Melorheostosis: Follow-up of a case Compounded with carcinoma prostate

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ABSTRACT

A case of melorheostosis, a relatively uncommon congenital condensing bone disease, developed adenocarcinoma of prostate – a relatively slow-growing tumour usually amenable to treatment with good prognosis. The malignancy in this case disseminated unusually fast spelling out the unfavourable prognosis. This patient also had a fracture neck of the femur which unexpectedly healed very early. We believe that these observations support the notion of the congenital local hypervascularity theory in the aetiology of the disease.

Key Words: Melorheostosis, TURP, secondaries, prostatectomy, orchietomy

INTRODUCTION

Melorheostosis, a rare congenital condensing bone disease, characterized by a flowing type of hyperostosis resembling “candle flow streaming” in the long bones, may be monomorphic and polyostotic. It is easily recognizable radiographically by irregular cortical thickening, periosteal proliferation and sclerosis that blends with irregular cortex involving one side of bone. The location is usually diaphyseal. The aetiology is unknown and the onset is usually insidious. Ordinarily, the prognosis of the disease is good.

The present case was initially reported with reference to a fracture neck femur with early healing of fracture, despite inadequate immobilization, supporting the hypervascularity theory originally proposed by Lorimier and Moore. We would like to clarify that this particular patient was reported earlier (1979) but this communication deals with another disorder that provides support for hypervascularity in the causation of the disease.

CASE REPORT

A 58-year-old male was a known case of monomorphic, polyostotic type of melorheostosis and was followed up diligently. In April 2000 the patient sought medical attention for complaints of increased frequency and difficulty in passing urine with distended bladder. He was provisionally diagnosed as a case of benign hyperplasia of prostate.

Digital rectal examination revealed prostatomegaly with flat anterior rectal wall mucosa, firm to hard in consistency at places, and the median sulcus was obliterated. A Foley’s catheter was placed for the continuous drainage of urine and further investigations and procedures were carried out as listed in Table 1.

DISCUSSION

This case is singularly interesting because firstly, it provided an opportunity for a long follow-up. Secondly, the natural process was complicated by two unusual events with unlikely outcomes, namely a fracture femoral neck that healed and rapid metastatic spread from a prostatic malignancy with low metastatic potential. More than 90% of prostatic metastases are osteoblastic. Secondaries may be so extensive as to produce...
diffuse chalkiness of all bones. Osteolytic lesions are rare and may be solitary.[5] It is believed that the metastatic process follows a network of valveless veins in the pelvis including the veni vasorum of the extremities and their tributaries. These veins go directly posterior to the sacral region, and also to the pelvis carrying tumour metastases with them.[6] The earlier area of bone involvement is on the inner aspect of the acetabulum, and it spreads with similar propensity in all directions from this site.

Carcinoma of the prostate is usually spheroidal-celled, with a varying degree of tubule formation and slow growth. Curiously enough, in the present case, it rapidly metastasized locally through lymphatic and venous channels. We feel it is reasonable to suggest that the hypervascularity of the pelvic bone due to melorheostosis explains the early and widespread metastases of the disease. Along with the earlier report of primary healing in a femoral neck fracture this finding supports the concept of hypervascularity as an aetiological factor in the development of melorheostosis.

**Table 1: Investigations done**

<table>
<thead>
<tr>
<th>Date</th>
<th>Investigation</th>
<th>Treatment/Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>21-04-2000</td>
<td>Abd.USG</td>
<td>BPH with residual urine of 210 ml.</td>
</tr>
<tr>
<td>25-04-2000</td>
<td>Radiograph Pelvis</td>
<td>Melorheostotic left pelvic bone (Figure 1).</td>
</tr>
<tr>
<td>02-05-2000</td>
<td>CT Scan</td>
<td>Metastatic lymph node at the level of rectosigmoid junction.</td>
</tr>
<tr>
<td>26-07-2000</td>
<td>Abd. USG</td>
<td>• Revealed enlarged lymph nodes rectosigmoid junction.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Both kidneys showed early hydronephrosis.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Post voidal urine-400 ml.</td>
</tr>
<tr>
<td>22-08-2000</td>
<td>--</td>
<td>• Post-op. radiotherapy at TMH, Mumbai</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Had incontinence of urine.</td>
</tr>
<tr>
<td>06-01-2001</td>
<td>Abd.USG</td>
<td>• Multiple secondaries in the liver (Figure 2)</td>
</tr>
<tr>
<td></td>
<td>Radiograph chest</td>
<td>• Multiple secondaries in the lung</td>
</tr>
<tr>
<td></td>
<td>Radiograph pelvis</td>
<td>• Multiple osteoblastic secondaries in the pelvic bones and vertebrae (Figure 3).</td>
</tr>
</tbody>
</table>

Melorheostosis

**Figure 1:** Radiograph pelvis showing the lesion and fracture neck femur (1977)

**Figure 2:** Large metastatic mass – right lobe of the liver

**Figure 3:** Radiograph pelvis showing osseous metastases of the pelvic bones and vertebrae
No malignant degeneration of the affected segment of bone with melorheostosis has been reported so far.2 However, more recently two cases of osteosarcoma in melorheostotic femur have been reported.8,9 Radio- 
graphs of this patient illustrate an unusual radiological combination of melorheostotic pelvic bone with secondary deposits from prostatic carcinoma in the pelvic bones.

Wahi et al10 have discussed at length the theories, viz. infective theory (Leri), developmental disorder (Zimmer and Junghagen), ischaemic theory (Putty) tel- 
angiectatic theory (Lorririer and Moore), etc. Among these theories the one suggesting “developmental dis- 
order” seems to be the most acceptable. Campbell et al2 have also pointed out the various aetiological the- 
ories for this disorder. They clearly point out the non- 
hereditary and congenital origin of the disease. The authors find malformation of the limb bud to be the 
most plausible aetiology of the disorder, and do not 
preclude involvement of the metameric mesoderm, 
some cells of which may have migrated to the limb 
bud. In view of the recorded history of this case, which 
involves differential course in two disorders viz. traum- 
atic and malignant, over a period of 25 years, all that 
can be reasonably said is that there seems to have been 
maldevelopment of the mesoderm involving unilater- 
al localised hypervascularity – mechanisms that have 
been suggested by Lorimier and Moore.5

With the present state of knowledge all that can be 
said with reasonable certainty is that melorheostosis 
is congenital in origin and involves unilateral malde- 
velopment of the mesoderm with an element of hy- 
evascularity. Previous authors have suggested hypervascularity as a causative factor in the development of 
melorheostosis.8,1 The present case has offered two 
events to support this suggestion, first a subcapital frac- 
ture of neck of femur on the affected side that healed 
rapidly and second, early dissemination of an adeno- 
carcinoma of the prostate, which is generally slow- 
growing. Melorheostosis is a benign disorder and com- 
patible with normal life expectancy, as the disease has 
been followed from childhood to the termination of 
life at the age of 58. The early termination may be as- 
cribed to carcinoma of prostate. This case further high- 
lights the need for a close follow-up of patients with 
melorheostosis.

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