Paget’s disease of bone in non-Caucasians in East London: a report of eight cases and a review of the literature

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Abstract

Paget’s disease of bone is more prevalent in populations of northern European origin and is thought to be rare in non-Caucasians. The population of the east end of London is multi-cultural with 45% from ethnic minorities. We report the case histories of eight non-Caucasian patients with Paget’s disease of bone.

Keywords

Paget’s disease of bone; non-Caucasians.
Introduction

Paget’s disease of bone (PDB) appears to be more prevalent in populations of northern European origin and is thought to be rare in non-Caucasians\[^1\]. The population of the east end of London is multi-cultural; 55% are Caucasians, 28% are of Bangladeshi origin, 3.5% are Black Caribbean, 3.2% Black African and 1% are Asian Indian\[^2\]. We report the case histories of eight non-Caucasian patients with PDB.

Case report

A 73-year-old Indian man, born and brought up in Kenya but resident in the UK since 1969, presented to the orthopaedic surgeons in January 1999 after falling downstairs and complaining of pain in the right hip. Examination revealed a reduced range of movement of the right hip with pain on abduction. X-Rays showed no bony injury had been sustained, but there was sclerotic change of the right hemipelvis suggestive of PDB. Serum alkaline phosphatase was elevated at 849 IU/l (normal range 70–320 IU/l). Serum prostate specific antigen level was within the normal range and an isotope bone scan confirmed the presence of PDB in the right hemipelvis with no evidence of metastatic disease.

He was later referred to the rheumatology department with continuing right hip pain and treatment with risedronate 30 mg daily for 2 months was commenced. This resulted in a marked improvement in pain and associated reduction in serum alkaline phosphatase to 304 IU/l.

A further seven patients of non-Caucasian ethnic origin with PDB have been identified. The age, ethnic origin, years of residence in the UK and sites affected are summarised in Table 1.
Table 1: Paget’s disease of bone in patients of non-Caucasian origin.

<table>
<thead>
<tr>
<th>Age</th>
<th>Sex</th>
<th>Ethnic origin</th>
<th>Years in UK</th>
<th>Sites affected</th>
</tr>
</thead>
<tbody>
<tr>
<td>73</td>
<td>M</td>
<td>Kenya (Asian Indian)</td>
<td>34</td>
<td>Right hemipelvis</td>
</tr>
<tr>
<td>55</td>
<td>M</td>
<td>Trinidad (Asian Indian)</td>
<td>33</td>
<td>Right hemipelvis, right femur</td>
</tr>
<tr>
<td>73</td>
<td>M</td>
<td>Sierra Leone (Black African)</td>
<td>60</td>
<td>Pelvis</td>
</tr>
<tr>
<td>80</td>
<td>F</td>
<td>Guyana (Black Caribbean)</td>
<td>23</td>
<td>Skull, sacrum, left tibia and femur</td>
</tr>
<tr>
<td>63</td>
<td>M</td>
<td>Bangladeshi</td>
<td>41</td>
<td>Sacrum</td>
</tr>
<tr>
<td>62</td>
<td>M</td>
<td>Jamaica (Black Caribbean)</td>
<td>41</td>
<td>Lumbar spine</td>
</tr>
<tr>
<td>58</td>
<td>M</td>
<td>Bangladeshi</td>
<td>?a</td>
<td>Left hemipelvis, femora, lumbar spine and right distal tibia</td>
</tr>
<tr>
<td>63</td>
<td>M</td>
<td>Indian</td>
<td>38</td>
<td>Right hemipelvis</td>
</tr>
</tbody>
</table>

The seventh patient’s case history has been reported elsewhere [19].

Discussion

PDB is common in Caucasians but rare below the age of 50 years [3]. Prevalence rates are estimated to be 4–5% in England; 10% of cases are over the age of 90 years [4]. Epidemiological surveys of abdominal X-rays of people over the age of 55 years have been performed in various prevalence surveys. The abdominal X-ray shows the pelvis, sacrum, lumbar spine and femoral heads, the most common sites affected by PDB.

A survey of 15 European cities found similar prevalence rates of 2–3% but a much lower prevalence in Norway and Sweden at 0.3% [5]. Prevalence rates in the north-west towns of Preston, Bolton, Wigan, Burnley and Blackburn were found to be particularly high at over 6%, with the highest rate of 8.3% in Lancaster [6]. A recent study within the UK suggests that the prevalence of PDB in patients over the age of 55 years has decreased to 2%, but the
increasing incidence with age was maintained\cite{7}. Another study suggests a similar decline in prevalence in New Zealand\cite{8}.

PDB is thought to be rare in non-Caucasians. However, a survey of radiographs of black and white hospital patients in New York and Atlanta found a different prevalence in the two cities but no difference between the races (see Table 2)\cite{9}.

In South Africa a survey of abdominal X-rays of 1003 white and 1355 black people aged over 55 years found rates of 1.3% in blacks and 2.4% in whites\cite{10}. In Africa, however, the disease is rare, although there may be geographical variation. A study of 82 000 X-ray films over a 4-year period in Freetown, Sierra Leone, identified 14 cases of PDB but no cases were seen over a similar period and number of radiological examinations in North Nigeria\cite{11}.

PDB is rare in Asians but there are reports of the disease in Chinese patients\cite{12,13}. A series of five cases of PDB collected over 8 years in Singapore also included an incidental diagnosis in an Indian man. This was a 72-year-old man in whom the diagnosis was made after he presented with a stroke and a CT scan of the head showed changes of the skull suggestive of PDB. Serum alkaline phosphatase was also raised\cite{14}. There are isolated reports of PDB in Indians and it has been postulated that the prevalence in India is underestimated as most cases are incidental findings\cite{15,16}.

In countries with European immigrant populations, the disease is found almost exclusively in those of European descent. For example, PDB is recognised in South American countries but the indigenous population is not affected\cite{17}.

\begin{table}
\centering
\caption{Rates of Paget's disease of bone in black and white residents of Atlanta and New York, USA\cite{5}.}
\begin{tabular}{|l|l|l|l|l|l|}
\hline
City & Ethnic group & No radiographs & Age standardised prevalence (%) & \\
& & & Men & Women & Both \\
\hline
New York & Black & 950 & 3.3 & 2.0 & 2.6 \\
New York & White & 1082 & 5.2 & 2.5 & 3.9 \\
Atlanta & Black & 1111 & 1.9 & 0.6 & 1.2 \\
Atlanta & White & 1563 & 0.9 & 0.8 & 0.9 \\
\hline
\end{tabular}
\end{table}
A 25-year study of PDB in four medical centres in Israel over 25 years (1961–1985) identified 278 cases in Jewish patients. All but six were immigrants, 74% from Eastern Europe, and none were Arab\[18\].

Our eight non-Caucasian patients represent only 14% of all patients with PDB attending our department, and this does not reflect the non-Caucasian population (45%) living in the east end of London. Our patients have also lived on average for 38 years (range 23–60 years) in London and may have acquired the predisposing environmental factors (if any) for the disease.

PDB is a relatively benign bone disorder common in Caucasians. The aetiology is unknown, although genetic and environmental factors are thought to be important. PDB is rare in Asians and Africans but is likely to be under-reported and no prevalence studies are reported in the literature. There is no obvious explanation for the high prevalence found in Lancashire but this is a region with a high proportion of Asian immigrants and a further epidemiological survey of rates in Asians and Caucasians may provide an indication of the relative importance of genetic and environmental factors in the aetiology.

References