
Extraadrenal Myelolipoma

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A rare case of presacral myelolipoma surgically resected from a 72-year-old woman, who also had megaloblastic anemia and breast carcinoma, is described and compared with mass-forming extramedullary hematopoiesis, and other reported cases of extra-adrenal myelolipoma. Extra-adrenal myelolipoma should be morphologically differentiated from mass-forming extramedullary hematopoiesis. The former is encapsulated or well-circumscribed, is composed of fat cells, and has normal marrow hematopoietic elements. The latter lacks circumscription, and fat is not an integral component of the process. Clinically, myelolipoma is usually asymptomatic, and shows no consistent associated disease process, while mass-forming extramedullary hematopoiesis is usually symptomatic, and is associated with myeloproliferative disorders, hemolytic anemia, or severe skeletal diseases. (Key words: Extraadrenal myelolipoma; Retroperitoneal myelolipoma) Am J Clin Pathol 1982; 78: 386-389.

MOST MYELOLIPOMAS occur in the adrenal gland. Extra-adrenal myelolipomas are rare, with only 15 acceptable cases reported in the literature.1-3,5-5,10,15,16,18-20,23,26 This report describes a surgically resected presacral myelolipoma. The literature is reviewed, and the terminology and differential diagnosis are discussed.

Report of a Case

A 72-year-old woman was found to have a left breast mass in a routine physical examination. An excisional biopsy showed infiltrating ductal carcinoma. She was also found to have macrocytic anemia with a hemoglobin of 8.6 g/dL, a hematocrit of 26%, and an MCV of 116. The serum vitamin B-12 level was normal. The serum folate level was reduced to 2.4 mg/mL (normal range, 4-20 mg/mL). A bone marrow examination showed erythroid hyperplasia with megaloblastic changes. The serum gamma glutamyl transaminase level was 63 U/mL (normal range, 9-38 U/mL). Other serum chemistry values were normal. She also had a history of easy bruisibility for which serum cortisol and ACTH levels were measured. The levels were all within normal range and there was normal diurnal variation. She underwent a metastatic workup which showed no evidence of metastatic lesion.

An unexpected finding of a midline pelvic mass was found in the intravenous pyelography. An exploratory laparotomy was performed and showed a large retroperitoneal fatty mass protruding from the presacral area. The mass elevated the colon out of the pelvis and deviated both ureters laterally. It was fixed to the presacral fascia.

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Pathologic Findings

The excised pelvic mass was well circumscribed and encapsulated. It measured 16 × 15 × 7 cm and weighed 790 g (Fig. 1). It was composed of light yellowish fatty tissue with irregular, brownish areas and irregular, dark red, friable, necrotic areas. Histologic examination showed mature fat cells admixed with cellular foci. The cellular foci were composed of megakaryocytes, red blood cell precursors, and mature and developing granulocytes (Figs. 2 and 3). Numerous sections showed no ectopic adrenal tissue.

The autopsy showed bronchopneumonia with abscess formation, marked intra-abdominal adhesion, left ventricular hypertrophy and benign nephrosclerosis. The adrenal, pituitary and thyroid glands were normal. No residual breast carcinoma or pelvic tumor was found.

Discussion

Most myelolipomas occur within the adrenal glands. Extra-adrenal myelolipoma is rare. A review of the literature revealed only 15 acceptable cases. The clinicopathologic features of these 15 cases and the present case are summarized in Table 1. The male to female ratio was 1:2. The age of the patients ranged from 40 to 81 years, with a mean of 64 years. Three tumors arose in parenchymal organs; two in the liver and one, in the stomach. Three tumors in two cases were located in the subpleural space of the chest wall. In the remaining 11 cases, the tumor arose in the retroperitoneum; 9 in presacral space and one each in the iliac fossa and perirenal space. The tumors ranged from 2.5 to 16 cm in size and weighed up to 790 g. All tumors were sharply circumscribed. With the exception of a hepatic myelolipoma, all tumors were encapsulated. None contained ectopic adrenal tissue. One hepatic tumor produced a slight hepatomegaly and the gastric tumor was associated with gastrointestinal bleeding. The remaining tumors were either incidental findings at autopsy or surgery, or found in physical or radiographic examinations performed for other medical conditions. The significant associated findings included nonmetastasizing carcinoma (three cases), pituitary adenoma (one case), hydromecephalus...
Table 1. Reported Cases of Extraadrenal Myelolipoma

<table>
<thead>
<tr>
<th>Cases</th>
<th>Authors</th>
<th>Age (yr)</th>
<th>Sex</th>
<th>Location and Size</th>
<th>Clinical Presentation</th>
<th>Associated Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Saleeby, 1925&lt;sup&gt;26&lt;/sup&gt;</td>
<td>81</td>
<td>F</td>
<td>Parietal pleura near the ribs, 2 tumors, 2.5 cm</td>
<td>Incidental finding at autopsy</td>
<td>Arteriosclerotic cardiovascular disease, pituitary adenoma</td>
</tr>
<tr>
<td>2</td>
<td>Blaisdell, 1933&lt;sup&gt;5&lt;/sup&gt;</td>
<td>64</td>
<td>F</td>
<td>Presacral, 11 cm, 250 g</td>
<td>Pelvic mass</td>
<td>Pyelonephritis, cystitis</td>
</tr>
<tr>
<td>3</td>
<td>Lyall, 1935&lt;sup&gt;35&lt;/sup&gt;</td>
<td>54</td>
<td>F</td>
<td>Presacral, fist size</td>
<td>Incidental finding at autopsy</td>
<td>Pernicious anemia</td>
</tr>
<tr>
<td>4</td>
<td>Dodge &amp; Evans, 1956&lt;sup&gt;10&lt;/sup&gt;</td>
<td>74</td>
<td>F</td>
<td>Presacral, 15 cm, 570 g</td>
<td>Pelvic mass</td>
<td>Paget’s disease</td>
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<tr>
<td>5</td>
<td>Foster, 1958&lt;sup&gt;12&lt;/sup&gt;</td>
<td>80</td>
<td>M</td>
<td>Intrathoracic paravertebral, 4 cm</td>
<td>Incidental finding at autopsy</td>
<td>Pneumonia</td>
</tr>
<tr>
<td>6</td>
<td>Benson &amp; Janko, 1965&lt;sup&gt;9&lt;/sup&gt;</td>
<td>52</td>
<td>F</td>
<td>Presacral, 6 cm</td>
<td>Palpable pelvic mass</td>
<td>None</td>
</tr>
<tr>
<td>7</td>
<td>Grosdidier, 1973&lt;sup&gt;13&lt;/sup&gt;</td>
<td>63</td>
<td>F</td>
<td>Right lobe of liver, 12 cm</td>
<td>Slight hepatomegaly</td>
<td>Obesity, diabetes mellitus, gout</td>
</tr>
<tr>
<td>8</td>
<td>LeBodic and associates, 1974&lt;sup&gt;20&lt;/sup&gt;</td>
<td>60</td>
<td>?</td>
<td>Gastric antrum, 4 cm</td>
<td>Gastrointestinal bleeding</td>
<td>—</td>
</tr>
<tr>
<td>9</td>
<td>Beraha and associates, 1974&lt;sup&gt;7&lt;/sup&gt;</td>
<td>64</td>
<td>M</td>
<td>Perirenal soft tissue, 13 cm</td>
<td>Incidental finding in arteriography for arteriosclerosis</td>
<td>—</td>
</tr>
<tr>
<td>10</td>
<td>Ishak, 1976&lt;sup&gt;16&lt;/sup&gt;</td>
<td>53</td>
<td>F</td>
<td>Left lobe of liver, 2 cm</td>
<td>Incidental finding at autopsy</td>
<td>—</td>
</tr>
<tr>
<td>11</td>
<td>Labow and associates, 1977&lt;sup&gt;19&lt;/sup&gt;</td>
<td>47</td>
<td>F</td>
<td>Presacral</td>
<td>Melena, mass found in barium-enema examination</td>
<td>Viral pericarditis treated with steroid</td>
</tr>
<tr>
<td>12</td>
<td>Farman and associates, 1978&lt;sup&gt;11&lt;/sup&gt;</td>
<td>72</td>
<td>M</td>
<td>Presacral</td>
<td>Lower abdominal pain, mass found in barium-enema examination</td>
<td>Emphysema, arteriosclerotic heart disease</td>
</tr>
<tr>
<td>13</td>
<td>Kapadia &amp; Kanbour, 1978&lt;sup&gt;18&lt;/sup&gt;</td>
<td>40</td>
<td>F</td>
<td>Presacral, 9 cm</td>
<td>Mass found in rectal examination</td>
<td>Anemia due to menometrorrhagia</td>
</tr>
<tr>
<td>14</td>
<td>DiBonito and associates, 1979&lt;sup&gt;9&lt;/sup&gt;</td>
<td>75</td>
<td>M</td>
<td>Right iliac fossa, 16 cm</td>
<td>Acute urinary retention</td>
<td>Anemia, hydronephrosis carcinoma of kidney, colon and prostate</td>
</tr>
<tr>
<td>15</td>
<td>Allen, 1981&lt;sup&gt;1&lt;/sup&gt;</td>
<td>77</td>
<td>M</td>
<td>Presacral, 13 cm</td>
<td>Mass found in cholecystectomy</td>
<td>None</td>
</tr>
<tr>
<td>16</td>
<td>Chen and associates, present report</td>
<td>73</td>
<td>F</td>
<td>Presacral, 16 cm, 790 g</td>
<td>Incidental finding in pyelography</td>
<td>Breast carcinoma, megaloblastic anemia</td>
</tr>
</tbody>
</table>

or pyelonephritis (two cases), mild Paget’s disease of the bone (one case), pernicious anemia or megaloblastic anemia (two cases), viral pericarditis with steroid treatment (one case), and anemia secondary to menorrhagia (one case).

The tumor resembles the adrenal myelolipoma<sup>24</sup> in its microscopic components. It is composed of an admixture of fat cells and hematopoietic cells including mature and developing myeloid, erythroid and megakaryocytic cells. The proportion of mature and immature hematopoietic cells varies from case to case. Unlike the adrenal myelolipomas, all extra-adrenal myelolipomas are sharply circumscribed and all soft tissue myelolipomas are encapsulated. Nests of adrenal cells are usually present within the adrenal myelolipomas, but not in the extra-adrenal tumors.

In the literature, the extra-adrenal myelolipomas were often confused with the more common mass-forming extramedullary hematopoiesis. These two terms were frequently used interchangeably. Our review indicates that these are two distinct clinicopathologic entities. They can and should be clearly separated in future reports. Clinically mass-forming extramedullary hematopoiesis represents a complication of myeloproliferative disorders,<sup>14,21</sup> severe skeletal diseases,<sup>17,27,30</sup> or severe anemia secondary to hemoglobinopathy, red-cell enzyme deficiency or carcinomatosis.<sup>7,22,25,29</sup> It involves various parenchymal organs and the soft tissue. Of the
soft tissue, the thoracic paravertebral space is the preferred site of involvement and the lesions often produce neurologic symptoms. In contrast, the extra-adrenal myelolipomas show no consistent association with other diseases. The retroperitoneal space, especially the presacral area, is the usual location for the lesions. The tumor is usually asymptomatic. Pathologically mass-forming extramedullary hematopoeisis is poorly circumscribed and nonencapsulated, while extra-adrenal myelolipomas are sharply circumscribed and, when involving the soft tissue, are encapsulated. In addition, the fat cell is not an integral part of extramedullary hematopoeisis.

As in the adrenal myelolipoma, the etiology of extra-adrenal myelolipoma is uncertain. The occasional associations of adrenal myelolipoma with Cushing's disease and congenital adrenal hyperplasia, the relatively frequent occurrence of the tumors in burn and cancer patients, and the production of myelolipoma in rats by injection of crude adrenocorticotropic hormone and testosterone suggest that prolonged stimulation with adrenocorticotropic hormone or cortisol may be etiologically related to myelolipoma. Of the 15 patients with extra-adrenal myelolipoma, one had pituitary adenoma, three had carcinomas and one received steroid treatment. These observations raised the possibility of excessive pituitary ACTH or ectopic ACTH production in these patients. However, in the present case the serum cortisol and ACTH studies were all normal, and the steroid treatment received by one patient was of short duration. The megaloblastic anemia secondary to folic acid deficiency documented in the present case, and the associated anemia in two other cases suggest that prolonged erythropoietin stimulation may play an etiologic role in some patients. Detailed hematologic and endocrinologic studies in future cases may reveal the pathogenesis of extra-adrenal myelolipoma.

References