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A Giant Solitary Fibrous Tumor of the Liver with Metastasis and Local Recurrence

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Solitary fibrous tumor (SFT) is a rare type of mesenchymal neoplasm. Although the majority of SFTs are benign, some cases have shown characteristics of malignant neoplasms. Weight loss, fatigue, and upper abdominal bloating are the main signs of these lesions. Clinical and radiographic features are not sufficient for the diagnosis of hepatic SFT and the definitive diagnosis depends on histopathological sampling and immunohistochemistry. One of the main issues in the diagnosis of this tumour is the ability of this tumour to grow to large sizes. A 69-year-old male presented to the clinic 2 years earlier with episodes of hypoglycemia and loss of consciousness. The symptoms improved after receiving glucose. The patient diagnosed as rare solitary fibrous tumour of the liver, a giant (10 × 10 cm in diameter) round and well-defined lesion in the left lobe of the liver which was obvious in magnetic resonance imaging (MRI). Surgery is the most common line of treatment for this disease and there is no evidence regarding the effectiveness of other approaches. According

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to the scarcity of hepatic SFT, long-term prognosis in these patients is highly challenging. Here, we present the case of a 69-year-old male patient with hepatic SFT with metastasis and local recurrence. In the very rare malignant form of liver solitary fibrous tumour which is surgically unresectable, liver transplantation is one of the potential options but maybe not amenable due to the malignant behaviour of the disease. Role of debulking surgery is also not clear in this situation.

Keywords: Solitary fibrous tumour; resection; mesenchymal neoplasms; malignancy.

1. INTRODUCTION

Mesenchymal neoplasms are soft tissue tumours that can develop in any part of the body [1]. Solitary fibrous tumours (SFT) are a rare type of mesenchymal neoplasms, which are originally described in the pleura. These lesions were initially differentiated from mesothelioma in 1931 [2]. SFT of the pleura is the most common site of this lesion, however, it may be seen in other anatomic sites, even in abdominal wall soft tissue and the liver. Although the majority of SFTs are benign, some cases have shown characteristics of malignant neoplasms. Despite adequate local control, the recurrence rate of malignant SFTs is estimated at 30% [2].

Clinical and radiographic features are not sufficient for the diagnosis of hepatic SFT and the diagnosis of this lesion, even via surgery, is a highly challenging issue. Commonly, SFT is observed as a heterogeneous solitary encapsulated highly vascular tumour in computed tomography (CT) scan. The essential tools for the diagnosis of hepatic SFT are histopathological sampling and immune-histochemistry. The tendency of this tumour to grow to large sizes is one of the main issues that should be considered at the time of diagnosis [3]. Solitary Fibrous Tumor / Hemangiopericytoma of Palate - Report of a Case with Immuno-histochemical Interpretation Using CD 34 was reported by Bajpei et al. [4].

In the process of diagnosis, the exclusion of other diagnosis is very important, especially regarding the hepatocellular carcinoma, gastrointestinal stromal tumours, leiomyoma, and sarcoma [5]. Although there are no certain clinical symptoms and signs for the diagnosis of hepatic SFT, weight loss, fatigue, and upper abdominal bloating are some signs of these lesions [5]. Long-term prognosis in patients with hepatic SFT has been rarely described due to the rarity of this problem. Here, we present a case of hepatic SFT with soft-tissue metastasis and his follow up.

2. CASE REPORT

A 69-year-old male presented to the clinic 2 years earlier with episodes of hypoglycemia and loss of consciousness. The symptoms improved after receiving glucose. The patient diagnosed as rare solitary fibrous tumour of the liver, a giant (10 × 10 cm in diameter) round and well-defined lesion in the left lobe of the liver which was obvious in magnetic resonance imaging (MRI), as we had described previously [2]. Under general anaesthesia, laparotomy was performed, and wide local excision of the lesion was done. Hypoglycemia was improved after resection of the tumour.

Eight months later, the patient was readmitted due to a bulging mass in the right inguinal hernia, which was identified as synovial sarcoma based on incisional biopsy. But based on permanent pathology report the lesion was diagnosed as SFT, which was 10×10 cm in diameter. The tumour was surgically removed, and until five-month follow-up, no signs of recurrence were observed.

After five months, he presented to the clinic again with progressive dyspnea, hypoglycemia, and loss of consciousness. On chest CT scan a mass was obvious and he underwent resection of a tumor mass in the right hemithorax, which was diagnosed as SFT. During another 10-month follow-up, he did not show any signs of hypoglycemia or dyspnea. But at 10 month, he came back with recurrence of previous symptom and large liver mass involvement were found and confirmed with needle biopsy as a recurrence of Solitary Fibrous Tumor. Surgery was not possible due to the involvement of multiple segments (Fig. 1). No evidence of liver failure has been seen. At this point the patient is treating with continuous oral and occasionally intravenous glucose.

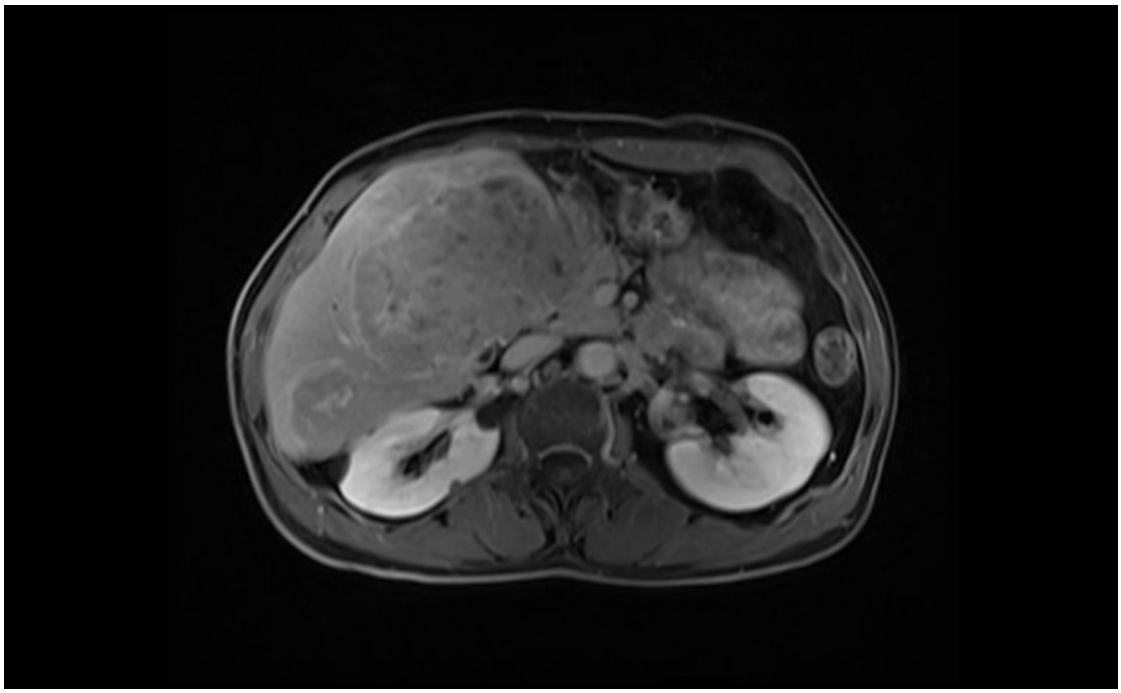

Fig. 1. Heterogeneous large liver mass surrounded with normal hyper-intense liver tissue in T1-weighted image, which represents a large liver solitary fibrous tumour

3. DISCUSSION

Benign SFT is a rare condition, even more so malignant hepatic SFTs. The majority of cases of hepatic SFT were reported in middle-aged and elderly patients. However, there have been some cases of SFT in patients aged 16-87 years old. These lesions are observed in both sexes and the female/male ratio is 1.4:1 [5,6]. Based on macroscopic examination, hepatic SFT is observable in different sizes, which can range from 0.5 to 35 cm [5-7].

In general, SFT mostly develops in the pleura, but it may be observed in different anatomic sites [2]. Nonetheless, it is rarely observed in the abdominal wall soft tissues and the liver [2]. Although extrathoracic SFTs are also found across different anatomic regions of the human body, but they are rarely observed in the abdominal cavity [8]. The majority of extrathoracic SFTs are benign; however, they may be aggressive and malignant in some cases [2]. So far, very few cases of hepatic SFT have been reported, the majority of which were in the left lobe of liver. Here, we reported a case with a history of hepatic SFT and recurrence of the tumour in the right hemithorax and the liver.

SFT lesions have different clinical presentations, such as abdominal pain, cholestasis, and hypoglycemia; however, it may be totally asymptomatic in some cases [9]. The secondary symptoms of SFT include dyspepsia, postprandial pain, nausea, vomiting, and jaundice [10-14]. In some cases, symptoms are resolved after resection of the tumour [11,15,16]. Hypoglycemia is the main reported symptom of hepatic SFT, which is resistant to medical management and usually, its recovery is related to resection of hepatic SFT. Hypoglycemia is caused by insulin-like growth factor 2 (IGF-2) expression induced by hepatic SFT. The severity of hypoglycemia caused by the tumour is related to tumour burden [17]. In the present case, metastasis occurred concomitant with hypoglycemia recurrence. However, hypoglycemia is not limited to cases with hepatic SFT [18]. Therefore, hypoglycemia cannot be considered as a specific marker of malignant hepatic SFT.

Most cases of hepatic SFT are benign, and less than one-fifth of them are malignant. There is insufficient data regarding SFT's malignant potential and invasive growth, as well as the natural history of these lesions. In many patients, these lesions are incidentally found during

pathology examination. In this regard, the exclusion of differential diagnosis (i.e., hepatocellular carcinoma, gastrointestinal stromal tumours, leiomyoma, and sarcoma) should be considered [5]. Similar to benign SFT, malignant SFT occurs more commonly in middle-aged and elderly patients, and its frequency in women is higher than in men.

Usually, hepatic SFT is observed in the form of a highly vascular solitary tumour. These lesions have local growth and they tend to grow to large sizes, which are resected at the time of diagnosis [2]. So far, no specific laboratory markers have been suggested for the diagnosis of hepatic SFT. Based on MRI findings, hypointense signals with a heterogeneous mass, which may be observed along with calcification, indicate a high content of collagenous tissue in T1 weighted image, and heterogeneous masses including hypo or hyperintense or both of them in T2 weighted are observable in patients with SFT of the liver [18,19].

Progressive heterogeneous enhancement in the arterial phase related to the hypervascular region, which is developed in the venous and delayed phases, was observed in the imaging studies [20]. Imaging methods are common and proper for the diagnosis of SFT; however, these approaches cannot be applied to distinguish between malignant and benign tumors [2]. Moreover, a preoperative percutaneous biopsy is a challenging issue for diagnosis due to its many side effects. For example, a preoperative biopsy may lead to needle tract seeding, pain, intrahepatic haematoma, and bile leakage [21-23]. Therefore, a preoperative biopsy can change the course of treatment [15].

One of the main problems in the diagnosis of SFT is the lack of sufficient association between SFT behaviour and histopathologic findings. Clinical and radiographic characteristics are not sufficient for a definitive diagnosis, and histopathological sampling and immune-histochemistry should be performed for this purpose. Long-term prognosis of hepatic SFT is rarely described due to limited data in the literature [2]. One the other hand, there are a few markers such as CD34 and CD99 for the diagnosis of hepatic SFT; Usually, c-Kit (CD117), CD31, cytokeratins, EMA, factor VIII, epithelial membrane antigen, and S100 are reported negative in patients with hepatic SFT [5].

Surgery is the first line of treatment and there is no evidence regarding the effectiveness of adjuvant medical therapy for this problem. Moreover, controversial findings have been obtained regarding the role of chemotherapy and radiotherapy, which are mostly used in patients with signs of malignancy or when resection is incomplete [11]. Although there is a risk of malignant transformation in hepatic SFT, resection is performed to obtain a margin-negative specimen and treat malignant SFT of the liver [24,25].

4. CONCLUSION

In general, given the limited understanding of the biological nature of hepatic SFT, the diagnosis of the disease is difficult and its prognosis is unknown, and complete surgical resection is known as the first line of treatment. In this disease, long-term follow-up of patients is recommended due to its uncertain prognosis.

In the very rare malignant form of liver solitary fibrous tumour which is surgically unresectable, like our case, liver transplantation is one of the potential options but maybe not amenable due to the malignant behaviour of the disease. Role of debulking surgery is also not clear in this situation. Due to its rarity, the optimal management is not clear and it's subject of further evaluation as well.

CONSENT AND ETHICAL APPROVAL

As per university standard guideline patient consent and ethical approval has been collected and preserved by the authors.

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COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Peng L, Liu Y, Ai Y, Liu Z, He Y, Liu Q. Skull base metastases from a malignant solitary fibrous tumor of the liver. A case report and literature review. Diagnostic Pathology. 2011;6(1):127.

2. Farazmand B, Fahimi H, Tajiknia V, Tajiknia N. Giant solitary fibrous tumor of the liver, case report: Unusual site, extravagant presentation, aggressive behavior. Academic Journal of Surgery. 2017;4(2):58-60.
3. Janssen J, Wagenaar S, Bosch J, Vanderschueren R, Planteydt H. Benign localized mesothelioma of the pleura. Histopathology. 1985;9(3):309-13.
4. Bajpai M, Pardhe N, Chandolia B, Arora M. Solitary fibrous tumor / hemangio-pericytoma of palate - Report of a case with immunohistochemical interpretation using CD 34. J Coll Physicians Surg Pak. 2017;27:457-458.
5. Chen N, Slater K. Solitary fibrous tumour of the liver—report on metastasis and local recurrence of a malignant case and review of literature. World Journal of Surgical Oncology. 2017;15(1):27.
6. Moran CA, Ishak KG, Goodman ZD. Solitary fibrous tumor of the liver: A clinicopathologic and immunohistochemical study of nine cases. Annals of diagnostic pathology. 1998;2(1):19-24.
7. Moser T, Nogueira TS, Neuville A, Riehm S, Averous G, Weber JC, et al. Delayed enhancement pattern in a localized fibrous tumor of the liver. American Journal of Roentgenology. 2005;184(5):1578-80.
8. Zong L, Chen P, Wang G-Y, Zhu QS. Giant solitary fibrous tumor arising from greater omentum. World Journal of Gastro-enterology: WJG. 2012;18(44):6515.
9. Thway K, Ng W, Noujaim J, Jones RL, Fisher C. The current status of solitary fibrous tumor: Diagnostic features, variants and genetics. International Journal of Surgical Pathology. 2016;24(4):281-92.
10. Patra S, Vij M, Venugopal K, Rela M. Hepatic solitary fibrous tumor: Report of a rare case. Indian Journal of Pathology and Microbiology. 2012;55(2):236.
11. Brochard C, Michalak S, Aubé C, Singeorzan C, Fournier HD, Laccourreye L, et al. A not so solitary fibrous tumor of the liver. Gastroenterologie Clinique et Biologique. 2010;34(12):716-20.
12. Guglielmi A, Frameglia M, Iuzzolino P, Martignoni G, Manzoni G, Laterza E, et al. Solitary fibrous tumor of the liver with CD 34 positivity and hypoglycemia. Journal of Hepato-Biliary-Pancreatic Sciences. 1998; 5(2):212-6.
13. Kim H, Damjanov I. Localized fibrous mesothelioma of the liver. Report of a giant tumor studied by light and electron microscopy. Cancer. 1983;52(9):1662-5.
14. Yilmaz S, Kirimlioglu V, Ertas E, Hilmioglu F, Yildirim B, Katz D, et al. Case report: Giant solitary fibrous tumor of the liver with metastasis to the skeletal system successfully treated with trisegmente-ctomy. Digestive Diseases and Sciences. 2000;45(1):168-74.
15. Silvanto A, Karanjia ND, Bagwan IN. Primary hepatic solitary fibrous tumor with histologically benign and malignant areas. Hepatobiliary & Pancreatic Diseases International. 2015;14(6):665-8.
16. Radunz S, Baba HA, Sotiropoulos GC. Large tumor of the liver and hypoglycemic shock in an 85-year-old patient. Gastro-enterology. 2012;142(2):e10-e1.
17. Chan G, Horton PJ, Thyssen S, Lamarche M, Nahal A, Hill DJ, et al. Malignant transformation of a solitary fibrous tumor of the liver and intractable hypoglycemia. Journal of Hepato-Biliary-Pancreatic Sciences. 2007;14(6):595-9.
18. Tapias LF, Lanuti M. Solitary fibrous tumors of the pleura: review of literature with up-to-date observations. Lung Cancer Management. 2015;4(4):169-79.
19. Bejarano González N, García Borobia FJ, Romaguera Monzonís A, García-Monforte N, Falcó-Fagés J, Bella-Cueto MR, et al. Solitary fibrous tumor of the liver. Case report and review of the literature. Rev Esp Enferm Dig. 2015;107:633-9.
20. Nath DS, Rutzick AD, Sielaff TD. Solitary fibrous tumor of the liver. American Journal of Roentgenology. 2006;187(2):W187-W90.
21. Maccio L, Bonetti LR, Siopis E, Palmiere C. Malignant metastasizing solitary fibrous tumors of the liver: A report of three cases. Polish Journal of Pathology. 2015;66(1): 72-6.
22. Chang S, Kim SH, Lim HK, Kim SH, Lee WJ, Choi D, et al. Needle tract implantation after percutaneous interventional procedures in hepatocellular carcinomas: lessons learned from a 10-year experience. Korean Journal of Radiology. 2008;9(3): 268-74.
23. Feng LH, Dong H, Zhu YY, Cong WM. An update on primary hepatic solitary fibrous tumor: An examination of the clinical and

pathological features of four case studies and a literature review. Pathology-Research and Practice. 2015;211(12):911-7.

24. Terkivatan T, Kliffen M, De Wilt JH, van Geel AN, Eggermont AM, Verhoef C. Giant solitary fibrous tumour of the liver. World Journal of Surgical Oncology. 2006;4(1): 81.

25. Jakob M, Schneider M, Hoeller I, Laffer U, Kaderli R. Malignant solitary fibrous tumor involving the liver. World Journal of Gastroenterology: WJG. 2013;19(21): 3354.

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