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Case report of a littoral cell angioma of the spleen and accessory spleens: A benign vascular tumour



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ABSTRACT

INTRODUCTION: Littoral- cell angioma (LCA) is a rare benign vascular tumour of the spleen. There have been less than 80 cases reported in the literature.

Recent reports have described it to be a malignant lesion with congenital and immunologic associations. We report a case of LCA of the spleen.

PRESENTATION OF CASE: A 52-year-old male patient was admitted to hospital with a three month duration of intermittent upper abdominal pain and nausea. Imaging studies, including computer tomography (CT) and magnetic resonance imaging (MRI), showed multiple lesions in the spleen as well as in the accessory spleens.

An open splenectomy was performed and his post-operative recovery was uneventful.

DISCUSSION: Littoral cell angioma of the spleen is a benign vascular tumour that has been infrequently reported in the English literature. While it does have malignant potential, the vast majority are benign. Diagnosis depends on the expression of endothelial markers like CD31 and histiocytic markers like CD68. Malignant potential is enhanced by the presence of splenomegaly as well.

CONCLUSION: This rare condition is made even more rare by the presence of the tumour in the two accessory spleens as well.

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1. Introduction

Littoral cell angioma (LCA) of the spleen is a rare benign vascular tumour. Since the initial report by Falk et al. in 1991 there have been less than 80 cases reported in the literature [1,2].

This is a case report of a LCA of the spleen and accessory spleens in a 52 year old male. His abdominal symptoms included early satiety and pain.

An open splenectomy effected resolution of his symptoms. His recovery was uneventful. This case report has been reported in line with the SCARE criteria [3].

2. Case report

Mr. WS is a 52 year old male who was referred with a complaint of abdominal pain of three month's duration. The pain was located in his left upper quadrant. Other complaints included early satiety and nausea. There was no history of trauma and no food pain association or change in bowel habits.

He had no constitutional symptoms such as fever, fatigue or weight loss and his medical history included hypertension and an elevated cholesterol level. Medications included lovastatin, atenolol and amlodipine for his high cholesterol and hypertension.

He did not smoke or drink.

Physical examination revealed an enlarged spleen extending to the umbilicus.

He had no clinical signs of peritoneal irritation. His complete blood count showed a leukocytosis while his haemoglobin and platelet counts were within normal limits. This leukocytosis resolved with surgical resection.

An ultrasound done at the referral hospital showed splenomegaly. This was confirmed by computerized tomography (CT) and magnetic resonance imaging (MRI) (Figs. 1–3). There was no hepatomegaly or lymphadenopathy. There was also an accessory spleen in the splenic hilum with the same benign vascular tumours.

A percutaneous trucut biopsy confirmed the diagnosis of a littoral cell angioma.

This was performed by an internist prior to his surgical referral.

After an extensive discussion with the patient and his family he decided on surgical treatment and an informed consent was taken. An open splenectomy was performed given the large splenic parameters. A laparoscopic procedure was discussed with the patient but given the size of the spleen it was decided against.

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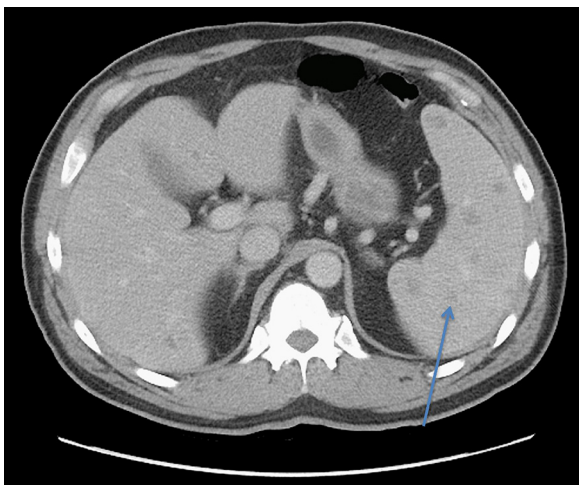


Fig. 1. CT scan with splenomegaly (blue arrow).

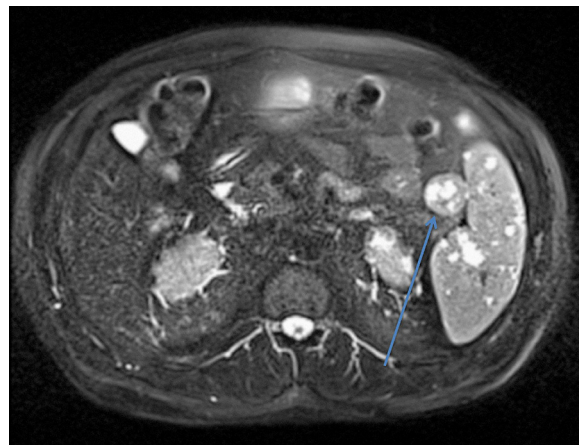


Fig. 3. MRI (T1) Angiomas in the accessory spleen. (blue arrow).



Fig. 2. CT scan saggital view showing the splenomegaly.

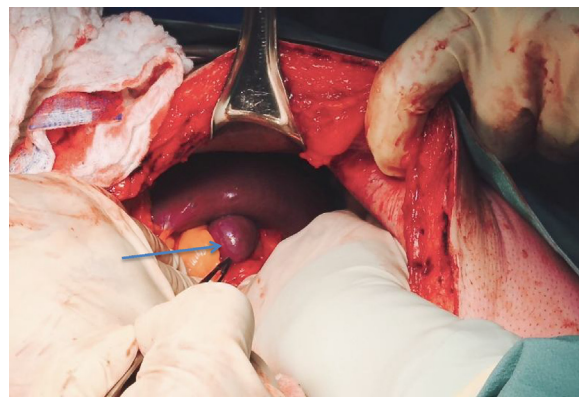


Fig. 4. Accessory spleen in the splenic hilum (blue arrow).

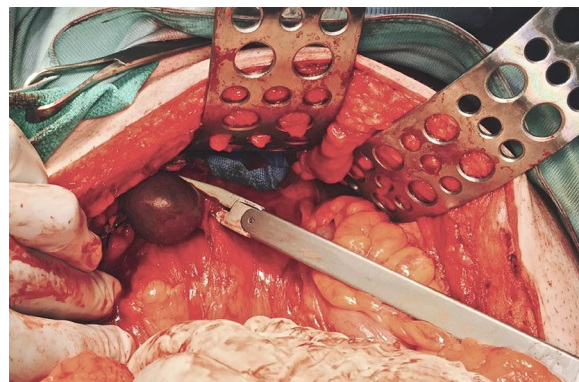


Fig. 5. Accessory spleen excision.

Surgery was uneventful and he was discharged home after five days (Fig. 4 and 5). Pathology confirmed a littoral cell angioma of the spleen (Figs. 6–9) as well as in the two accessory spleens.

3. Discussion

Littoral cell angioma (LCA) initially described by Falk and colleagues is a rare benign vascular tumour of the spleen [4].

The endothelial cells which line the sinus channels in the splenic red pulp are thought to be precursors of the tumour.

50% of all patients present clinically with splenomegaly or signs of hypersplenism like anemia or pancytopenia [4].

Endothelial markers like CD31 as well as histiocytic markers such as CD68 and lysozyme is thought to be pathognomic of LCA [4–6] and establishes the pathological diagnosis.

The pathological diagnosis can vary between the benign littoral cell angioma (LCA), the potentially malignant littoral cell haemangioendothelioma(LCHE) [1] and the malignant littoral cell angiosarcoma(LCAS) [7]. LCA remains the commonest variant. The etiology remains nebulous although there is an association with



Fig. 6. Excised spleen with two accessory spleens demonstrating spongy blue sub-scapsular tumours (blue arrows).

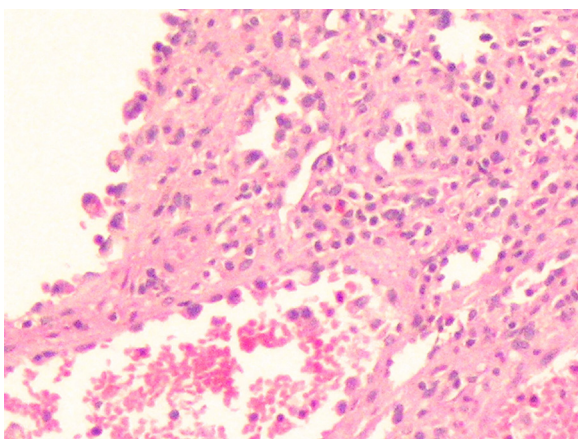


Fig. 7. Medium power photomicrograph demonstrating irregular vascular channels lined by hobnail epithelial cells.

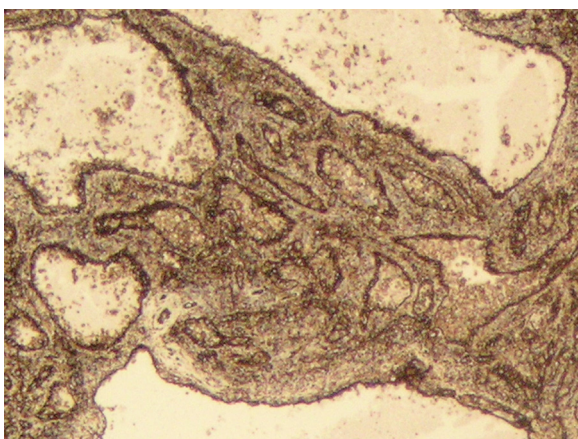


Fig. 8. Antibody to CD31 decorating the littoral endothelial cells of the angiomas.

visceral organ tumours and immune-mediated diseases in one-third of the eighty reported cases to date [8].

Splenomegaly of greater than 10 cm is significantly associated with the presence of malignancy [9].

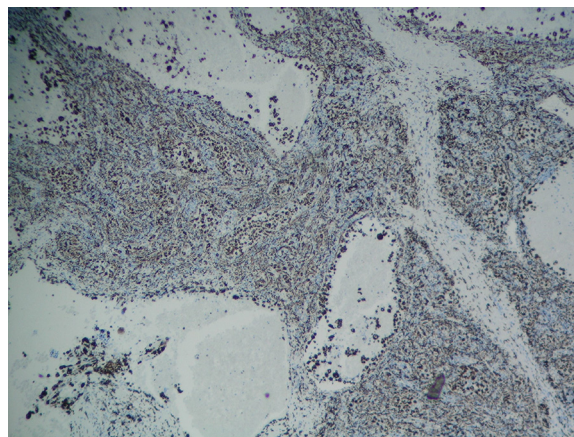


Fig. 9. Antibody to CD 68 demonstrating dual staining of the littoral endothelial cells and macrophages in the vessel lumina.

While causes of splenomegaly such as lymphoma are common it behooves us to keep in mind the rare causes of malignant tumours of the spleen in our clinical armamentarium.

A rare benign vascular lesion [10], this condition is made even more rare by the presence of the tumour in the two accessory spleens as well. A recent literature search of pubmed and medline by the authors shows this to be the first case report with the LCA in the two accessory spleens as well. There have been two case reports with LCA in the English literature with one accessory spleen but no reports in the English literature with two accessory spleens. We believe this to be the first such case report. This only adds to the rarity of this condition.

Conflicts of interest

No conflicts of interest.

Funding

No known funding sources.

Ethical approval

Not applicable as it is a case report and not a research study.

Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Author contribution

Yagan Pillay (YP): Wrote the article including a literature search and collected the data and photos. Agrees to be responsible for all aspects of the work. He also performed the surgery on the patient.

M. Omar Shokeir (MOS): Provided the pathology slides and read the draft manuscript. He was involved in proof reading the final draft for publication.

Guarantor

Yagan Pillay is the guarantor.

Acknowledgement

Not applicable.

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