



Giant Hepatic Hemangioblastoma - Report of a Rare Sporadic Case

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Abstract

We report here a sporadic case of giant hepatic hemangioblastoma. The patient has no history and other signs of Von Hippel-Lindau disease. This is the 3rd reported case of hepatic hemangioblastoma and probably the largest hemangioblastoma ever reported.

Introduction

Hemangioblastoma is a benign tumor of mesenchymal origin which occurs predominantly in the cerebellum. About 1/5 of all the cases is associated with *VHL* gene mutation, the so called Von Hippel-Lindau disease. Occasional cases have been reported in other parts of the central nervous system and organs such as kidney and soft tissue [1-5]. However, so far only two cases of hepatic hemangioblastoma have been reported and were both associated with Von Hippel-Lindau disease [6,7]. We here report a case of giant liver tumor of hemangioblastoma. The patient showed no signs of Von Hippel-Lindau disease.

Case Presentation

Clinical data

A male patient aged 43 was admitted to the hospital with main complain of discovery of liver space-occupying lesion. In December of 2021, the patient visited the clinic for "hydronephrosis with renal and ureteral stone". Abdominal computer tomography (Figure 1) revealed a giant space-occupying lesion in the right lobe of liver with a dimension of 20.7 cm × 18.7 cm × 11.9 cm. The liver was enlarged and the left margin pushed to the left end of abdomen. The patient was with no complaining of fatigue, poor appetite, abdominal distention or pain. Laboratory investigation after admission revealed no abnormality of blood routine, urine routine, stool routine, coagulation function, CRP, renal function, electrolytes, and the triple liver cancer biomarkers AFP, AFP-L3, and PIVAKII. Digital radiography showed no special findings of heart, lungs and diaphragm. Head and brain MRI revealed no abnormality of brain. The radiologists considered a diagnosis and differentials of mesenchymal originated tumor, angiomyolipoma, solitary fibrous tumor, or hepatic hemangioma. Three days after admission, the right lobe of liver and gallbladder of the patient were dissected out under total anesthesia. The patient recovered smoothly and was discharged 10 days later.

Pathological findings

A giant tumor with very thin margin of liver tissue was seen measuring 20 cm × 19 cm × 12 cm. The tumor was solid, dark red, not encapsulated but well circumscribed. Microscopically, the tumor was composed of large amounts of compact capillaries and oval or fusiform eosinophilic or vacuolated stromal cells. The nuclei of stromal cells were small and compact, mitotic figures rarely found. Distended venules and hemorrhages were frequently seen. Hematopoietic foci (Figure 2) were also occasionally spotted. Immunohistochemical (Figure 3) investigation demonstrated that tumor cells were positively stained for vimentin, CD56, and TFE3. GFAP were spottily stained. CD163 and CD68 stain revealed abundant existence of histocytes distributed among the tumor cells. Endothelial cell markers CD34 and CD31 positively labeled large blood vessels but not capillaries and stromal cells. Negatively stained markers included CK, S100, HMB45, Melan A, stat6, NSE, RCC, Pax 8, and α-inhibin. Cell proliferation index Ki67 was about 10%.

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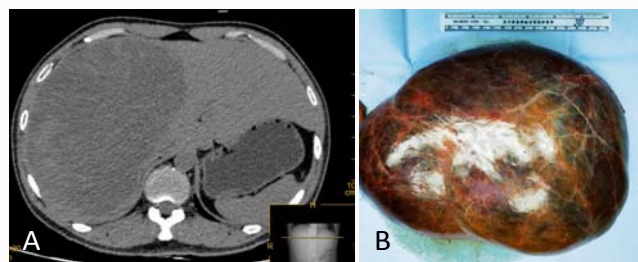


Figure 1: A. Computer aided tomography of the liver reveals a huge space occupying lesion measuring around 20.7 cm x 18.7 cm x 11.9 cm with a well-defined margin. The left lobe of the liver was pushed to the left end of the abdominal cavity. B. The right lobe of the liver was dissected out.

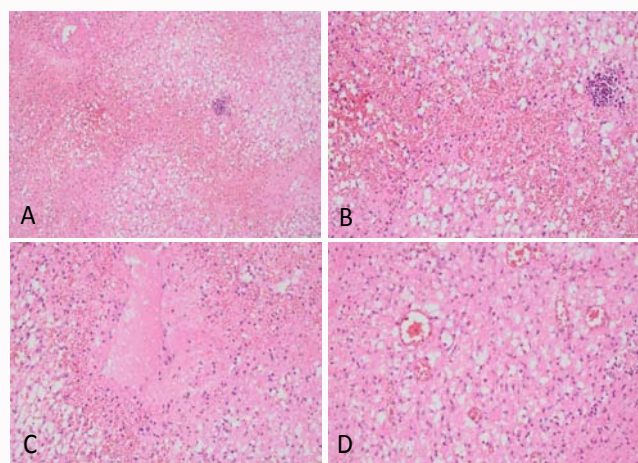


Figure 2: Histological images show the tumor was with many hemorrhagic regions (A and B), occasional extramedullary hematopoietic focus (B), dilated blood vessels (C), capillaries and stromal cells (D).

Based on the morphological features and Immunohistochemical findings, a diagnosis of hemangioblastoma was made after excluding the differentials of hemangioma, solitary fibrous tumor, angiomyolipoma and metastasis of renal clear cell carcinoma.

Discussion

hemangioblastoma is a rare benign tumor that mostly occurs in the cerebellum, and about one fifth of the cases were in Von Hippel-Lindau patients, with mutations of the *VHL* gene. It also occurs in other parts of the central nervous system, but in much lower frequency. Outside the central nervous system, there were cases reported in the soft tissues and kidneys. We found only 2 cases of hepatic hemangioblastoma in the literature and both of them were associated with Von Hippel-Lindau diseases, and were much smaller.

Hemangioblastoma generally were very small due to the restriction of cerebellar location. But even outside the central nervous system, such as inside the kidney, the tumors were generally not large either. Our case is probably so far the largest one ever reported. Interestingly, with such a large tumor, the patient did not manifest any symptoms. The tumor did not destruct the normal liver tissue but rather enlarged it and pushed the left margin of the liver all the way to left end of the abdomen, indicating the benign nature of the tumor.

The diagnosis of hemangioblastoma in cerebellum is rather direct.

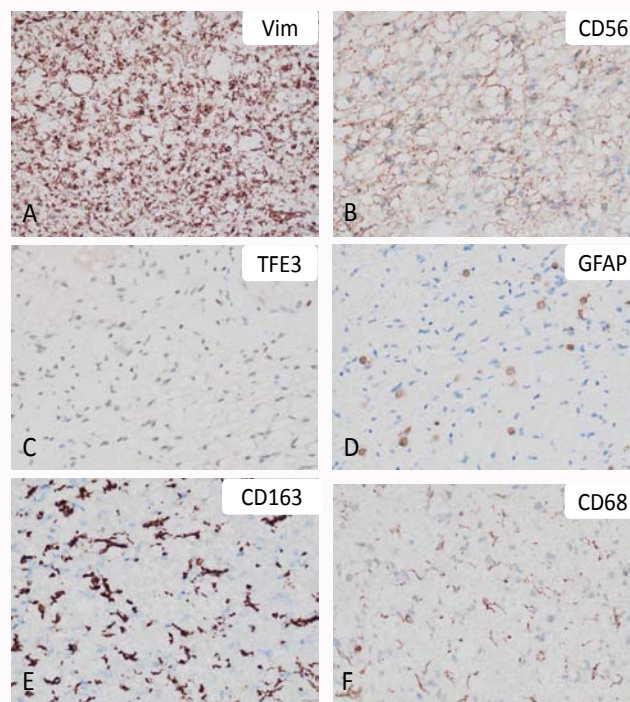


Figure 3: Immunohistochemical phenotype of the tumor. The tumor cells were positively stained by vimentin, CD56, TFE3, spotty GFAP, CD163, and CD68.

But in other parts of the body, it could pose a serious challenge because the rarity of incidence. In the present case, the initial differentials were hemangioma, angiomyolipoma, solitary fibrous tumor, and metastatic renal cell carcinoma. The final diagnosis of hemangioblastoma was in fact the last one came to mind after excluding all the others. Although Immunohistochemistry is helpful in the diagnosis, there are no signature markers for the diagnosis of hemangioblastoma. It has been reported in most of the cases that S100, NSE and α -inhibin was positive, but not in the present case. On the other side, the positive staining of vimentin, CD56, TFE3, and spotty positivity of GFAP seen in this case has been reported in some studies and lays support for the diagnosis [8]. But the fundamental basis for the diagnosis is the histological feature and the exclusion of other differentials rather than Immunohistochemistry.

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