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Diffuse Hepatic Hemangiomatosis in an Adult

Diffuse hepatic hemangiomatosis without extrahepatic lesions is extremely rare in adults. A case of diffuse hepatic hemangiomatosis involving right lobe in a 50-year-old woman was presented. The hemangiomatosis was demonstrated by ultrasonography, computerized tomography (CT) and magnetic resonance image (MRI), and was confirmed histopathologically. Although diffuse hepatic hemangiomatosis is a rare disease in adults, its diagnosis should be considered in patients with diffuse tumor growth in one or both hepatic lobes and distinguished from malignant tumors. The present case is the first documented case of diffuse hepatic hemangiomatosis in an adult in Korea.

Key Words: *Liver; Hemangioma; Adult*

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INTRODUCTION

Diffuse hepatic hemangiomatosis usually presents in infancy as an abdominal mass or as unexplained congestive heart failure (1). In some patients, the disease is associated with Rendu-Osler-Weber's disease or skeletal hemangiomatosis (2-4). Isolated diffuse hemangiomatosis of liver in adult without extrahepatic lesions is extremely rare (5-8). We describe a surgically resected case with diffuse hemangiomatosis of the right hepatic lobe in a 50-year-old woman.

CASE REPORT

A 50-year-old woman was admitted to our hospital with one-year history of post-prandial epigastric discomfort and indigestion. She also suffered from repeated episodes of multi-focal joint pain for about three years. She has been taking medication for arthralgia irregularly. There was no history of oral contraceptives usage. On physical examination, there was tenderness in the right upper abdomen, but no palpable mass. Cardiac or abdominal murmur was not heard. Laboratory findings revealed hemoglobin 12.6 g/dL, hematocrit 36.7%, platelet count 220,000/μL, alpha-fetoprotein 2.3 ng/mL, which were normal.

On abdominal ultrasound, numerous, ill-defined, hyperechoic nodules, ranging from a few millimeters to several centimeters in diameter as well as echo-lucent intervening tissue replaced the right lobe of the liver. Abdominal CT scan with contrast enhancement showed intense enhancement of the lesion and dilated hepatic artery with its branches that supply the tumor. Generally, all tumors showed low signal intensity on T1-weighted MR images, and high signal intensity on T2-weighted MR images (Fig. 1). Some large tumor nodules, however, had foci of lower intensity than the remainder of the tumor on T1-weighted images and higher intensity than the remainder of the tumor on T2-weighted images. Those foci were considered as regions of cystic change,

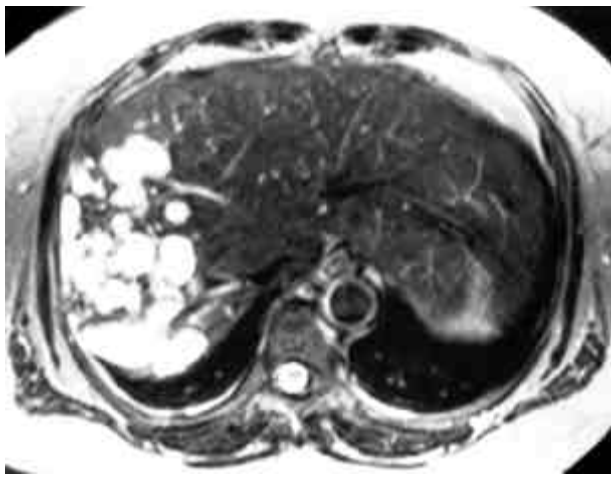

Fig. 1. T2 weighted spin echo image shows numerous, variable-sized, high-signal intensity nodules with intervening normal hepatic parenchyme in the same lobe.

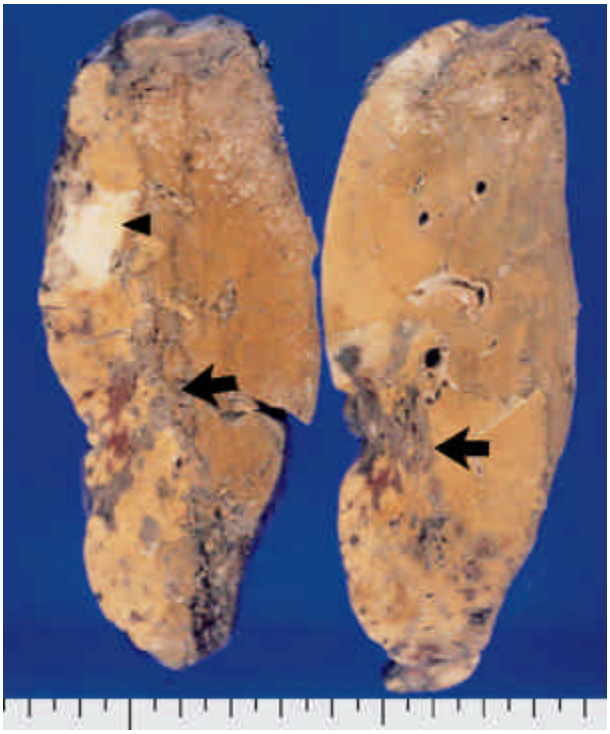

Fig. 2. Photograph of resected lesion. The lesion consists of multiple cavernous hemangioma with spongy appearance (arrow) and solid area of hyaline degeneration (arrow head).

but confirmed as hyaline degeneration by histology. On dynamic MR imaging, tumors demonstrated complete dense enhancement or intense peripheral contrast enhancement with a centripetal filling pattern and several foci of non-enhancing portion, within some nodules. In ^{99m}Tc-labeled red blood cell scanning, blood pool images showed multi-focal increased activity in right lobe of the liver by 1 hr.

Clinical impression was giant liver hemangioma. In the operative field, the lesion was located at entire right lobe of the liver, which was closely attached to right main hepatic vein. The lesion was solid and well demarcated with no encapsulation. Although much bleeding occurred during operation, mass was entirely removed by right hepatectomy. Grossly, the specimen measured 17×14×9 cm in dimension and multiple hemorrhagic blood-filled honeycomb areas from 2-3 mm up to 3 cm in diameter were scattered throughout entire right lobe (Fig. 2). Histologically, most of the individual lesion showed ordinary cavernous hemangioma alternated with hepatic parenchyme. Blood filled vascular channels were lined by flattened endothelial cells without cellular atypia (Fig. 3). In some areas, sprouting blood vessels in the vicinity of the portal tracts were observed. Two weeks after operation, the patient was well and could be discharged. Nine months after the surgery, clinical evolution was favorable, and no mass was visible on ultrasound examination.

DISCUSSION

Cavernous hemangioma is the most common type of hepatic tumor and are most frequently seen in women between third and fifth decade (5, 6). In some patients, diffuse hepatic hemangiomatosis was reported in relation with hereditary hemorrhagic telangiectasia (4). Heredi-

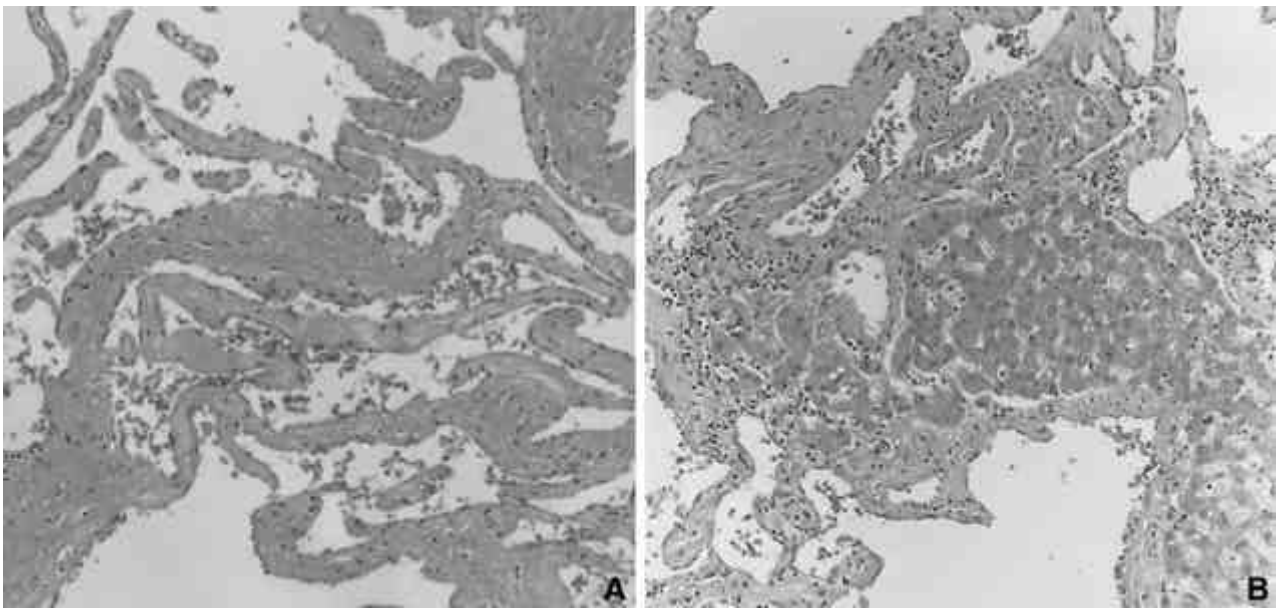

Fig. 3. **A:** Multiple cavernous vessels without angiosarcomatous characteristics are noted (H&E, ×100). **B:** Cavernous hemangioma alternating normal hepatic parenchyme (H&E, ×100).

tary hemorrhagic telangiectasia, also called Rendu-Osler-Weber's disease, is a systemic fibrovascular dysplasia with autosomal dominant inheritance (4, 9). Epistaxis and cutaneomucous telangiectasia of the face, the hands, and the oral cavity are the most frequent symptoms. Involvement of the gut with hemorrhage and of other organs, including lung, central nervous system, and liver have been described (9). Our patient did not show any clinical features of Rendu-Osler-Weber's disease. In other cases, intestinal and hepatic hemangiomatosis was associated with skeletal hemangiomatosis (2, 3). In infancy, hemangiomatosis usually involves entire liver diffusely. It acts as an intrahepatic arteriovenous shunting, leading to high cardiac output condition and thus congestive heart failure (1). In contrast, diffuse liver hemangiomatosis is extremely rare in adults, and only a few patients with isolated liver hemangiomatosis have been reported in English literature (5-8).

Small hemangiomas of liver usually present no symptoms, but giant hepatic hemangiomas and diffuse hemangiomatosis present symptoms of abdominal pain, discomfort, and palpable mass, and often intrahepatic arteriovenous shunts, leading to high cardiac output state and eventually to heart failure (5, 6). Rare complications include rupture, thrombocytopenia and consumptive coagulopathy (10).

There have been only few report about imaging findings of diffuse liver hemagiomatosis (5, 7). In this case, CT and MR imaging features of each nodule were similar to that of hemangioma. However, the nodules were variable in size, from several millimeters to several centimeters, and also, conglomerated and intermingled. In addition, normal hepatic parenchymal tissue was identified among enhancing nodules and it looked like infiltrating hypervascular malignant tumor. Although the first choice of differential diagnosis at the stage of presentation was diffuse hemagiomatosis, but angiosarcoma and epithelioid hemangioendothelioma could not be ruled out confidently because imaging characteristics of these malignant vascular tumors could parallel to those of hemangioma (11). The diagnosis of diffuse liver hemangiomatosis can be suspected by ultrasonography, CT, MRI and hepatic arteriography, and has to be confirmed histologically.

The histological features are characterized by the presence of large vascular channels, which are not restricted to the cavernous tumor parts, but are also found in the otherwise normal appearing liver parenchyme (5). The findings of focal portal fibrosis and sprouting slitlike vessels in the vicinity of the central vein are also described (7).

The etiology and natural history of diffuse liver hemangiomatosis are not well understood. Lehmann et al. reported that hemangiomatosis was regarded as an angiogenesis-dependent condition with phase of activation and regression (5). Factors that modulate tumor growth are poorly understood. The role of sex hormones and steroid medication during the development of hepatic cavernous hemangiomas are reported (12), but the use of estrogen has not been described in the reported cases of diffuse liver hemangiomatosis (5-8). One patient who had received medication with metoclopramide developed diffuse hepatic hemangiomatosis and arteriovenous shunting (7). There was no history of steroid or estrogen intake in our patient. The prognosis of reported isolated diffuse liver hemangiomatosis is uncertain because of the rarity of this disease. In the literature, one patient developed hepatorenal syndrome and finally died (6), but in another patient, severe arteriovenous shunting and cholestasis resolved spontaneously (7). Lehmann et al. described a case with diffuse hemangiomatosis of the left hepatic lobe, who developed progressive tumor growth in the right hepatic lobe after left hepatectomy (5). The most recent follow-up in nine months after surgery revealed that our patient was well. Until more cases of diffuse liver hemangiomatosis are intensely studied, the natural history and prognosis cannot be established.

There are several treatment methods for diffuse hepatic hemangiomatosis (13-16). Surgery is not generally recommended because of poor delineation of the tumor boundaries and risk of intraoperative hemorrhage. Our patient underwent right hepatectomy because of well localization of tumor in one lobe and possibility of heart failure. Surgical resection was effective, the disturbing symptoms were completely resolved and there was no evidence of recurrence after the operation. A previous study has shown that a symptomatic giant hemangioma within one lobe should be treated by hepatectomy, but if it involves both lobes, ligation of the hepatic artery, with or without radiation, should be considered (13). We believe that non-surgical treatments are effective for unresectable cases and that they decrease the large arteriovenous pooling in hemangiomatosis of the liver. Liver transplantation should be considered in patients with congestive heart failure and in whom the other treatment methods will not be applicable (6).

Our case suggested that diffuse liver hemangiomatosis can affect patients at any age and it should be considered in patients with multiple tumor growth in one or both hepatic lobes.

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