HEPATIC AND RENAL ANGIOMYOLIPOMA - A RARE CLINICAL ENTITY

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ABSTRACT

Hepatic and renal angiomyolipoma is a rare clinical condition to diagnose radiologically. We are presenting a case with typical radiological features on CT scan, which were later confirmed by the histopathological examination. Our case has involvement of both liver and kidneys with tuberous sclerosis of dorsolumbar vertebrae. The typical features are highlighted in our case report.

Key words: Angiomyolipoma, Tuberous sclerosis, CT scan, Liver and Kidney

Introduction

Angiomyolipoma (AMLs) is a rare benign tumor of kidney and less commonly found in the liver and rarely in other organs. It represents 1% - 3% of all renal tumors and about 80 - 90% of cases are sporadic and these are most commonly found in middle-aged women.[1] Multiple AMLs are extremely rare and are typically seen in patients associated with tuberous sclerosis. An autopsy study and tuberous sclerosis clinic survey found prevalence of 67% and 85% respectively for patients with tuberous sclerosis. Both genders are affected equally.[2] AMLs are composed of abnormal vasculature, smooth muscle, and adipose cells. Renal AML presents with flank pain, a palpable mass and gross hematuria.

Case Report

A 22 year- old man presented to us with a history of abdominal pain and dyspepsia for the last one year. He had no significant medical or surgical history. His ultrasound abdomen revealed multiple hyperechoic lesions in the right lobe of liver, one of them measured 1.9 cm. His both kidneys were enlarged with multiple hyperechoic areas bilaterally, largest one measured 3.4 cm in right kidney. No calculus or hydronephrosis was seen on either side. No lesion was seen in other solid organs.

His CT scan abdomen was also done for further evaluation which revealed multiple hypodense areas of fat density in the right lobe of the liver, one of them measured 2.2 cm in the right lobe of the liver. Both kidneys were enlarged in size, multiple hypodense and fat density areas were seen in both kidneys, one of them on right side measured 3.9 cm. Small sclerotic lesions were also seen in the dorso-lumbar vertebrae. No calculus or hydronephrosis was seen in either kidney. Rest of the scan was unremarkable.

On the bases of above investigation we suspected the case as Hepatic and Renal Angiomyolipoma associated with tuberous sclerosis but for further confirmation we did histopathological examination by ultrasound guided trucut biopsy from kidney and liver, which revealed spindled and histiocytoid shape tumor cells with slightly eosinophilic cytoplasm.The tumor cells were arranged alongside the vessels and scattered among the inflammatory background. Sinusoid structure was also seen in the tumor. Mature adipocytes and thick-walled blood vessels were focally obser-

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ved at the boundaries between the tumor and surrounding liver tissues. The tumor cells were positive on immunostaining for HMB-45.The diagnosis of hepatic and renal Angiomyolipoma was made.

Discussion

Angiomyolipoma is a rare benign mesenchymal tumor composed of a mixture of smooth muscle cells (myoid), blood vessels (angioid), and a variable amount of fat (lipoid). The appearance of an angiomyolipoma has often been reported in the kidney and occurs at least nine times more commonly in the kidney than any other organ.[3]

AMLs may also occur, however, in extra-renal sites, such as the liver, spleen, abdominal wall, retroperitoneum, uterus, oral cavity, penis, spermatic cord, fallopian tube, vagina, skin, and lung. The renal AML is seen in normal individuals, but occurs more frequently in patients with the genetic disease tuberous sclerosis, where incidence in adults is around 70%. Renal AMLs of TSC patients are found in both sexes, in the third and fourth decades of life, with predominance in females.

The first hepatic angiomyolipoma was reported by Ishak in 1976[4] a very rare benign neoplasm. It's incidence in the general population was 0.3%.[3] The natural history of hepatic angiomyolipoma is unknown. The course of patients with un-resected renal angiomyolipoma has been much better documented. Steiner et al demonstrated tumor growth in 27% of renal angiomyolipomas measuring less than 4 cm in diameter and in 46% of those greater than 4 cm over a 4-year period. Patients with lesions less than 4 cm in diameter were more likely to be asymptomatic than were individuals with tumors 4 cm or larger.[5]

Although hepatic AML has various types or variants and mimics various hepatic neoplasm, it can still be recognized or suspected on morphologic grounds. The clues to the diagnosis are the three characteristic components (blood vessels, smooth muscle, and fat tissue) and diagnostic myoid component which may exist in epithelioid, spindle, and intermediate forms. It is most frequently diagnosed on ultrasound which shows an echogenic, smoothly contoured lesion, with a well-defined border that separates it from adjacent normal hepatic tissue or CT scan where the density of the lesion varies and depends on the relative properties of the tissue components, on the bases of high fat content AML can be differentiated from a malignant one, as hepatocellular carcinoma and some metastatic lesions can rarely contain a significant amount of fat.[6]

The histologic patterns described in the literature include lipomatous, myomatous, angiomatous, trabecular, pelioid, inflammatory and mixed pattern.[7] It has been speculated that the distinctive epithelioid cells are primitive mesenchymal cells with an ability to differentiate toward both myoid and adipose cells. Immunohistochemically, these cells are strongly positive for HMB-45 and smooth SMA.

Surgical resection should be considered for all symptomatic patients. Conservative management with close follow-up is suggested in patients with asymptomatic tumors, good compliance and absence of hepatitis virus infection, as well as small hepatic angiomyolipoma (< 5 cm) that are diagnosed through fine-needle aspiration biopsy.[8]

Conclusion

We concluded that angiomyolipoma should be suspected in those cases with multiple hyperechoic lesions in liver as well as kidneys and clinical suspicion of tuberous sclerosis. CT scan of the abdomen with multiple hypodense lesions with fat density in liver and kidney with tuberous sclerotic lesions in vertebrae is the hall mark of angiomyolipoma.

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