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Rare solid and cystic presentation of hemangiopericytoma/ solitary fibrous tumor: A case report

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ABSTRACT

Hemangiopericytoma/Solitary Fibrous Tumor (HPC/SFT) is a rare fibroblastic sarcoma characterized by hypervasculature and STAT6 trans-nuclear localization. Cystic HPC/SFT is extremely rare. Due to the scarcity of cystic HPC/SFT cases, diagnostic and treatment guidelines are not well established. To our knowledge, we present the first case of cystic HPC/SFT observed in the liver. In addition, the patient had over 6 years of recurrent hypervascular solid HPC/SFT in the brain, bone, leptomeninges, liver and lung prior to developing a cystic HPC/SFT. Briefly, a 37-year-old Caucasian female with a history of HPC/SFT presented with several enlarging cystic hepatic lesions on surveillance MRI. The cystic/nonenhancing nature of these liver metastases were confirmed by contrast-enhanced ultrasound. Due to diagnostic uncertainty, two of these hepatic cysts were removed laparoscopically and pathology confirmed cystic HPC/SFT with a high MIB-1 index. Previously, in 2014, the patient was diagnosed with solid intracranial grade III pseudopapillary mesenchymal HPC/SFT in the posterior fossa and underwent subtotal resection followed by external beam radiation. In 2017, she had recurrent intracranial, vertebral, and intraspinal intradural extramedullary HPC/SFTs followed by surgery, proton therapy, and SRS radiotherapy. In 2019, after an uneventful pregnancy and birth, routine surveillance revealed metastases in the liver requiring an extended right hepatectomy. In 2020-2021 two solid hypervascular hepatic HPC/SFT were found and treated with microwave ablation. Shortly afterwards, several rapidly growing hepatic cystic HPC/SFT lesions developed. Of note, she has not taken any systemic therapy, indicating the cystic tumors are from metastases rather than cystic degradation as a sequela of therapy. Overall, this case highlights that cystic metastasis are a potential clinical manifestation of solid HPC/SFT. Moreover, cystic HPC/SFT can co-exist with the more typical primary solid hypervascular HPC/SFTs in the same patient. Lastly, in this case cystic HPC/SFT had a higher growth rate and propensity to metastasize as compared to the solid equivalent.

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Introduction

Solitary fibrous tumors (SFT), also referred to as hemangiopericytoma (HPC) if the tumor started in the CNS, is a sub-type of soft tissue sarcoma that affects approximately 0.6 per 1 million people annually in the US (Ratneswaren et al., 2018; Pouchieu et al., 2018; Wang et al., 2020). These tumors can occur anywhere capillaries are present; however, most originate within the cerebral meninges. Prognosis for patients with intracranial HPC/SFT is poor, with a median overall survival (OS) of 13 years (Kubicky et al., 2014). Intracranial HPCs have high rates of local recurrence (87%) and metastases (64%) (Chen et al., 2019). The median OS after the first recurrence is 4.6 years (Galanis et al., 1998; Chan et al., 2012). Distant metastases of intracranial HPC/SFT have been reported in the bones, liver, and lungs (Galanis et al., 1998; Sheehan et al., 2002; Ali et al., 2016).

The first line of treatment against intracranial HPC/SFT is surgery with adjuvant radiation therapy. For surgically inoperable HPC/SFT or post-surgery/radiation recurrences the therapeutic effects of chemotherapy are severely limited (Park et al., 2013). Anti-angiogenic drugs, alone (Park et al., 2013; Lee et al., 2014; Penel et al., 2012) or in combination with temozolomide chemotherapy (Park et al., 2011), have been used on HPCs with limited success. No chemotherapies enable complete remission, with the best response being a partial response or stable disease for months. The average OS of patients on chemotherapies is 2 years (Park et al., 2013). Therefore, more research is needed to improve systemic therapy to treat HPC.

In 2013, researchers discovered that nearly all HPCs/SFTs harbor a hallmark intrachromosomal gene fusion between NAB2 and STAT6 on chromosome 12 (Park, 2013; Robinson, 2013). Research has identified 6 common and up to 40 distinct fusion types that account for pathologic variation and tumor aggressiveness in HPCs (Bieg, 2021; Lee, 2014). Preliminary research suggests that NAB2-STAT6 fusion may promote the cancer via increasing Early Growth Response-1 (EGR-1) and Insulin-like Growth Factor-2 (IGF-2) expression (Pavelić, 1999; Penel et al., 2012) and drive cancer progression(Park et al., 2020).

Intracranial HPC/SFTs commonly present as extra-axial hypervascular solid tumors similar as meningiomas on magnetic resonance imaging (MRI). To date there have been 7 case studies of the unusual cystic presentation of HPC/SFT; three cases in the orbit (Gheorghisan-Galateanu et al., 2019; Feuerman et al., 2010), one intracranial (Clarençon et al., 2007), two pleural (Watanabe et al., 2020; Kishi et al., 2001), and one in the spleen (Gomes and Kothari, 2013). In all cases, the tumor burden was localized so surgical removal was the only treatment without prophylactic radiation or recurrence. Herein we present a unique case of 20+ metastatic cystic HPC/SFTs in the liver. These cystic lesions developed after an extensive history of primary solid intracranial HPC/SFT with recurrence and multiple distant metastatic HPC/SFT throughout her body.

Case presentation

In 2014, a 30-year-old Caucasian female was found to have a HPC/SFT in the right cerebellar pontine angle after presenting with slight hearing loss in the right ear. The details of this patient's cancer history and surgical and radiation treatments can be found in Hayenga et al. (2019). Briefly, after subtotal resection of a lobular 3cm grade III hypervascular pseudopapillary mesenchymal HPC, the patient received 30 fractions, totaling 60Gy, of external beam radiation to the surgical bed. In late 2017, a routine surveillance MRI revealed intracranial progression of residual tumor cells and three spinal cord drop metastasis. Subsequently the patient had gross total resection of the largest spinal cord metastasis, and proton therapy to the surgical bed and other drop metastasis. The intracranial local recurrences along the posterior fossa were treated by gamma knife radiosurgery totaling 18Gy to the 50% isodose line.

Histopathological reports indicate the primary cerebellar pontine angle tumor resected in 2014 was an epithelioid variant HPC/SFT with high mitotic rate (>8 mitoses/10 high power fields), fulfilling the anaplastic HPC, WHO grade III designation. Tumor cells were strongly positive for nuclear STAT6 reactivity, BCL2, CD34, Vimentin, NSF, GRAP, EMA, CAM5.2, CD99, CD57, and CARB ANA-IX. The tumor was negative for S-100, AE1/AE3, CK20, CK7, chromogranin, D2-40, GFAP, PR, SMA, HMB45, MEL A, Synaptophysin, Inhibin, TTF1, ISG15, and CD10. The 2017 spinal cord drop metastasis specimen showed similar morphology. Tumor cells were diffusely positive for STAT6 and vimentin, as well as patchy CD34 reactivity. Tumor cells were negative for meningothelial marker SSTR2 and EMA. Other negative markers include CAM5.2, synaptophysin, inhibin and CD10. The KI-67 (MIB-1) proliferation index was approximately 20%.

Next generation sequencing tests from Tempus, Caris and Foundation One were performed. The tumor cells had the hallmark fusion between NAB2 intron 5 and STAT6 intron 16; which results in NAB2exon6-STAT6exon17 mRNA expression, supporting the diagnosis of HPC. Otherwise, the tumor cells were microsatellite stable with a low tumor mutational burden of 0-0.68 mutations/Mb, PD-L1 negative, and had no clinically targetable features. The DNA repair mechanisms were sufficient as indicated by positive IHC for MLH1, MSH2, MSH6, and PMS2. The patient's tumor had some gene variants of unknown significance (AHSA1 (p.T298I), ELF4 (p.R1775), ERBB2 (p.A440T), ESR1 (p.H6Y), MYH11 (p.V695M), NAB2 (p.G167R), PDGFB (p.H228R), RAP1GDS1 (p.L98V), STAT6 (c.1955+4A), TNFRSF11A (p.P10L)) that may be clinically meaningful in the future. Next-generation sequencing (NGS) reported 76 genes with indeterminate mutational results due to low coverage of some or all the exons (e.g. AXL, BCL10, BCL3, ETV4, EWSR1, FGF3, FLT3, FOXO1, MAP2K4, MYCL, PIK3R2, RUNX1, TAL1, TERT, TRIM33, VEGFB, etc.). The Tempus RNA-seq report aggregated the mRNA molecular profile from the tumor and the patient's germline to provide insights into the patient's tumor biology. Interestingly, the tumor had high mRNA overexpression for AKT2, HER2, FGFR2, and KRAS. However, follow-up histological verification of HER2 surface protein on tumor cells was negative.

In 2018, the patient became pregnant and gave birth to a healthy boy in early 2019. Three months postpartum, the patient was found to have 3 large solid hepatic masses (7.3 cm, 3.0 cm, and 1.5 cm in diameter) and a T12 vertebral body lesion (0.8 cm) by abdominal computed tomography (CT) scan. An extended right hepatectomy was performed for the liver lesions, as well as stereotactic body radiation therapy (SBRT) of 25Gy to the entire T12 vertebral body. Pathology confirmed solid epithelioid HPC/SFT with an immunohistochemical profile of STAT6+/CAM 5.2-/CD34+(patchy), similar to the patient's prior CNS HPCs. The MIB-1 proliferation index was high (approximately 20%).

Between 2020 and 2021, the patient received fractionated proton beam therapy and 7 gamma knife radiosurgeries for multiple CNS drop metastases; as well as microwave ablation therapy for 2 new solid liver metastases. She also has a slow growing 5 mm solid lung mass that is currently being monitored.

In January 2021, a 0.4cm subcapsular cystic liver lesion appeared along the cranial most aspect of segment 2 adjacent to the hepatic vein branch and abutting the diaphragm. It was originally thought to be a benign simple cyst, and thus no treatment action was initiated. A polycystic liver disease panel of 7 genes (GANAB, LRP5, PKD1, PKD2, PRKCSH, SEC63 and ALG8, from Blueprint Genetics) was negative. However, the August 2021 MRI revealed the initial cystic lesion grew to 1.5cm, and at least 15 new cystic lesions were scattered throughout the liver. These cystic lesions were new and not associated with the prior treatment sites Fig. 1. shows contrast ultrasound images, and T1 and T2 weighted abdominal MRI scans of the various lesions. From these imaging modalities an obvious difference between the solid and cystic hepatic HPC/SFT is observed. The hepatic solid HPC/SFT in November 2020 was hypervascular with contrast enhancement on the ultrasound and in the MRI. Conversely the cystic HPC/SFT have no contrast enhancement by

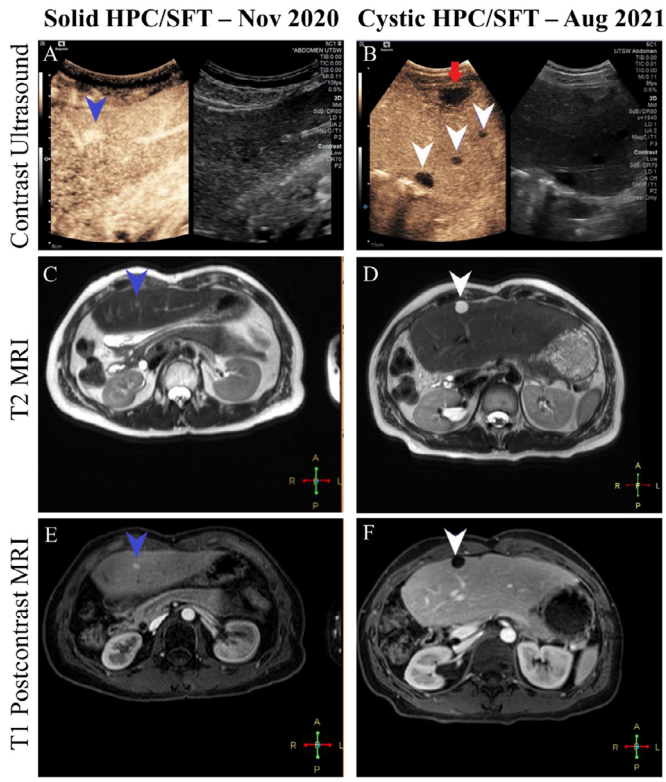

Fig. 1. After a history of solid hepatic HPC/SFTs, several cystic HPC/SFT occurred in the same liver and appeared benign on imaging. Left column labelled, "solid HPC/SFT – Nov 2020" illustrates the initial post extended right hepatectomy metastasis. (A) Hypertense contrast ultrasound indicates the lesion is vascular, (C) hyperintense T2-weighted single shot fast spin echo (SSFSE) indicate the lesion has high fluid content, and (E) hyperintense post-contrast (Eovist) arterial phase T1-weighted Dixon, followed by hypointense washout venous phase (not shown), indicate the lesion is solid and malignant. The right column labeled, "cystic HPC/SFT – Aug 2021" shows several cystic HPC/SFTs that appeared after treatment of the solid hepatic lesions with microwave ablation. On imaging, these cystic HPC/SFTs appear benign. (B) Absent enhancement by contrast-enhanced ultrasound indicates they are avascular without any peripheral enhancement, (D) hyperintense and homogeneous MRI T2-weighted images indicate a high fluid content, and (F) hypointense post-contrast T1-weighted images without peripheral enhancement suggest a benign cystic lesion. The contrast ultrasound, B, was taken after surgery to remove the cystic lesion (in D and F) for diagnostic pathology. Red arrowhead indicates the surgical resection bed. Blue arrowheads indicate solid HPC/SFT lesions. White arrowheads indicate cystic HPC/SFT lesions. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.).

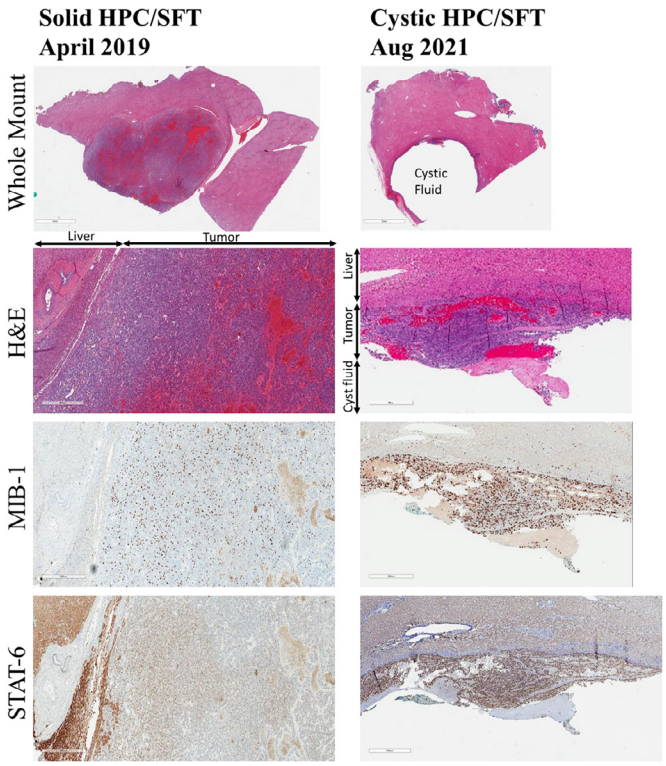

Fig. 2. Histologic features of a solid HPC/SFT and entirely cystic HPC/SFT in the patient's liver. The left column labeled "solid HPC/SFT, April 2019" shows the gross bisected solid tumor and histological stains from the extended right hepatectomy in April, 2019. The right column labelled "cystic HPC/SFT, Aug 2021" demonstrates the H&E, MIB-1, and STAT-6 pathology stains from the surgically excised cystic HPC/SFT in August 2021. Of note, the cystic lesion has a higher MIB-1 proliferation index and only 0.5-1mm layer of tumor cells surrounding a 1.5cm fluid-filled cystic lesion. All scale bars are 500μm, except the whole mounts which are 4mm (solid HPC) and 5mm (cystic HPC).

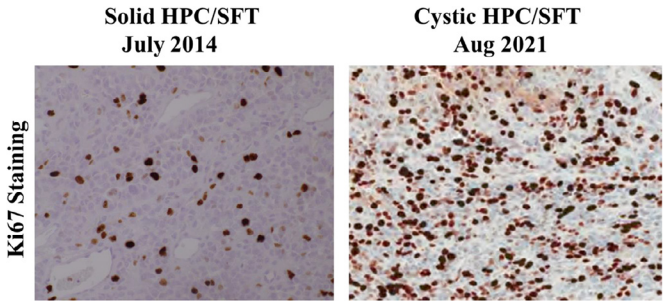

Fig. 3. Proliferation is increased in the cystic HPC. The original intracranial HPC/SFT has an MIB-1 index of 20% (left subfigure). The cystic HPC/SFT has a significantly increased MIB-1 index of at least 30% (right subfigure).

ultrasound or MRI. The lesions do however have bright T2 signal, characteristic of a cyst. Due to the lack of any peripheral enhancement, the cysts were thought to be benign, not tumors. However, new ones appeared, and the cysts were growing rapidly. Therefore, to differentiate the pathology, cystic lesions in segment 3 and 4b were laparoscopically removed. The red arrow in Fig. 1 indicates the surgically excised cystic lesion (as shown in the MRI images below). Pathology confirmed that the cystic lesions were cystic cavities lined by epithelioid tumor cells similar to her prior HPCs, with positive STAT6 nuclear reactivity. The pathology of the patient's 2019 solid liver metastasis and 2021 cystic liver metastasis are illustrated in Fig. 2. Compared to the prior solid HPC, the cystic HPC/SFT had a higher number of MIB+ proliferating tumor cells as illustrated in Fig. 3. For treatment, microwave ablation was used to treat 7 hepatic lesions Fig. 4. demonstrates the 7 target lesions and post-ablation CT showing successful treatment of all these tumors.

Discussion

Incidence and outcome of cystic HPC/SFT

This case study demonstrates an uncommon cystic presentation of a rare cancer. Perhaps more uncommon is that the cystic HPCs were preceded by a 6 year history of multiple solid HPCs. To our knowledge there have only been 7 reported cases of entirely cystic HPC/SFTs. Three cases arising in the orbit (Gheorghisan-Galateanu et al., 2019; Alam et al., 2018; Feuerman et al., 2010), one case intracranial (Clarençon et al., 2007), two cases from the pleura (Watanabe et al., 2020; Kishi et al., 2001), and one case in the spleen (Gomes and Kothari, 2013). In all

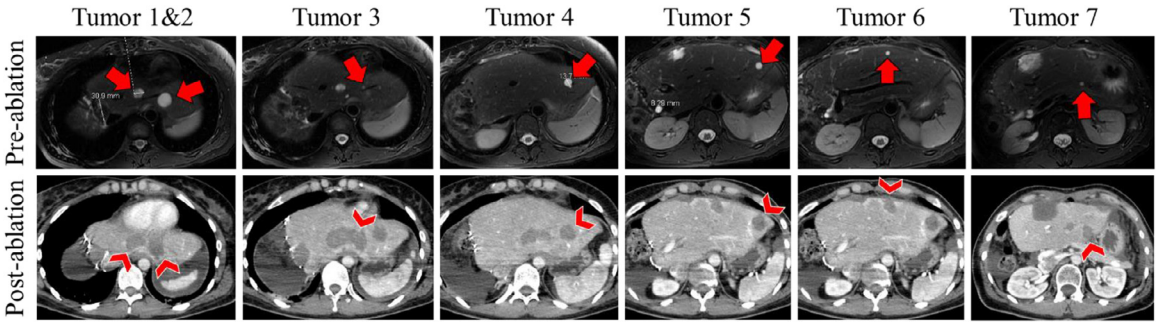

Fig. 4. Microwave ablation treated 7 hepatic cystic HPC/SFT lesions. Pre- and Post- procedural imaging showing successful microwave ablation of 7 cystic HPCs. Arrow indicates the pre-ablation target tumor on the MRI. Arrowhead corresponds with ablation zone for each arrowed tumor noted.

prior cases the cystic HPC/SFT were treated definitively with surgical excision with no local or distant recurrence at the time of publication (which was ~3 months for all case studies except for (Gheorghisan-Galateanu et al., 2019) which had a 15 month recurrence free follow up). However, the female in this case continues to experience cystic recurrences in the liver. Thus, the clinical outcome of patients with cystic, as compared to solid HPC/SFT is unclear. Cystic degeneration of solid HPC/SFTs in response to antiangiogenic treatment or chemotherapy has been observed (Martin-Broto et al., 2019). This is due to a tumor density reduction (as indicated by the Choi criteria) in relation to several factors, such as a decrease in the number of tumor blood vessels. These cystic observations are in response to treatment, however the treatment-induced cystic presentation is different than the de novo cystic tumors presented herein, and in the above mentioned 7 case reports. Of note, the patient has not taken any systemic pharmaceuticals for this cancer.

To our knowledge, this is the first case of multiple cystic HPC/SFT in the liver as a result of distant metastasis. Moreover, to our knowledge this is the first case of both solid and entirely cystic HPCs occurring in the same patient. The mechanisms of cystic formation in HPC/SFT are unknown. Several cystic lesions appeared after the first hepatic microwave ablation treatment; however, this is more likely to be a coincidental correlation since there is no logical explanation for a causative relationship. It has been suggested cystic fluid, in general, can harbor a high concentration of growth factors encouraging growth of tumor cells lining the perimeter (Hazelton et al., 1999; Stockhammer et al., 2000). This could explain the higher proliferation index in cystic HPC/SFTs compared to solid HPC/SFTs.

Treatment for solid and cystic HPC/SFT

The first line of treatment against HPC/SFT is surgical resection, radiation and/or ablation. The patient has undergone an extended right hepatectomy, robotically assisted surgical biopsy resection, 9 microwave ablations, and 2 histotripsy ablations (clinical trial: NCT04572633) to treat the hepatic lesions. If the cancer persists and metastasizes, systemic pharmaceuticals should be considered. Martin-Broto et al. recently published a good review article on the various systemic pharmaceuticals tried on solid HPC/SFT and their limited success rates (Martin-Broto et al., 2021). These include chemotherapies and antiangiogenics (primarily tyrosine kinase inhibitors (TKIs)). Chemotherapies, are typically recommended for dedifferentiated HPC/SFT (a type that may lose STAT6 nuclear immunopositivity) (Martin-Broto et al., 2021; Stacchiotti et al., 2013, 2017). Chemotherapies tried on HPC/SFT include doxorubicin, ifosfamide, dacarbazine and temozolomide, trabectedin and erilbulin with median progression free survival (PFS) ranges between 2 and 12 months. Several antiangiogenics used to treat solid HPC/SFT include temozolomide and bevacizumab, sunitinib, sorafenib, pazopanib, and axitinib with median PFS ranging from 4 to 10 months (Park et al., 2013, 2011; Stacchiotti et al., 2012; Maruzzo et al., 2015).

It remains unknown how cystic HPC/SFT will respond to antiangiogenic or cytotoxic therapies. The higher proliferation rate may make cytotoxic pharmaceuticals, like temozolomide, more effective. Likewise, if growth factors are more concentrated in the cystic fluid, pharmaceuticals that block VEGF, FGFR, PDGF, etc may be effective at slowing the growth as well. RNA sequencing on HPC/SFT shows IGF-2 is orders of magnitude higher than any other factor (Lee et al., 2014; Stacchiotti et al., 2013). Moreover blocking IGF-2 in primary HPC/SFT cells in-vitro reduced proliferation significantly (Penel et al., 2012). Therefore, IGF-2 regulation may provide some benefit. Preferentially expressed antigen in melanoma (PRAME) is also commonly expressed on HPC/SFT and could be considered as an immunotherapy target (Wang et al., 2021). Another potential treatment involves targeting the hallmark NAB2-STAT6 fusion protein (Guseva et al., 2016). Research suggests NAB2-STAT6 is the oncogenic driver (Park et al., 2013; Lee et al., 2014; Penel et al., 2012). Physiologically, wildtype NAB2 functions as a negative feedback loop. That is EGR-1 activates NAB2, NAB2 in turn represses oncogenic EGR-1 target genes (Park et al., 2013). However, in the case of the NAB2-STAT6 fusion, the activation domain of STAT6 replaces the repression domain of NAB2. Therefore, instead of repressing oncogenic EGR-1 target genes, the fusion protein activates them (Stacchiotti et al., 2012). Thus blocking the fusion protein mRNA with an antisense oligonucleotide (ASO) or CRISPR-Cas13 adeno associated virus (AAV) has the potential to reduce the activation effects of the fusion protein.

Attempt to establish a HPC/SFT cell model

There is an urgent need to develop a preclinical model of the tumor type for exploration of therapeutic options for HPC/SFTs. Previously, Ogihara et al. established a HPC/SFT cell line from a canine HPC/SFT tumor. They found several cell types in the culture with different morphologies including multi-nucleated cells, spindle-shaped cells, bipolar and multiple cells in addition to fibroblast-like cells, suggesting the complex nature of the tumor. Recently, a HPC/SFT cell line (HPC3) was established from an intracranial tumor (NAB2ex6-STAT6ex16 fusion type) (Tang et al., 2019). Several mutations were found in the tumor including a mutation in the DSTYK gene (M296Ile) in HPC3. It was shown that the DSTYK mutation promoted cell migration and invasion by activating ERK1/2 signaling pathway in HPC3 cells (Tang et al., 2019).

In this paper, we attempted to establish a HPC/SFT cell line from the patient's surgically resected hepatic cyst. The tumor specimen used had a dimension of 0.2 × 0.2cm and was blue marked by pathologist for tumor orientation. The specimen was minced and cultured in RPMI-1640 medium supplemented with 5% Fetal bovine serum in a humidified atmosphere of 5% CO_2 at 37°C. The culture was not moved for the first 7 days to promote attachment of cells. Subsequently, the medium was changed every week. Unfortunately, there were no tumor cells after 6 weeks of culturing despite the culture conditions similarity to the

condition of the two HPC/SFT cell lines reported previously. The failure to start a cell-line may be due to the limited quantity of tumor cells in the thin layer around the perimeter of the cyst. Alternatively, the blue mark in the specimen may have damaged the cells in the tissue although no such claim had been reported.

Conclusion

We recommend that HPC/SFT be considered in the differential diagnosis of growing cystic hepatic lesions in a patient with an appropriate history. Note that even if the patient has had a history of solid HPC/SFT, cystic HPC/SFT years after the initial resection is still possible.

Patient consent statement

The authors received written, informed consent from the patient to publish this case.

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Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

CRediT authorship contribution statement

Heather N. Hayenga: Conceptualization, Writing – original draft, Project administration. **Chunyu Cai:** Investigation, Data curation, Writing – review & editing. **David Fetzer:** Investigation. **Sarah White:** Investigation. **Joshua Kuban:** Investigation. **Zabi Wardak:** Investigation. **Robert S. Benjamin:** Supervision. **Edward Pan:** Supervision. **James Strauss:** Supervision. **Boning Gao:** Investigation, Writing – review & editing. **John Minna:** Supervision. **Javier Martin-Broto:** Supervision. **J Louis Hinshaw:** Investigation, Data curation, Writing – review & editing.

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