# Splenic Littoral Cell Haemangioendothelioma: Case Report

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## *Background*

Littoral cell haemangioendotheliomas (LCHE) are vascular tumours that originate from littoral cells lining splenic red pulp sinuses with intermediate features between benign littoral cell angioma and frankly malignant angiosarcoma. Only scanty case reports of LCHE are available in literature, including those of minimal atypical histological features, but presented with disseminated disease or with late recurrence or metastasis. We present herein a local case of LCHE.

## *Case report*

The patient was a 67-year-old female who presented with abdominal discomfort, weight loss and splenomegaly. Blood investigations showed anaemia and leukocytosis. Computer tomography with contrast revealed marked splenomegaly with reduced splenic enhancement. There were scattered hyperdense haemorrhagic areas in the spleen, and small peripheral splenic hypodense regions with mild rim-enhancement. The peripheral splenic vein was poorly enhancing suggesting thrombosis. Splenectomy was subsequently performed. Grossly, the spleen was enlarged to 17.0 x 11.5 x 7.5 cm and weighed 675 gm. Cut surface showed that the well-capsulated spleen was occupied by a firm red tumour, measuring 17.0 x 11.0 x 7.0 cm, with vaguely multinodular appearance and multiple focal necrosis.

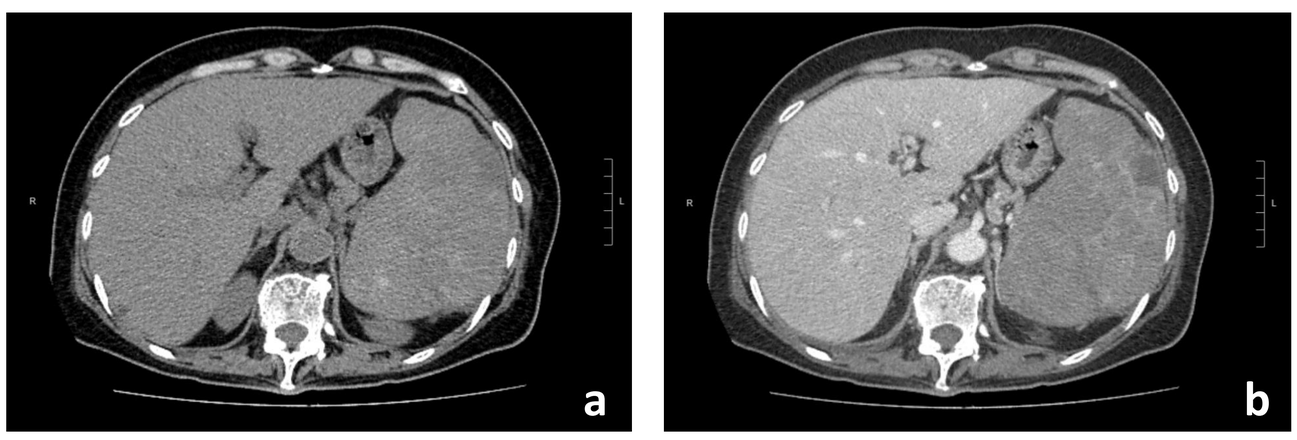


Figure 1. Computer tomography plain (a) and with contrast (b) showing the spleen.

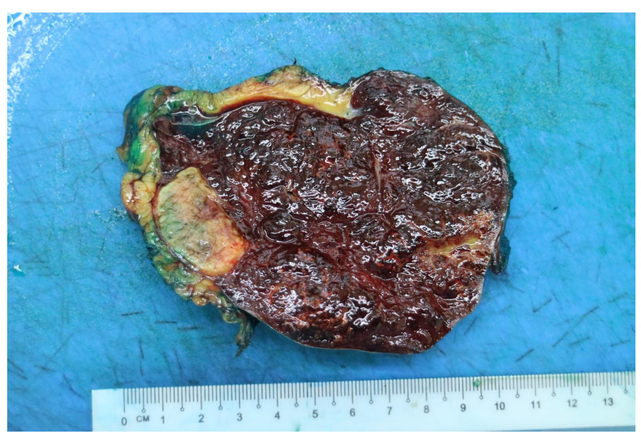

Figure 2. Cut surface of the spleen.

Histological examination shows the spleen is almost completely replaced by a vascular neoplasm which exhibits vaguely nodular growth with permeative borders. The tumour comprises spindly cells forming slit-like or sinusoidal blood vessels. The tumour cells possess obviously atypical nuclei with varying fine to hyperchromatic chromatin, focal distinct nucleoli and exhibit mitotic activity (up to 4 mitoses/10 high power fields). Scattered degenerative bizarre cells are also seen. On immunostaining, the neoplastic spindle cells are CD31+, CD8+(some), ERG+(some), HHV8- and CD34-. Rich population of perivascular myoid cells are demonstrated by SMA. The overall features are consistent with LCHE. There is no recurrence or metastasis in a one-year follow-up.

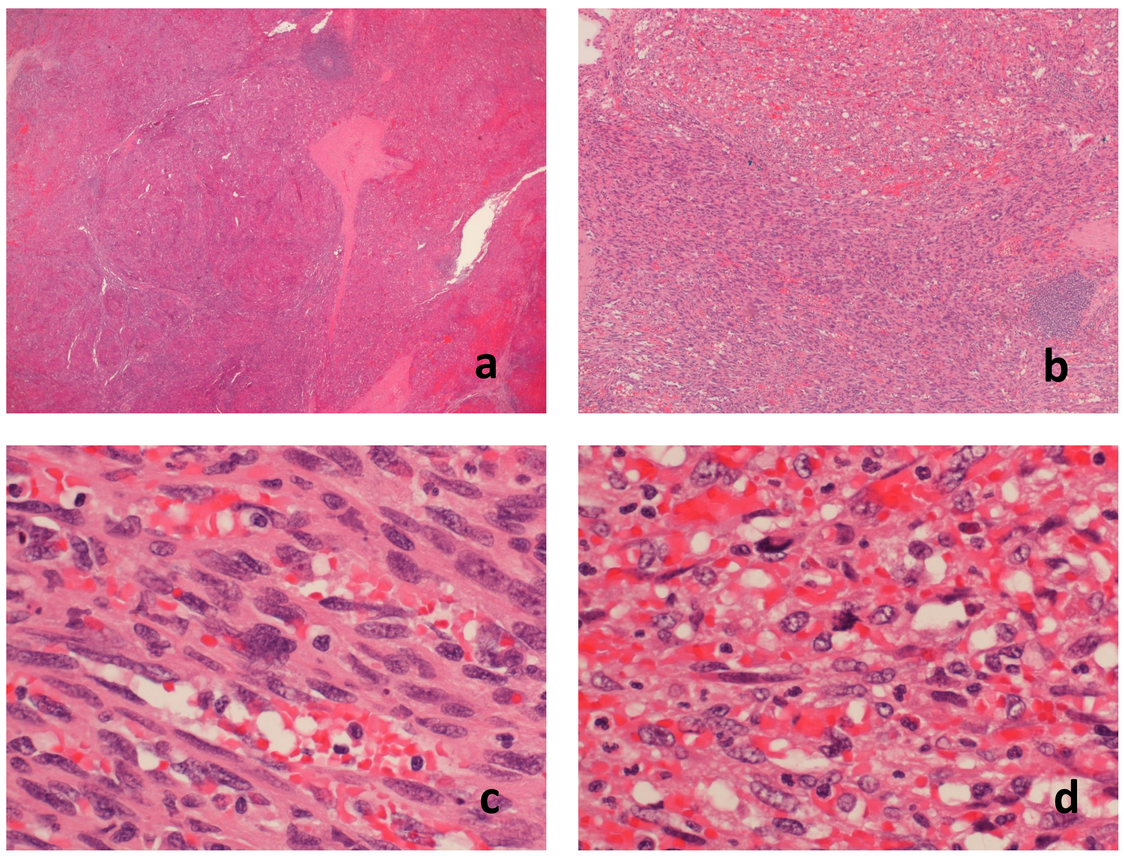


Figure 3. Microscopic features: vaguely nodular growth (a, 10x); sinusoidal (upper) and slit-like (lower) blood vessels (b, 40x); cytology (c & d, 400x).

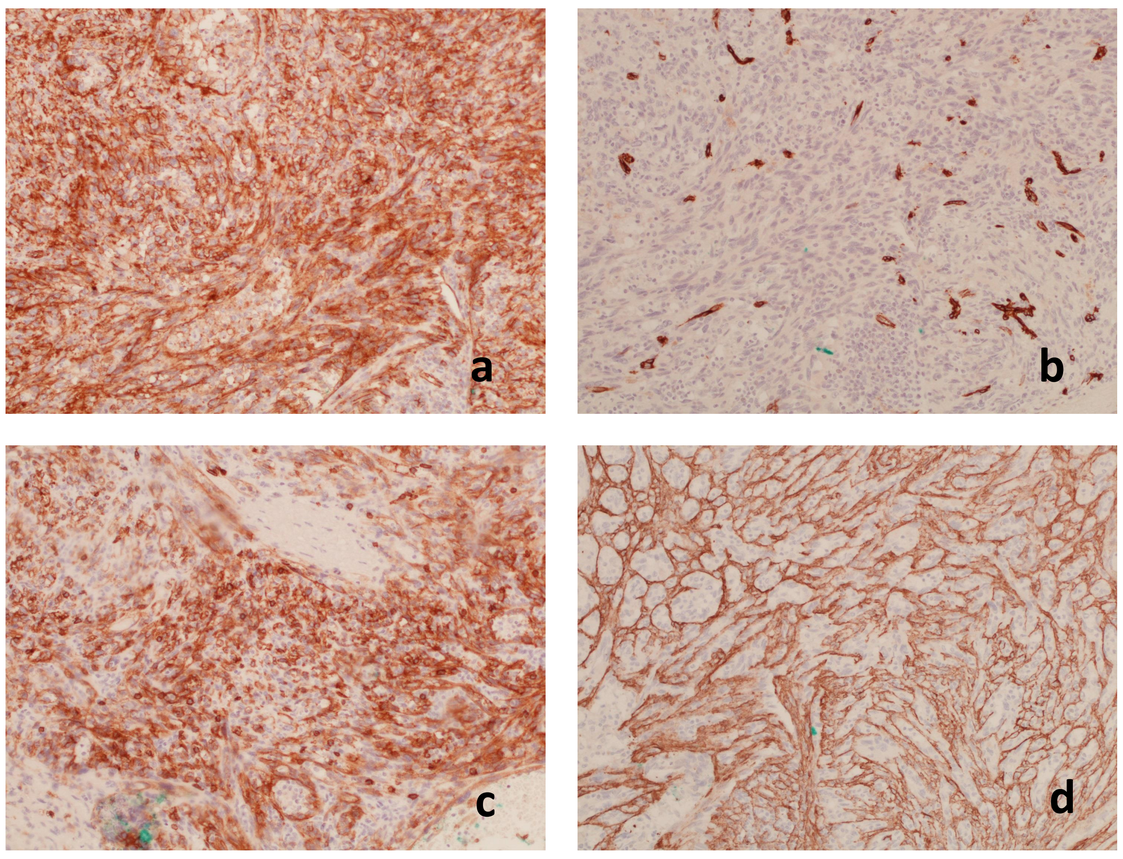


Figure 4. Immunostaining for CD31 (a), CD34 (b), CD8 (c) and SMA (d).

## *Discussion*

In the normal splenic red pulp, three distinctive types of blood vessels are present: sinusoids (CD8+, CD31+, CD34-), capillaries (CD8-, CD31+, CD34+) and small veins (CD8-, CD31+, CD34-). In splenic hamartoma and sclerosing angiomatoid nodular transformation, all three types of blood vessels are present. Splenic haemangioma and lymphagioma show a usual endothelial immunophenotye (CD8-, CD31+, CD34+). Littoral cell angioma is CD8-, CD31+, CD34-, CD68+, CD163+ and CD21+, while angiosarcoma is CD8-/+, CD31+ and CD34+. In addition, angiosarcoma lacks perivascular myoid cells. Kaposi sarcoma is associated with human herpesvirus 8 infection and stains positive for HHV8. The present case adds another example of rare LCHE. Considering the indeterminate behavior, long-term follow-up is deemed necessary, especially in view of the high-grade morphology.

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