should be considered secondary until an underlying pathologic condition has proven negative. Based upon this assertion we advocate the value of HOA as an aid to the early diagnosis of vascular graft infection.

\begin{thebibliography}{9}
\item Sorin SB, Askari A, Rhodes RS. Hypertrophic osteoarthropathy of the lower extremity as a manifestation of arterial graft sepsis. \textit{Arthritis Rheum} 1980; 23: 768–770.

\end{thebibliography}

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