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Case Study
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Ablation Therapy for Hepatic Epithelioid Hemangioendochelioma

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Keywords

Ablation; Hemangioendochelioma; Therapy

Abbreviations

AFP: Alpha-Fetoprotein; CEA: Carcinoembryonic Antigen; TAE; MWA: Microwave Ablation; RFA: Radio Frequency Ablation

Background

Hepatic Epithelioid Hemangioendochelioma is rare all over the world. There few effective strategies for it. Single lesion can be performed by resection. Hepatic transplantation is another effective treatment way for extensive epithelioid hemangioendochelioma. In our institution, one patient was treated by interventional therapy with good response.

Case Study

A 48-year-old male patient was referred to our department with liver lesions without hypodynamia, nausea, vomiting and wasting in October 2018. He got checkup one month ago. He had no special past illness. He had no family history of cancer. On physical examination, there were no abnormal. Abdomen enhanced Computer Tomography image showed that: there were multiple lesions in liver, among them; the biggest one was 39 mm × 36 mm, CT value was 33 Hu. Cycling enhancement style in artery phase. The enhancement area became bigger in venous phase (Figure 1). Contrast enhanced ultrasound showed: multiple metastasis lesions in liver. Investigations revealed normal AFP (2.25 ng/mL) and CEA (2.68 ng/mL), the other laboratory findings had no significance. PET-CT showed high hyper metabolism lesions, delay phase showed enhanced more, malignant lesions. Fine needle liver biopsy showed (2018.10.17): neoplastic cell infiltrated in loose fibrous tissue, the neoplastic cells are epithelial-like, arranged in cavities partially, hyperchroma

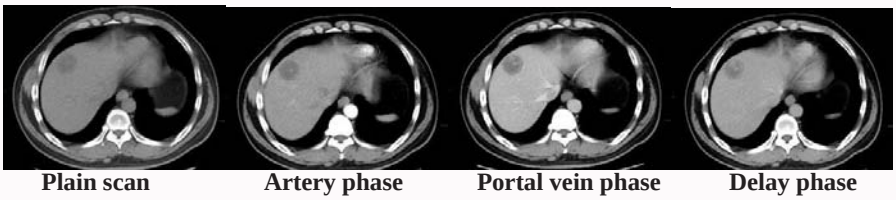

Figure 1: CT showing multiple nodular lesions scatters in different segments of liver. The margin is enhanced. There are arteries in them.

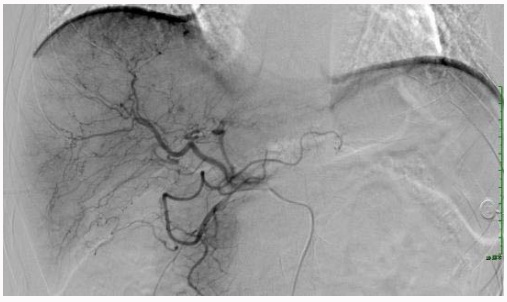

Figure 2: Angiography of epithelioid hemangioendothelioma.

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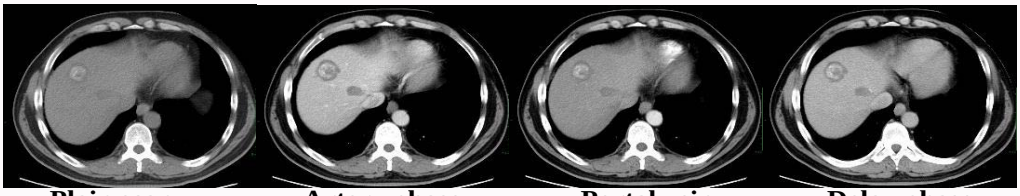

Figure 3: Follow up enhanced CT: no recurrence.

of the nucleus, mild abnormity. The immune histochemical study showed: HBsAg (-), HBcAg (-), Hepa and GPC-3 were negative, while CK19 was bile ducts positive, CD34 was double positive, proliferative index was low (Ki6710%), P53 was mutation type, CK7 was positive. The diagnosis was hepatic epithelioid hemangioendochelioma. TAE (2018.10.31 see (Figure 2)) and MWA (2018.11.14) RFA (2018.11.21) were performed subsequently. The lesions were ablated completely with 5 mm security margin. Protect hepatic medicine was administrated. After several days later, the patient was discharged without complications.

3 months later, follow-up abdomen enhanced CT findings approved no recurrence. The interval checkup would be obeyed every 3 months (Figure 3).

Discussion

Hepatic epithelioid hmangiendochelioma was a rare disease (the prevalence is 1/1,000,000). The etiology was unknown. The clinical syndrome was non-specific. It was a low to intermediate malignant neoplasm which was origin from vascular. It composed of epithelioid-appearing endothelial cells.

Radiological characters were as following: CT showed multiple lesions with retractive capsular scatters in peripheral portion of the liver parenchyma. The rim enhancement in early phase, the center enhancement in delay phase, like centripetal enhancement in hepatic haemangiomas. While for the small ones, they were mimic metastasis. In the center, heamorrhage and necrosis can be detected if the lesion was big. AV shunting could be found in some cases [1]. MRI showed the lesions were T1-Weighting hypointense, T2-Weighting hypertense, while not as intense as hepatic haemangiomas [2]. Some diagnostic enhancement signs could be found such a score pattern, ring-like arterial enhancement, layered enhancement, and entrapment-like pattern [3], "Target sign"," Lollipop sign" [4,5]. The special pattern of ADC map was the tumor with a peripheral low signal intensity area surrounding the central high signal intensity area [6]. Angiography showed tumor stain. There was prolonged pooling of contrast within the mass. Internal AV shunting with large draining veins might be seen. Lipiolol scattered in the lesion if TAE was performed.

The diagnosis depended on immune histochemical biopsy. By immune histo chemistry, the findings of positivity for CD31, CD34 and factor-VIII-related antigen were the key for diagnosis. Microscopic findings of characteristic proliferation of dendritic or epithelioid cells with abundant eosinophilic cytoplasm and occasional intracytoplasmic vacuolization in myxoid or fibrous stroma.

The differential diagnosis of was extensive and broad, as abscesses, cholangiocarcinoma, liver metastasis, haemangioma, metastases etc.

Although a little case could spontaneous regressions, surgical resection was recommended if possible. Target, immune or chemotherapy were administrated such as thalidomide, interferon, apatinib, sorafenib, pazopanib, sirolimus and different chemotherapy regimens (carboplatin, paclitaxel, and bevacizumab; carboplatin and etoposide; Adriamycin, dacarbazine, and ifosfamide; cyclophosphamide, adriamycinandvincristine; bevacizumab and nab-paclitaxel).

Orthotopic liver transplantation was performed in many cases. The outcome was effective. For young-aged patients with big bulk lesion in liver, transplantation was recommended even metastasis exist [7].

Recently, there were several cases report relative with haemangioendothelioma treatment: DongyueGu reported one patient was treated with radiation by the guiding of ultrasound, follow up 2 years without recurrence. Li Janjun [8] reported that Microwave Ablation could be a new way to cure haemangioendothelioma. RC Cerba used 6 cases showed different outcome with different regimens. The treatment tactics depended on the cases [9].

This case was asymptomatic and discovered by checkup. Because it was multiple lesions, it was not fit for liver resection. According to the treatment strategies, chemotherapy could be performed with little response. It should be candidate for liver transplantation. However, the economic issues would be the obstacles. In our department, TAE was performed to trace the lesions by lipiodol. After that, the lesions were ablated divided into two series in order to avoid major complications. According to the follow up CT, complete respond was obtained.

Conclusion

Hepaticepithelioid hemangioendochelioma can be controlled effectively by interventional therapy which is economic and performed easily. More samples needed to confirm it.

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