Cystic angiomatosis of bone with sclerotic changes mimicking osteoblastic metastases

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Abstract. Five unusual cases of cystic angiomatosis of bone which presented with the radiologic appearance of osteoblastic lesions are reported. Three patients were female (ages 37, 41, and 65 years) and two were male (ages 24 and 66 years). Although cystic angiomatosis of bone usually produces widespread osteolytic lesions with a honeycombed appearance in the skeletal system, multiple osteoblastic lesions mimicking metastatic osteoblastic carcinoma are sometimes seen. This radiological presentation has not been well emphasized in previous reports. Histologically, in addition to the angiomatous lesions, both mature thickened lamellar bone trabeculae and immature trabeculae of woven bone were found. In one of our patients, increasing density of the osteoblastic lesions was noted over time. One previous study has suggested that the age of the lesions of cystic angiomatosis is related to radiographic density. It is important to recognize this uncommon variant of cystic angiomatosis and to include this entity among the radiologic differential diagnoses when multiple osteoblastic lesions are encountered.

Key words: Cystic angiomatosis – Hemangiomatosis – Lymphangiomatosis – Osteosclerosis – Bone

Cystic angiomatosis is a rare disorder in which multiple angiomatous lesions affect the skeleton with or without visceral involvement. The sites of extraskeletal involvement include soft tissue, lung, liver, and, particularly, the spleen. The angiomatous lesions consist of hemangiomas [1–7], lymphangiomatosis [8–13], or a combination of both [14, 15]. Most cases are recognized in the first three decades of life. This condition is usually detected incidentally in radiographs performed for other reasons.

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Radiologically, widespread osteolytic lesions showing a honeycombed appearance in the skeleton are characteristic. The axial skeleton, particularly the skull, spine, ribs, and pelvis are commonly affected; however, peripheral involvement does occur [3, 16]. Although osteoblastic lesions in cystic angiomatosis have been described in several previous reports [6, 17–21], this finding is uncommon, as it is in solitary hemangioma of bone [22]. Its existence is not widely recognized. We report five cases of cystic angiomatosis which showed such osteoblastic lesions mimicking osteoblastic metastases.

Materials and methods

Cases 1, 2, and 3 were encountered in the consultation files of one of us (H.D.D.) and case 5 was found in the consultation files of another author (G.C.S.). Case 4 was retrieved from the files of the Hospital for Joint Diseases Orthopaedic Institute. Plain radiographs and hematoxylin-and-eosin-stained histological sections were available in all cases. Bone scans performed with technetium-99m methylene diphosphonate (⁹⁹mTc-MDP) were available in four cases and computed tomography (CT) scans were available for review in two cases. In one case (case 4), a 13-year follow-up has been carried out with serial radiographic studies and repeat biopsy. In one case (case 3), immunohistochemical studies on paraffin-embedded sections were performed using antibodies for factor VIII-related antigen and cytokeratin.

Results

Clinical features

Clinicopathological data of the five cases are summarized in Table 1. The patients' age ranged from 24 to 66 years with an average of 46.6 years. Three patients were female and two were male.

Two patients complained of back pain for several months. One patient (case 2) had occasional slight pain related to bone and joints for 16 years. Two patients (cases 1 and 5) had back pain that was attributed to lumbar disc disease and fracture of lumbar vertebrae due to
Table 1. Clinicopathological data of patients with sclerosing cystic angiomatosis

<table>
<thead>
<tr>
<th>Case</th>
<th>Age (years)/sex</th>
<th>Symptoms (duration)</th>
<th>Laboratory data</th>
<th>Skeletal distribution</th>
<th>Bone scintigraphy</th>
<th>Visceral involvement</th>
<th>Histology</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>41/F</td>
<td>a Incidental finding</td>
<td>N.A.</td>
<td>Ribs, pelvis, femora</td>
<td>Increased uptake of isotope in chest wall</td>
<td>–</td>
<td>Mixed hemangioma and lymphangioma; sclerotic lamellar bone</td>
</tr>
<tr>
<td>2</td>
<td>37/F</td>
<td>Mild joint pain 16 years ago; initially diagnosed as &quot;osteopoikilosis&quot;</td>
<td>Anemia</td>
<td>Humerus (left), sternum, vertebrae, pelvis, femora</td>
<td>Multiple areas of increased uptake of isotope</td>
<td>Spleen</td>
<td>Predominant hemangioma with lymphangiomatic component; sclerotic lamellar bone and immature woven bone</td>
</tr>
<tr>
<td>3</td>
<td>66/M</td>
<td>Back pain (2 months)</td>
<td>Increased serum alkaline phosphatase; slight increase in total bilirubin, aspartate aminotransferase, and alanine aminotransferase</td>
<td>Ribs, vertebrae, pelvis, femora</td>
<td>Multiple areas of increased uptake of isotope</td>
<td>Lung, parapancreatic soft tissue</td>
<td>Predominant hemangioma with lymphangiomatic component; sclerotic lamellar bone and immature woven bone</td>
</tr>
<tr>
<td>4</td>
<td>24/M</td>
<td>Low back pain (5 months)</td>
<td>Normal</td>
<td>Scapula, ribs, vertebrae, pelvis, sacrum, femora, ? skull</td>
<td>Increased uptake of isotope in right frontal region of skull</td>
<td>–</td>
<td>Hemangioma; sclerotic lamellar bone</td>
</tr>
<tr>
<td>5</td>
<td>65/F</td>
<td>b Incidental finding</td>
<td>N.A.</td>
<td>Pelvis, sacrum, femora</td>
<td>N.A.</td>
<td>–</td>
<td>Hemangioma; sclerotic lamellar bone and immature woven bone</td>
</tr>
</tbody>
</table>

N.A., not available

a Back pain due to lumbar disc disease
b Back pain due to fracture of second and third lumbar vertebrae secondary to osteoporosis

Osteoporosis, respectively. One patient (case 2) had originally been diagnosed 16 years previously at another hospital as having osteopoikilosis on the basis of radiographic findings of osteoclerotic lesions; however, these earlier radiographs were not available for review.

Laboratory studies showed no abnormality in case 4 and no abnormality except for anemia in case 2. In one patient (case 3), who had a past history of hepatitis and amebic dysentery, laboratory studies revealed increased serum alkaline phosphatase (349 U/l) as well as slight abnormality of liver function including total bilirubin (1.6 mg/dl), aspartate aminotransferase (81 U/l), and alanine aminotransferase (66 U/l).

The pelvic bones and proximal femora were the most commonly affected sites and were involved in all cases. The vertebral bodies and ribs were affected in three cases. The humerus, sternum, and scapula were each involved in one case.

Extraskeletal involvement was found in two cases. One case had involvement of the spleen and another had lung and parapancreatic soft tissue involvement.

Radiographic features

Multiple, irregularly distributed osteoblastic lesions were scattered throughout the skeleton in all cases (Figs. 1–4). A variable combination of patchy radiodense lesions and hazy osteosclerotic areas was the most common finding (Figs. 1, 3A,B,D,E). Most patchy radiodensities were 0.5–2 cm in size and some of them showed a tendency toward central lucency. The pattern of nodular sclerotic foci was predominant in case 4 (Fig. 4). Ill-defined diffuse sclerotic lesions with patchy central radiolucencies throughout the pelvis and proximal femora were found in case 2 (Fig. 2A). Although multiple osteoblastic lesions were predominant in all cases, several lesions had associated osteolytic foci, producing a mixed osteolytic and sclerotic appearance (Fig. 3A,B). Despite the presence of purely osteoblastic lesions, the finding of additional lytic foci surrounded by sclerotic zones of varying thickness would favor the diagnosis of angiomatosis over osteoblastic metastases. The lesions of the ribs were expansile and were lytic rather than sclerotic. The degree of osteosclerosis of the lesions increased progressively during the 13-year follow-up in case 4 (Fig. 4A–C).

Bone scintigraphy was done in four cases and increased uptake of isotope corresponded to multiple sclerotic areas in bone in two cases (Fig. 3C). In the other two cases, a single hot spot was demonstrated by bone scan and one of these could be correlated with a lytic focus in a rib. The osteosclerotic lesions in the pelvic bo-