Large Telangiectatic Focal Nodular Hyperplasia Presenting with Normal Radionuclide Studies: Case Report

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A 9 cm-lesion of telangiectatic focal nodular hyperplasia was incidentally identified in a 31-yr-old female. Despite a typical appearance by X-ray computed tomography and ultrasonography, scintigraphy with technetium-99m-(^{99m}Tc) colloid, ^{99m}Tc-diethyliminodiacetic acid, and ^{99m}Tc-labeled red cells failed to demonstrate any abnormalities. These findings are felt to reflect the relative lack of architectural disruption that histologically characterizes this particular lesion. The present report described the imaging characteristics of the telangiectatic form of focal nodular hyperplasia.

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The typical solid lesion of hepatic focal nodular hyperplasia (FNH) is hypervascular, nonencapsulated, and often lobulated with a heterogeneous internal structure on ultrasonography (US) and X-ray computed tomography (CT). A stellate central vascular scar is commonly present (*1,2*). Technetium-99m-colloid (^{99m}Tc-SnC) uptake within the lesion varies from absent to normal relative to surrounding liver tissue. The most common hepatobiliary manifestation with ^{99m}Tc-iminodiacetic acid (^{99m}Tc-IDA) analogs is increased transit through the lesion in the perfusion phase, normal or enhanced hepatocyte uptake, and prolonged retention during washout; although the degree of concentration can also be absent or decreased (*3–6*). A telangiectatic form of FNH has recently been distinguished pathologically (*7*). In a search of the literature, however, we were unable to find any description of its imaging characteristics.

In the present report we describe a 9-cm lesion of telangiectatic FNH that was easily disclosed by CT, US, and angiography, but indistinguishable from adjacent normal liver tissue with respect to ^{99m}Tc-SnC, ^{99m}Tc-diethyl-IDA, and ^{99m}Tc-labeled red cell perfusion and blood-pool imaging. Pathologic correlation is provided.

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CASE REPORT

A 31-yr-old female was incidentally found to have elevated serum liver enzymes (alkaline phosphatase = 632 U/l, gammaglutamyl transferase = 142 U/l, bilirubin = 6.0 μmole/l) during examination for chronic headache and recurrent epistaxis. CT revealed a 9-cm, peripheral, well marginated, hypodense mass in the right lobe of the liver, demonstrating heterogeneous enhancement with intravenous contrast. No other lesions were identified. Sonography of the liver confirmed the mass, and duplex Doppler analysis demonstrated a high velocity, very low impedance, intralesional blood flow that is often seen in ectatic sinusoidal vessels associated with certain tumors (*8*). On hepatic arteriography, the lesion received a hypervascular supply from the right hepatic artery, but relatively little portal flow (Fig. 1). Numerous tortuous and ectatic arteries were visualized within the lesion. The tumor demonstrated normal uptake of ^{99m}Tc-SnC (Fig. 2A), and normal uptake and excretion of ^{99m}Tc-IDA (Fig. 2B). Despite abnormal findings on angiography, a ^{99m}Tc-labeled red blood cell (RBC) perfusion and blood-pool study was normal (Fig. 2C). A right hepatectomy was performed. The mass was firm and showed areas of ectatic sinusoids lined with endothelial cells and Kupffer cells on histology (Fig. 3). Portal spaces contained large muscular arteries and hyperplastic bile ducts, but liver cell plates were relatively normal. Apart from a few necrotic hepatocytes associated with some inflammatory cells and minimal fibrosis in the areas of most extensive sinusoidal dilatation, very little architectural distortion was present. Additional lesions were not identified.

DISCUSSION

Focal nodular hyperplasia is a benign liver tumor of unknown etiology usually found incidentally in young adult females. Next to hemangioma, it is the most common benign tumor of the liver, and has an estimated prevalence of 0.8% (*7*). Multiple lesions are present in 20%–30% of cases (*7,9,10*). FNH is typically asymptomatic, and invariably follows a benign course (*1,11*). Accordingly, the preferred management is conservative, and excision is generally reserved for large symptomatic lesions, or when the diagnosis remains uncertain. This is in contrast to hepatocellular adenoma (HCA), a less common benign liver tumor, which is associated with a 30%–75% incidence of necrosis and

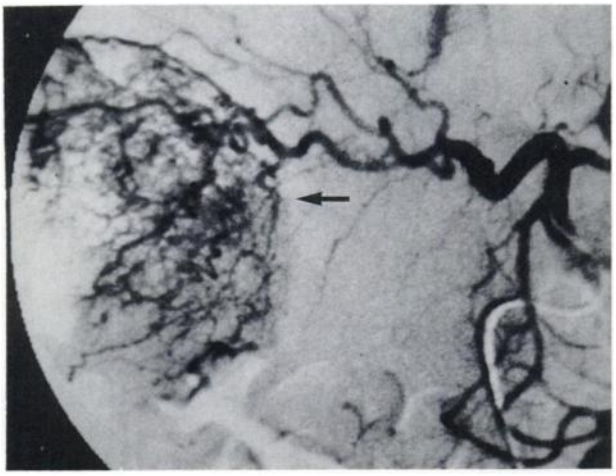

FIGURE 1
Contrast hepatic arteriogram with digital subtraction demonstrated hypervascular supply to the lesion from the right hepatic artery. Numerous tortuous and ectatic arteries were visible in the mass (arrow), but relatively few portal vein radicles were present.

potentially life-threatening hemorrhage, and accordingly treated by surgical excision (*1,11*). Differentiation of these two lesions by diagnostic imaging is not always possible, and surgery is often unavoidable. Although complications of untreated FNH are exceedingly rare, deaths during excision of this tumor have been reported (*12*).

The pathophysiology of FNH is not understood. It is hypothesized that it is not a true neoplasm, but a

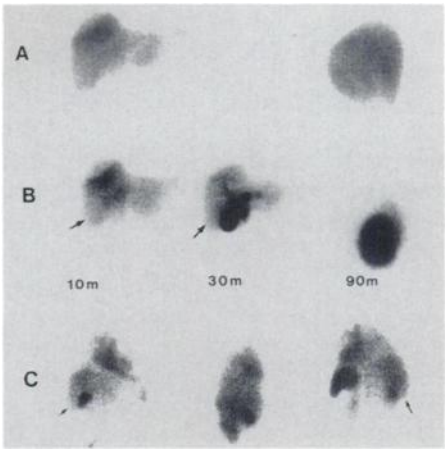

FIGURE 2
(A) Technetium-99m-SnC scan showed normal, uniform hepatic uptake of tracer. No focal lesions were demonstrated. The curvilinear band across the upper right lobe of the liver was caused by breast scatter. (B) Hepatobiliary scan with ^{99m}Tc-IDA showed normal uptake and excretion by the liver. At 90 min, most of the tracer was seen in the gall bladder. Arrow indicates location of the lesion. (C) Anterior, right lateral, and posterior ^{99m}Tc-labeled red-cell blood-pool images. There is normal concentration in the area occupied by the lesion (arrow). (The gall bladder is filled with ^{99m}Tc-IDA from the study performed earlier in the day.)

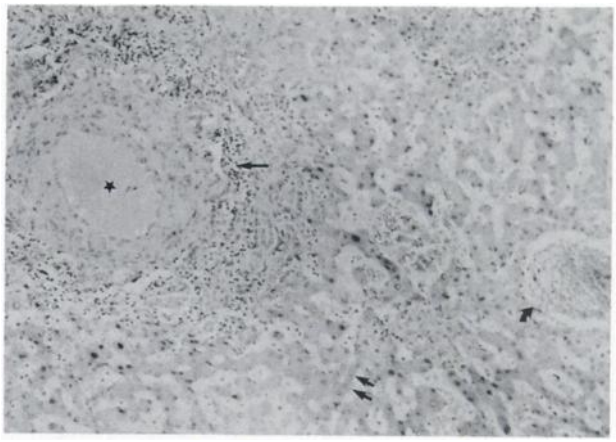

FIGURE 3
Lesion histology demonstrated well-demarcated, nonencapsulated areas of diffuse prominent sinusoidal ectasia (curved arrow), extending in some places to the portal spaces, and sparing most of the pericentral vein areas. Sinusoids were lined by endothelial and Kupffer cells. Portal spaces were prominent, and contained large muscular arteries (star). The liver cell plates (double arrows) were not involved; only a few necrotic hepatocytes were present at a site of maximal sinusoidal distention. A slight lymphocytic infiltrate (single arrow) not associated with necrotic hepatocytes was present. Bile ducts displayed ductular hyperplasia, but were otherwise unremarkable. The lobular architecture of the liver was well preserved.

hyperplastic response of liver parenchyma to abnormal blood flow through anomalous vessels at the center of the lesions (*13,14*). Two histologic forms have recently been distinguished; a solid form, in which abnormal vessels are often occluded by myointimal hyperplasia, and associated with fibrosis and distorted hypertrophic bile ducts; and a telangiectatic form characterized by large ectatic sinusoids and bile duct hypertrophy, but relatively less distortion of internal architecture (*7*). Hepatocytes are generally normal in both forms. The two subtypes may coexist, but whether the telangiectatic form represents a precursor of the solid lesion is not known.

Recent interest in FNH has focused on a possible association of cases with multiple lesions of FNH and the presence of meningioma, astrocytoma and other neoplasms (*7*). In a study of 2,500 consecutive autopsies, Wanless et al. (*7*) noted that FNH lesions, when multiple, were often of the telangiectatic form and usually very small: the largest telangiectatic lesion they reported was 2 cm, and most were less than 1 cm. These measurements are at the limits of resolution for current imaging methods, and it is unlikely that such lesions would have been detected in vivo.

Scintigraphic findings in cases of solid FNH are variable, and reflect the degree of distortion of the normal hepatoportobiliary architecture in the lesions. Bile duct hyperplasia and increased resistance to biliary flow causes intralesional retention of ^{99m}Tc-IDA (*3–6*).

Most lesions exhibiting abnormal biliary dynamics in this way also demonstrate normal uptake of ^{99m}Tc-SnC, indicating functional preservation of Kupffer-cell phagocytosis. Decreased ^{99m}Tc-SnC uptake develops when either delivery of tracer to the lesion is impaired by altered blood flow dynamics through anomalous or occluded vessels, or when phagocytic function in Kupffer cells becomes impaired by ischemia. Similar explanations have been proposed to account for decreased ^{99m}Tc-SnC uptake by HCAs, which have been shown to contain Kupffer cells, often in abundant supply (*15*). A search of the literature failed to find a description of the imaging characteristics of telangiectatic FNH.

The case described in the present report represents a telangiectatic FNH large enough to have been reliably examined by diagnostic imaging. It was distinctive in that the scintigraphic methods normally used to characterize FNH, ^{99m} Tc-SnC uptake and ^{99m}Tc-IDA excretion, failed to reveal any functional abnormality in this 9-cm lesion. This was consistent with the relative lack of histologic distortion, apart from sinusoidal ectasia, present in the lesion. Despite the demonstration of multiple tortuous and ectatic intralesional vessels by angiography, ^{99m}Tc-labeled RBC studies failed to resolve any abnormality. A probable explanation is that although individual hepatic arteries in the lesion were enlarged and ectatic, they remained too small to be resolved by the ^{99m}Tc-labeled RBC blood-pool technique, and that the hypervascular supply from the hepatic artery balanced the corresponding paucity of the normally dominant portal supply such that the overall blood-pool density within the lesion was not altered relative to that of surrounding normal liver tissue.

If the appearance described in the present report is typical of the telangiectatic form of FNH, it may offer a clue to the presence of additional hepatic lesions too small to be visualized. This may identify patients at risk for meningioma or glioma, and allow detection of these brain lesions at a stage when they are still clinically silent. A corollary of these findings is that lesions which are clearly disclosed by CT, US, and angiography, and of a size that is resolvable by planar or SPECT imaging, but exhibit normal ^{99m}Tc-SnC and ^{99m}Tc-IDA dynamics, do not represent a radionuclide failure. Rather, they render insight into the pathophysiology of these lesions, which may represent an early stage in the evolution of FNH.

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