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## CASE REPORT

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# Follicular Dendritic Cell Sarcoma of the Liver

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### ABSTRACT

*Follicular dendritic cell sarcomas are a rare form of malignancy of the follicular dendritic cells. More than half of follicular dendritic cell sarcomas are of nodal origin, while the rest can involve the tonsils, nasopharynx, pancreas, and peritoneal tissues. Follicular dendritic cell sarcoma mimics a number of tumours and tumour-like conditions, and should be considered in the differential diagnoses of a bulky soft tissue mass in relatively asymptomatic individuals. This report is of a patient with follicular dendritic cell sarcomas of the liver and, to the best of the authors' knowledge, is the first reported case in the Asian population. The radiological and pathological findings are described.*

*Key Words: Dendritic cells, follicular; Liver; Sarcoma; Tomography, X-ray computed; Ultrasonography*

## 中文摘要

### 肝臟濾泡樹突狀細胞肉瘤

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濾泡樹突細胞肉瘤是一種罕見的惡性腫瘤。一半以上的濾泡樹突細胞肉瘤為淋巴結來源，其餘的侵犯扁桃體、鼻咽、胰腺和腹膜組織等。濾泡樹突細胞肉瘤與多種腫瘤和瘤樣組織類似；對於臨床發現體積大的軟組織腫塊，但無明顯症狀的病例，應將濾泡樹突細胞肉瘤列為鑑別診斷之一。本文報告首名病灶位於肝臟的濾泡樹突細胞肉瘤的亞洲病人，描述該病相關的放射學和病理特徵。

### INTRODUCTION

Follicular dendritic cell sarcomas (FDCSs) are classified as histiocytic and dendritic cell neoplasms. These tumours are a rare form of malignancy of the follicular dendritic cells. Most patients are adults, and there is no particular sex predilection. More than half of the FDCSs are nodal in origin, predominantly affecting the cervical lymph nodes; 30% are extranodal, involving the tonsils, nasopharynx, pancreas, and peripancreatic and peritoneal tissues.<sup>1,2</sup> One report of FDCS involving the breast has been published.<sup>3</sup>

We report on a patient with FDCS of the liver, and the radiological and pathological findings are described. To the best of the authors' knowledge, this is the first report of FDCS involving the liver in Asia.

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#### Clinical Features

A 47-year-old Chinese woman presented in 2009 with a history of weight loss for 1 month and abdominal distension and discomfort for 2 weeks. The patient had normal bowel opening with no nausea or vomiting or

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Submitted: 26 Jan 2012; Accepted: 11 Jul 2012.

blood or mucus in her stool. An epigastric mass was palpated on physical examination. Complete blood count revealed microcytic hypochromic anaemia with a haemoglobin level of 84 g/l (reference range, 120–150 g/l), elevated platelet count of  $512 \times 10^9$  /l (reference range,  $150\text{--}450 \times 10^9$  /l), and normal white cell count of  $9.4 \times 10^9$  /l (reference range,  $4.5\text{--}11.0 \times 10^9$  /l). Her alfa-fetoprotein level was normal, and hepatitis B and C carrier status was negative.

### Radiological Features

Ultrasound examination (Figure 1) revealed an enlarged left lobe of the liver associated with hyperechoic lesions with dilated vessels and ascites, as well as an incidental finding of gallstones and gallbladder polyps. The hyperechoic lesions appeared ill-defined with dilated vasculature, and no haemorrhage, fat, or calcifications were seen within the lesions. Subsequent computed tomography (CT) showed a  $16.4 \times 9.5 \times 8.0$ -cm heterogeneous hypoattenuating lesion with intense arterial enhancement (Figure 2), and confirmed the presence of abnormally dilated vessels and aneurysms, suggestive of a hypervascular lesion. There was no haemorrhage, fat, or calcifications within the lesions. A moderate amount of ascites was noted. The kidneys were unremarkable. Systemic staging examination (positron emission tomography–CT) was not performed.

### Histopathology

During hepatectomy, a  $16 \times 9$ -cm vascular tumour was seen astride segment 2/3 of the liver, and 4 litres of peritoneal fluid with no definite peritoneal or omental deposits was noted. Cytology of the peritoneal fluid showed no malignant cells.

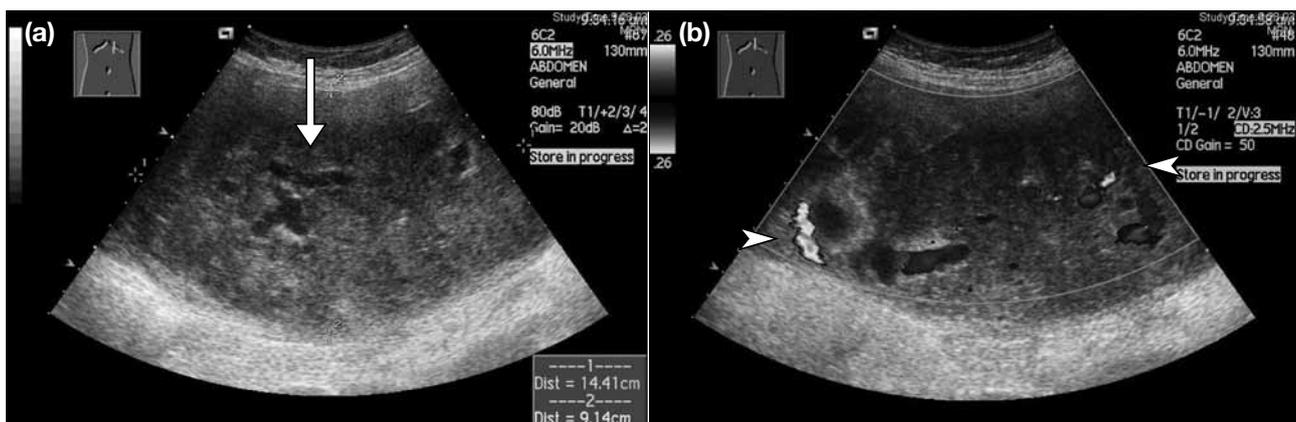
Macroscopic examination of the vascular tumour revealed a tumour with pushing margins and heterogeneous brownish surfaces with areas of haemorrhage. Medium-sized thick-walled prominent blood vessels were present within the tumour and were associated with haemorrhage, which could have been the abnormally dilated vessels and aneurysms seen on ultrasound and CT.

Microscopic examination revealed tumour cells with proliferation of spindle cells arranged in whorls, fascicles, diffuse sheets, and a focally storiform pattern. The cells also displayed poorly defined borders and eosinophilic cytoplasm, associated with moderate nuclear pleomorphism, and coagulative necrosis. Mitotic figures were seen at  $<1$  per 10 high-power fields (HPFs). No peritumoural vascular invasion was seen. Dense lymphoplasmacytic infiltrates obscuring the spindle cells were noted. No features of Castleman's disease were found. No atypical lymphoid cells were seen.

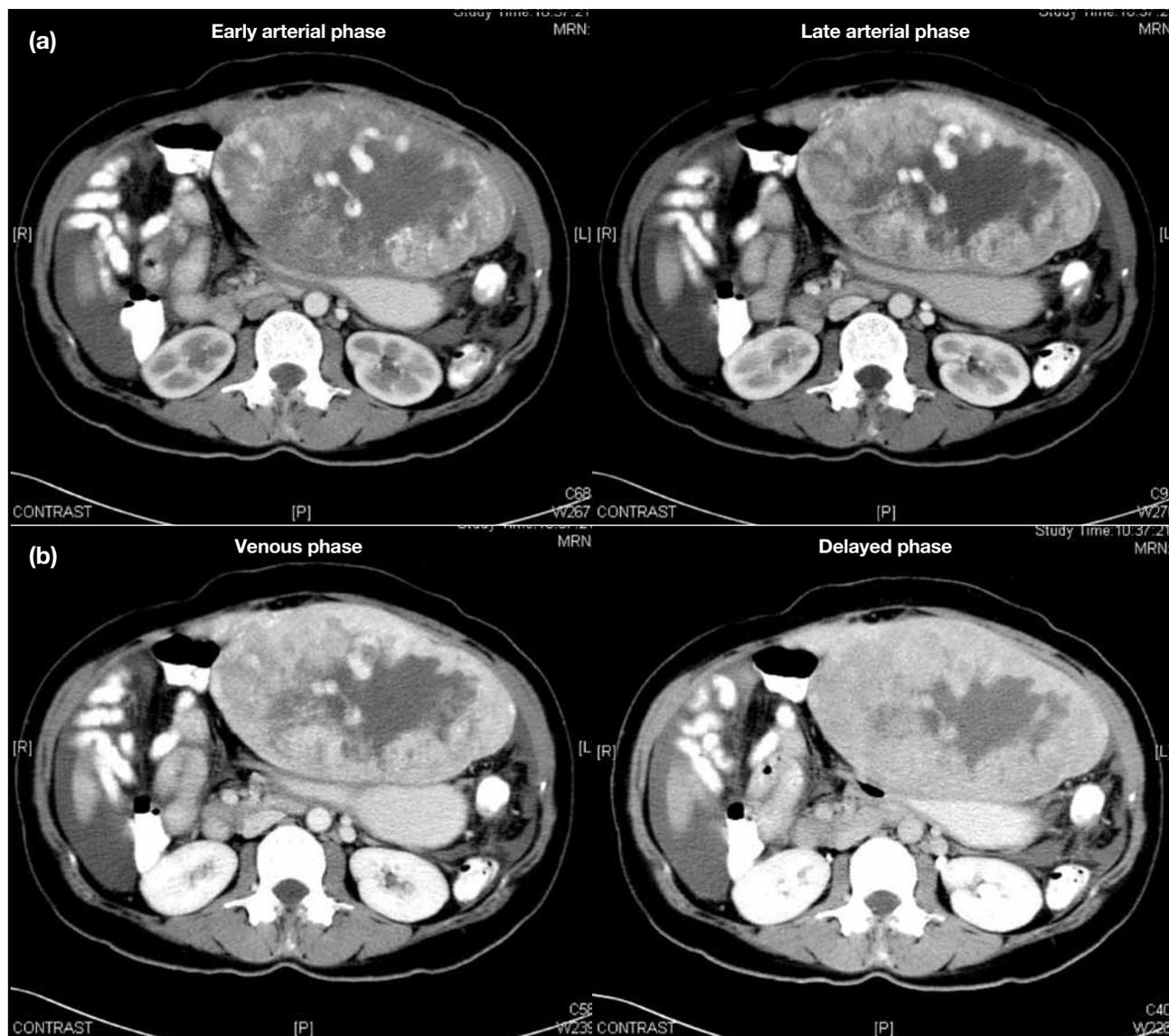
Immunohistochemical findings were diagnostic of FDCCS according to the special stains. In particular, the spindle cells were positive for follicular dendritic cell markers, and negative for actin, anaplastic lymphoma kinase, and hepatocyte paraffin 1 (Her Par 1). Staining with CD3, L26, kappa, and lambda showed unremarkable lymphoid cell distribution with no obvious lymphoid proliferation.

### Patient Outcome

The patient underwent partial hepatectomy. She was offered close monitoring and follow-up for over 2 years.



**Figure 1.** Ultrasound images of follicular dendritic cell sarcoma of the liver (a) show an enlarged left lobe of the liver associated with hyperechoic lesions (arrow) — the hyperechoic lesions appear ill-defined with dilated vasculature, and no haemorrhage, fat, or calcifications within the lesions; and (b) a Doppler image showing multiple dilated vessels (arrowheads).



**Figure 2.** Computed tomography images of follicular dendritic cell sarcoma of the liver. (a) Early and late arterial phases show intense arterial enhancement, and confirming the presence of abnormally dilated vessels and aneurysms (arrow); and (b) venous and delayed phases of the tumour.

However, 18 months after the surgery, she was found to have a local recurrence requiring further surgery for resection.

**DISCUSSION**

FDCS is a rare neoplasm of follicular dendritic cells. Follicular dendritic cells are the antigen-presenting cells of B-cell follicles in lymph nodes and extranodal lymphoid tissue.<sup>4</sup> Patients are usually asymptomatic, presenting with a slow-growing painless mass. Nearly 10 to 20% of cases are associated with a hyaline-vascular variant known as Castleman’s disease, a

tumour precursor of hyperplastic or dysplastic follicular dendritic cells.

Macroscopically, FDCS is similar to other sarcomas, with a well-circumscribed tan-grey cut surface and areas of necrosis and cystic change in larger tumours. Histology often reveals spindle and ovoid cell proliferation forming fascicles and a storiform pattern with whirling. In this patient, the tumour had pushing margins and heterogeneous brownish surfaces. Medium-sized thick-walled prominent blood vessels were present within the tumour and were associated

with haemorrhage. This correlated with the CT findings of a heterogeneous hypoattenuating hypervascular lesion with abnormally dilated vessels and aneurysms. Preoperative biopsy is not recommended given the hypervascular nature of the lesion.

Pathologically, tumour cells have been shown to have eosinophilic cytoplasm and indistinct cell borders. The nuclei are usually elongated with vesicular or finely granular chromatin and distinct nucleoli. Although most cases are cytologically bland with a low mitotic rate (0-10 per 10 HPFs), cases with greater cytologic atypia and higher mitotic rates may be seen. Often there is a sparse infiltrate of mature lymphocytes and plasma cells, predominantly in a perivascular distribution.

Awareness of FDSC is important as the tumour closely mimics other tumours and tumour-like conditions. Even with immunohistochemical examination, the diagnosis may be missed as follicular dendritic cell markers are often not included in the routine panel of antibodies used for investigation of undifferentiated neoplasms. At least seven cases of extranodal FDC sarcomas of the head and neck were initially misdiagnosed as inflammatory pseudotumour, ectopic meningioma, malignant schwannoma, poorly differentiated carcinoma, fibrous histiocytoma, carcinoma showing thymus-like elements, and acinic cell carcinoma. The key histological feature suggestive of FDSC is the perivascular distribution of small lymphocytes within the tumour. All cases need confirmation by immunohistochemistry with specific stains for FDSC.

The immunohistochemical profile shows immunoreactivity for markers specific for follicular dendritic cell differentiation: CD21, CD23, and CD35. The cells are usually positive for vimentin, fascin, human leukocyte antigen-D related, and epithelial membrane antigen (the latter causing confusion with meningioma and carcinoma), and variably positive for S100 and CD68. Staining for CD1a, lysozyme, CD34, CD3, CD79a, CD30, human melanoma black-45, and cytokeratin is consistently negative. The small lymphocytes are predominantly T or B cells. Tumours arising in the liver and spleen often show only weak or

focal positivity for follicular dendritic cell markers.

FDSC is treated by surgical excision, with or without adjuvant chemotherapy. Local recurrences occur in about 40 to 50% of cases and metastases occur in 25% on follow-up. This patient developed local recurrence 18 months after initial surgery. Given the aggressive nature of the tumour, closer monitoring is suggested. Cases with intra-abdominal presentation, higher cytological atypia, or greater mitotic activity are associated with a more aggressive clinical course. A patient with FDSC of the breast that demonstrated high-grade histological features (necrosis, mitoses) followed an uneventful clinical course of up to 19 years following initial treatment.<sup>3</sup> At least 10 to 20% of patients die of the disease, often after a long period of time.

## CONCLUSION

This report describes the first case of FDSC involving the liver in Asia, an uncommon soft tissue tumour of follicular dendritic cells of nodal or extranodal origin. FDSC, although unusual, should be considered in the differential diagnoses of a bulky soft tissue mass in relatively asymptomatic patients as FDSCs mimic other tumours and tumour-like lesions. A confirmed diagnosis involves immunohistochemical investigations. Treatment is primarily surgical, with a high recurrence rate and a long disease course.

## DECLARATION

No grants have been received with regard to this study.

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