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Case Series

Hepatic Angiosarcoma: Radiological Difficulties of a Rare Vascular Tumor

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Abstract

Introduction: Hepatic angiosarcoma accounts for less than 2% of primary liver tumors. Exposure to carcinogenic factors is classically described as responsible for the development of this tumor in 25% of cases.

Clinical Cases: We report three (3) clinico-radiological observations of 3 patients, including one female with hepatomegaly with signs of portal hypertension and the others male with multinodular hepatosplenomegaly. The lesions reported are multiple, polymorphic in appearance and hyper vascular for some. The pathological results are in favor of hepatic angiosarcoma.

Conclusion: Hepatic angiosarcomas are rare tumors with nonspecific clinical signs. Some radiological aspects must be understood in order to approach the diagnosis before the pathological step. The latter remains the only means of diagnostic confirmation before possible surgery, in case the tumor is resectable.

Keywords: *Angiosarcoma; Liver; CT*

Introduction

Angiosarcoma is a subtype of soft tissue sarcoma, arising either from the endothelium, lymphatic vessels or blood [1]. It represents 4% of angiosarcomas all localizations combined and less than 2% of primary liver tumors [2,3]. Although symptoms are generally nonspecific, abdominal distension, weight loss and signs of hepatocellular failure are generally found [4]. As angiosarcoma of the liver is a hypervascular tumor, its radiological differential diagnosis from other hepatic vascular tumors, such as adenoma, remains difficult [1].

Observations

Case 1

This is a 42-year-old patient admitted for an increase in abdominal volume, asthenia and weight loss progressing for 4 months. The initial clinical examination found a conscious patient who was stable, afebrile, anicteric, with declining dullness of the flanks and hepatomegaly. The ultrasound performed revealed a liver with chronic multi-nodular hepatopathy with signs of portal hypertension and ascites of great abundance. The Blood tests indicated correct renal function, hemoglobin at 10 g/dl, leukocytes at 8000/mm^3, platelets at 98000/

mm^3, HBsAg negative, AC anti HCV negative, AFP at 2.99 and prothrombin level at 44%. The abdominal CT scan found a heterogeneous liver, with several well-circumscribed lesions of the left liver, with an intense nodular petechial enhancement in the arterial phase and in the portal phase but however other lesions presented an annular contrast enhancement in the arterial phase with ascites (Figure 1). Thus, given the polymorphism of the lesions, a percutaneous biopsy puncture was performed, followed immediately by percutaneous embolization of the biopsy path with a gelatin emulsion. Histological examination of the biopsy cores revealed a malignant tumor proliferation, made up of irregular vascular cavities, forming an anastomotic network and dissecting the adjacent hepatic parenchyma, bordered by atypical endothelial cells, large in size, provided with abundant eosinophilic cytoplasm and nuclei voluminous vesicular strongly nucleoli. The immunohistochemical study carried out showed strong expression of vascular markers (CD31 and CD34 suggesting hepatic angiosarcoma. The patient died after 8 months.

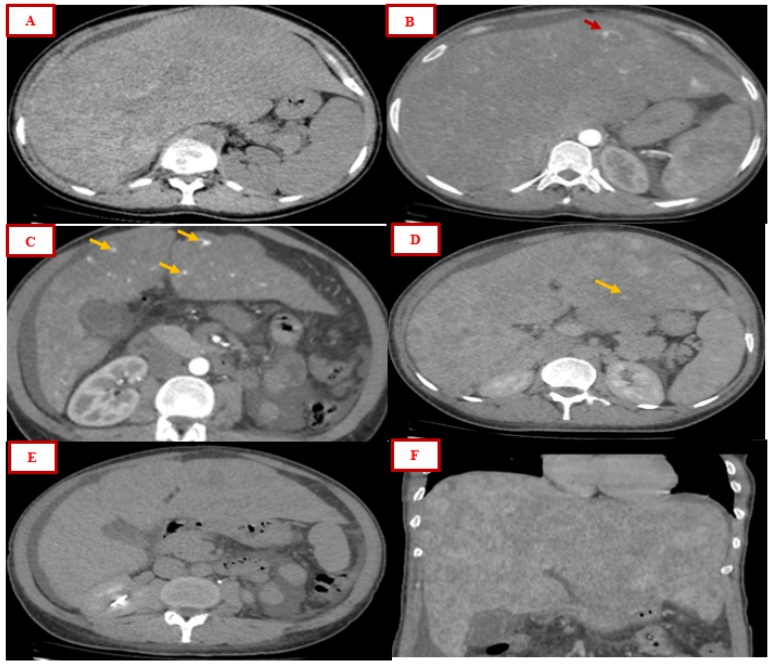

Figure 1: *CT axial sections without contrast (A), with contrast in arterial (B and C), portal (D), Tardif (E) and coronal Tardif (F). Heterogeneous dysmorphic liver, with several well-circumscribed lesions of the left liver, with intense nodular petechial enhancement in the arterial phase and in the portal phase (yellow arrows) but however other lesions presented an annular enhancement (red arrow) at the time arterial with ascites.*

Case 2

This is a 44-year-old patient referred for abdominal distension and found on ultrasound to multi-nodular hepatosplenomegaly and ascites. Clinical examination found a conscious patient who was stable, asthenic, afebrile and anicteric with declining dullness of the flanks.

The investigations found correct renal function, hemoglobin at 8 g/dl, leukocytes at 6000/mm^3, platelets at 58000/mm^3, viral serologies were negative, and prothrombin level at 35%. The abdominal CT scan found an enlarged liver of heterogeneous size, site of multiple hypodense lesions with petechial, annular and nodular contrast enhancement with a globular spleen also seat of multiple lesions present-

ing the same characteristics as the hepatic ones (Figure 2). Histological examination revealed a malignant tumor proliferation, made up of irregular vascular cavities, and immunohistochemical study showed strong expression of the vascular markers CD31 and CD34. The patient's consequences ended in death after a year.

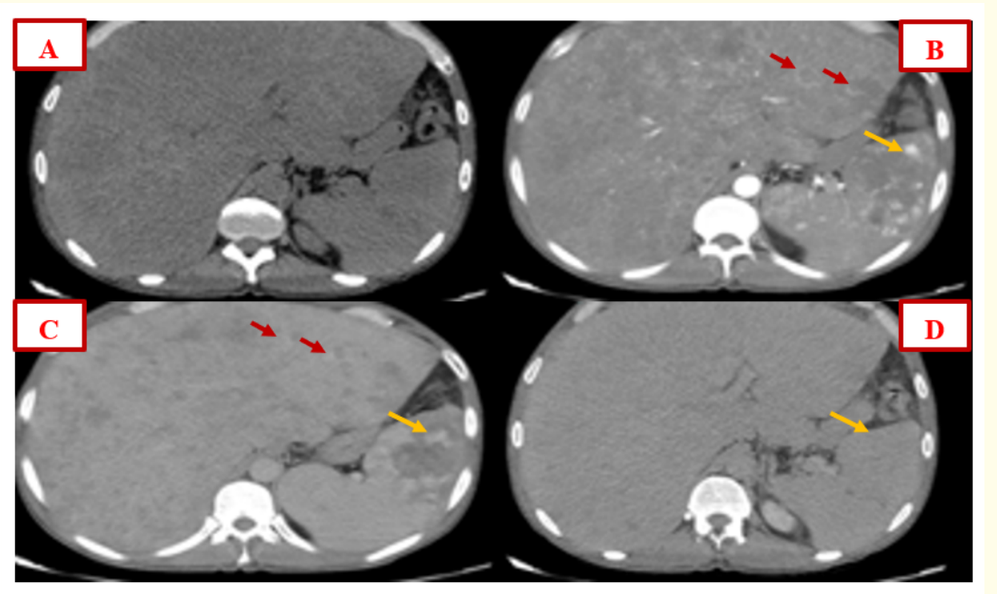

Figure 2: *CT axial sections without contrast (A) and with contrast in the arterial (B) portal (C) and late (D) phase. Increased liver of heterogeneous size, site of multiple hypodense lesions with annular contrast enhancement (red arrows) and lump (yellow arrows) with a globular spleen also site of multiple lesions presenting the same characteristics as hepatic ones.*

Case 3

This is a 50-year-old patient referred for abdominal pain and found on ultrasound to have multiple liver and splenic lesions. Clinical examination found a conscious patient to be stable, with general condition maintenance. The Blood tests found correct renal function, hemoglobin at 10.5 g/dl, leukocytes at 5000/mm^3, platelets at 110,000/mm^3, viral serologies were negative, and prothrombin level at 42%. The abdominal CT scan found a heterogeneous liver, site of multiple hypodense lesions with petechial contrast uptake, nodular for some and clodded for others, a spleen with lesions presenting the same characteristics as those of the liver, adrenal lesions bilaterally as well as osteolytic and osteocondensing lesions involving the vertebral bodies (Figure 3). The pathological examination carried out objectified a malignant tumor proliferation, made up of vascular cavities correlated with a strong expression of vascular markers by immunohistochemistry. The course was marked by death in less than six (6) months.

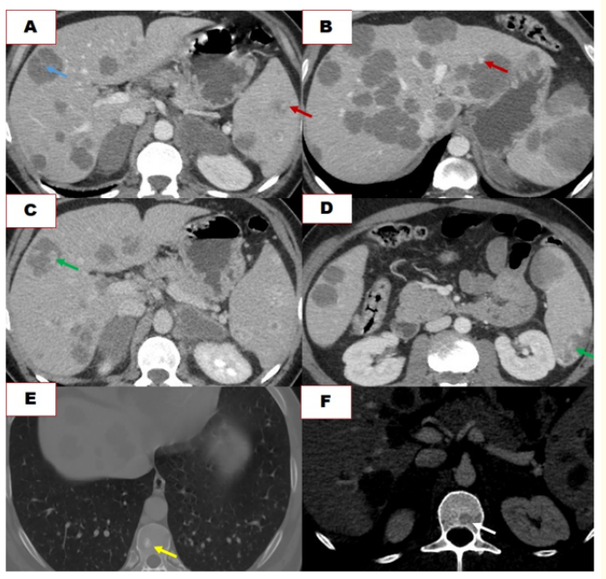

Figure 2: *Axial CT slices in the portal phase and with bone window (E). Heterogeneous liver and spleen, site of multiple hypodense lesions with petechial (blue arrow), nodular (red arrows), and root ball (green arrows) contrast enhancement, Bilateral adrenal lesions (orange arrows), Bone lesions of the osteolytic vertebral bodies (white arrow), and osteocondensing (yellow arrow).*

Discussion

Angiosarcoma is a rare vascular tumor occurring at a median age of 60 years, with a male predominance and a sex ratio of 4/1 [1]. Studies have shown a link between angiosarcoma of the liver and exposure to chemicals in the workplace or the environment. Three substances are currently known to cause angiosarcoma of the liver including vinyl chloride monomer, thorium dioxide and inorganic arsenic. In addition to these substances, anabolic androgenic steroids, estrogen and oral contraceptives may also cause angiosarcoma of the liver [5]. No exposure link with these substances could be identified in our patients. The revealing clinical manifestations are abdominal pain, weight loss, hepatomegaly, whether or not associated with splenomegaly and ascites [5]. Our patients presented with abdominal distension, hepatomegaly, splenomegaly, weight loss and ascites for some. Hepatic angiosarcoma is often associated with thrombocytopenia, disseminated intravascular coagulation or hemolytic anemia [2,6].

Which is consistent with our cases who had thrombocytopenia and hemolytic anemia. Radiologically, angiosarcoma is either single (40%) and most often large, or multi-nodular (50%) or diffuse (10%). The nodules are hypodense and heterogeneous before injection with a progressive contrast price [2]. CT and angiography to observe that the tumor takes up contrast material around the periphery and in puddles [7]. While all imaging techniques can in most cases differentiate hemangiomas, hypervascular metastases and HCC, the differential diagnosis between the latter two lesions and angiosarcoma may be impossible. There are frequently metastatic lesions at the time of diagnosis, preferably localized at the splenic and pulmonary level [7]. A precautious percutaneous biopsy can be performed for pathological confirmation. On the CT scan our patients presented for the first, a heterogeneous liver, with several well-defined lesions of the left liver, with an intense nodular petechial enhancement in the arterial phase and in the portal phase (arrow) but however other lesions presented a ring contrast at arterial time. On the other hand, the others presented an enlarged liver of heterogeneous size, site of multiple hypodense lesions of polymorphic appearance with petechial contrast enhancement, nodular annular and root ball with a spleen presenting lesions having the same characteristics as those hepatic. In one of the latter we found metastatic osteolytic and osteocondensing lesions. These polymorphic radiological aspects show the non-specificity of radiological semiology. Benign lesions such as forms of diffuse hepatic angiomatosis, multiple hepatic adenomas, multiple focal nodular hyperplasias, or multiple angiomyolipomas can lead to this malignant tumor. In contrast, other malignant lesions may be offered in the range of radiological diagnoses such as infiltrating hepatocellular carcinomas, secondary locations of neuroendocrine tumors, and metastases of hypervascular tumors. Thus, the recourse to a percutaneous biopsy with a histological study is inevitable. The survival of patients with hepatic angiosarcoma is very poor with a median survival of 6 months without treatment. Even after treatment, only 3% of patients reported living longer than two years [4]. The possibility of complete surgical resection is rarely achieved due to diagnostic delay in most cases [4].

Conclusion

In short, it appears that hepatic angiosarcoma is a tumor with non-specific clinical signs and polymorphic radiological appearances. The use of anatomopathology is unavoidable and represents the means of specific exploration for diagnosis.

Conflicts of Interest

The authors declare no conflict of interest.

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