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Case Report

Giant Extra-Adrenal Retroperitoneal Myelolipoma With Incidental Gastric Mesenchymal Neoplasias

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Extra-adrenal myelolipomas are rare, benign tumors composed of adipose tissue and hematopoietic cells. Almost all myelolipomas occur within the adrenal gland. Only 50 cases of myelolipomas were described in literature and none of these were associated with gastric mesenchymal neoplasia. A 72-year-old male patient presented to a family medicine outpatient clinic with dyspnea and urinary urgency. His abdominal sonography revealed a 9-cm intra-abdominal mass. An incidental finding was 2 separate masses 1 cm each on the serosal surface of the stomach. The pathology specimen of the retroperitoneal mass revealed myelolipoma histopathology while gastric masses were reported as spindle cell mesenchymal neoplasias. The association of gastric spindle cell tumor and myelolipoma was not reported before in medical literature. Extra-adrenal myelolipomas are rare lesions, but should be considered in the differential diagnosis of fat containing retroperitoneal masses that are well circumscribed.

Key words: Myelolipoma – Retroperitoneal – Neoplasia

Extra-adrenal myelolipomas are rare, benign tumors composed of adipose tissue and hematopoietic cells. When these lesions occur outside the adrenal gland, they can be confused with other soft tissue tumors. Differentiating extra-adrenal myelolipomas from well-differentiated liposarcomas, retroperitoneal myelolipomas, and extramedullary hematopoietic tumors can be difficult. There are only 50 cases of myelolipoma described in literature[1] and none of these is associated with gastric mesenchymal neoplasia.

Here we present a symptomatic extra adrenal retroperitoneal myelolipoma presenting along with incidental multiple gastric stromal tumors.

Case

A 72-year-old male patient presented to a family medicine outpatient clinic with dyspnea and uri-

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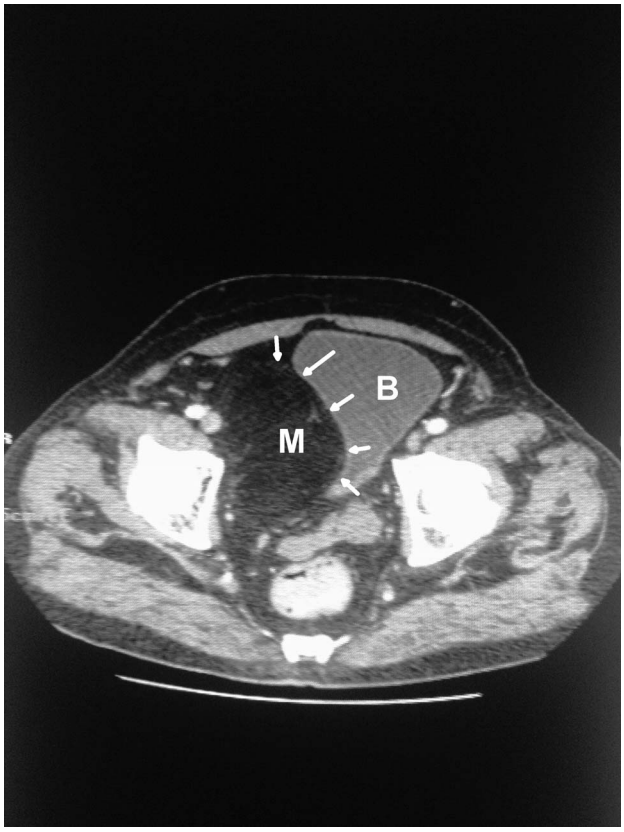

Fig. 1 Computed tomography showing the myelolipoma (M) and its indentation (small arrows) on the bladder (B).

nary urgency. His work-up included abdominal sonography which revealed a 9-cm intra-abdominal mass that resulted in referral to a general surgery clinic. His laboratory work-up did not reveal any significant finding other than an elevated CA 19-9 level of 48.47 U/mL (Range: <27). A computed tomography of the abdomen showed a 10-cm mass inside his pelvis impinging on the bladder and external iliac vein on the right side (Fig. 1). Surgical exploration was undertaken and it was seen that the mass was retroperitoneal, pushing the external iliac vein laterally but not invading it, neighboring the bladder and overlying the obturator nerves. An incidental finding was 2 separate masses 1 cm each on the serosal surface of the stomach. After careful dissection, the retroperitoneal mass was removed en bloc (Fig. 2) without any damage to the surrounding structures. The gastric masses were resected with a wedge of gastric tissue. The postoperative course of the patient was uneventful and he was discharged on postoperative day 4. The pathology specimen of the retroperitoneal mass revealed myelolipoma

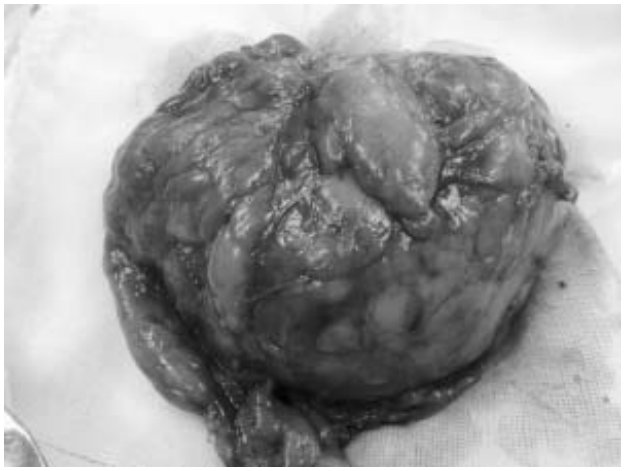

Fig. 2 Resected myelolipoma.

histopathology while gastric masses were reported as spindle cell mesenchymal neoplasias.

Discussion and Review of Literature

Myelolipomas are well-circumscribed lesions that contain various combinations of myeloid and erythroid precursors mixed with mature adipose tissue. Myelolipomas also may contain areas of internal hemorrhage which may calcify. Almost all myelolipomas occur within the adrenal gland. Myelolipomas manifest in 4 distinct clinicopathologic patterns: isolated adrenal myelolipoma, adrenal myelolipoma with hemorrhage, extra-adrenal myelolipoma, and myelolipoma associated with other adrenal diseases. Only 50 cases of extra-adrenal myelolipomas have been reported in the literature.[1] These tumors, which are usually present in the presacral retroperitoneum, also have been reported in the perirenal retroperitoneum, mediastinum, liver, stomach, and muscle fascia.[2] The median age for diagnosis of extra-adrenal myelolipoma is 66.5 years.[3] The cause of myelolipomas is unknown. Theories include embolization of bone marrow tissue and differentiation of primitive hematopoietic stem cell rests in response to a triggering stimulus such as endocrine dysfunction.[4] Extra-adrenal myelolipomas predominate in females (2:1) and are found in the middle aged to the elderly (mean age, 61 years).[5]

The natural history of extra-adrenal myelolipoma is unknown. Extra-adrenal myelolipomas range from 2 to 26 cm in greatest dimension. Smaller lesions are typically asymptomatic. Large tumors may cause symptoms from mass effect or hemorrhage. Malignant degeneration has not been reported. Extra-

adrenal myelolipomas are difficult to diagnose preoperatively and radiographic differentiation between this entity and other fat containing retroperitoneal tumors can be difficult. Most fat-containing retroperitoneal tumors are well-differentiated liposarcomas. Unlike extra-adrenal myelolipomas that are usually well-encapsulated liposarcomas tend to be poorly marginated and infiltrative. Histologically, well-differentiated liposarcomas are not hemorrhagic. Moreover, liposarcomas have lipoblasts and zones of cellular atypia, features that are not present in extra-adrenal myelolipomas.

Other than liposarcoma, there are a number of entities that should be differentiated from myelolipoma. One of these is extramedullary hematopoiesis (EMH), which is the development of blood cells outside the bone marrow. Extramedullary hematopoiesis can be found within neoplasms involving both hemic and nonhemic tissues.[6] Microscopically, extramedullary hematopoiesis is composed of hematopoietic cells and erythroid hyperplasia. The lesion is typically devoid of lymphoid cell aggregates and there is little to no adipose tissue, unlike myelolipoma.[7]

The second entity is primary retroperitoneal teratoma that accounts for 1 to 11% of all retroperitoneal neoplasms. Retroperitoneal teratomas are mostly located near the upper pole of the left kidney. The most characteristic features of mature teratoma of the retroperitoneum are a mass that contains a fluid component of variable volume, adipose tissue, sebum and calcifications.[8,9] The third entity is mesenchymal tumors, but as teratomas, these also also contain other tissue subtypes as well.[5]

If suspected preoperatively, a needle biopsy can establish a diagnosis and an asymptomatic patient can avoid surgery. In our patient, the large size of the extra-adrenal myelolipoma and his associated symptoms necessitated resection of the tumor.

The role of preoperative biopsy for any tumor that is presacral in location is debated. Traditionally, many surgeons consider presacral biopsy a contraindication in any lesion that is resectable due to the risks of seeding the biopsy needle tract with potentially malignant cells and infecting the lesion. Others suggest that a preoperative biopsy should be considered, as the pathologic results have the ability to strongly impact management, particularly for lesions that are large or invade adjacent structures.[11]

Concurrent presentation of myelolipomas with other neoplasms (adrenal adenoma, focal nodular hyperplasia, non-Hodgkin lymphoma) are reported in literature.[12] An incidental finding in our patient was 2 spindle cell tumors on the gastric surface. This association was not reported before in medical literature.

We conclude that extra-adrenal myelolipomas are rare lesions, but should be considered in the differential diagnosis of a fat-containing retroperitoneal mass that is well circumscribed. Additionally, a secondary neoplasm should not be missed.

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