Littoral cell angioma disguised as huge ovarian cyst: A thought to ponder

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SUMMARY

Littoral cell angioma (LCA) of the spleen is a rare vascular tumour. It was thought to be a benign and incidental lesion. Given the relative lack of specific symptom in many cases, these tumours are found incidentally during abdominal surgery during a non-related procedure. Clinical manifestation of a huge adnexal mass and the atypical appearance of splenomegaly, explains the often misguided diagnosis. To the best of our knowledge, there has been no case of incidental finding of LCA disguised as a huge ovarian cyst in Malaysia. We present a case report of a focal solitary LCA incidental finding after splenectomy.

INTRODUCTION

Vascular tumours are the most common primary neoplasms of the spleen. Among these, littoral cell angioma (LCA) is a rare vascular tumour that occurs exclusively in splenic tissues and was first described by Falk et al. in 1991.[1] Due to its rarity, it may be mistakenly diagnosed as a benign cystic lesion, particularly resulting from a gynaecological disorder. In this paper, we present a case of LCA with clinically manifested as an adnexal mass in Hospital Sultanah Nora Ismail, Batu Pahat, Johor, Malaysia.

CASE REPORT

A 15-year-old young woman with no known premorbid illness was seen in Gynaecology Clinic at Hospital Sultanah Nora Ismail, Batu Pahat, Johor, Malaysia. The patient came with a two week history of abdominal distension and constipation. She also had a frequency of micturition. She denied any gynaecology symptoms and attained menarche at the age 12. During physical examination by our Gynaecologist, her abdomen was found to be distended with clinically manifested as huge adnexal mass with 20-week size, soft and non-tender. No hepato-splenomegaly was noted during during percussion. Bedside ultrasonography examinations by the gynaecologist revealed an uniloculated ovarian cyst with the size of 10x14cm with no significant abdominal free fluid. Ultrasonography studies done during the 2 week follow up was consistent with the first ultrasonography finding and normal uterus visualised anteverted position, sized 4x5cm. Her laboratory tests showed hypochromic microcytic anaemia. Her full blood picture had a non-specific finding. Her tumour marker shows elevated in CA125, but Alpha-fetoprotein and LDH were normal.

She was diagnosed with a symptomatic huge ovarian cyst and to ruled out malignancy. She was then scheduled for elective cystectomy keeping in view salphingo-oophorectomy.

Intra-operative findings noted a large splenic tumour occupying the inferior pole of the spleen. Uterus and both ovaries were normal. The case was subsequently referred to the surgical team during the operation. Splenectomy was performed. On gross pathological examination, a focal solitary mass of the anterior splenic capsule measuring 17×18cm with clear margins was found. Histological examination of the pathological specimen from the splenectomy specimen showed a low-grade vascular tumour compatible with LCA. A baseline contrast-enhanced CT scan of the thorax, abdomen and pelvis was done postoperatively and showed unremarkable findings. She was then given triple vaccination and was started on prophylactic penicillin until the age of 21 years old. She was under our surveillance follow up 6 monthly.

DISCUSSION

LCA of the spleen is a rare vascular tumour that was first described in 1991. LCA may occur at any age and have no gender predilection. However, in the original paper Falk et al., state that LCA affects both men and women (female: male ratio = 5:3) with no specific age predilection (1-77 years, median age: 50 years).[1] Majority of patients with LCA are usually asymptomatic and the lesion is discovered incidentally as in the case with our patient. Typically, patients with LCA are found to have a splenic abnormality, in which 50% of all patients present with signs of hypersplenism such as anaemia or pancytopenia. Other systemic symptoms such as fever, chills, weakness, fatigue, and pain have been reported.[1] Given the relative lack of specific symptoms, in many cases, these tumours are found incidentally during abdominal surgery during a non-related procedure. However, most of the cases were incidentally found during emergency settings, such as traumatic solid organ injuries and ruptured haemorrhagic ovarian cysts.[2,3]

In our case, LCA was clinically manifested as a huge adnexal mass causing abdominal distention and external compression symptom such as constipation. Considering the atypical manifestation of splenomegaly from gross pathological examination, that might explain the misguided diagnosis pre-operatively by our Gynaecologist. The gross

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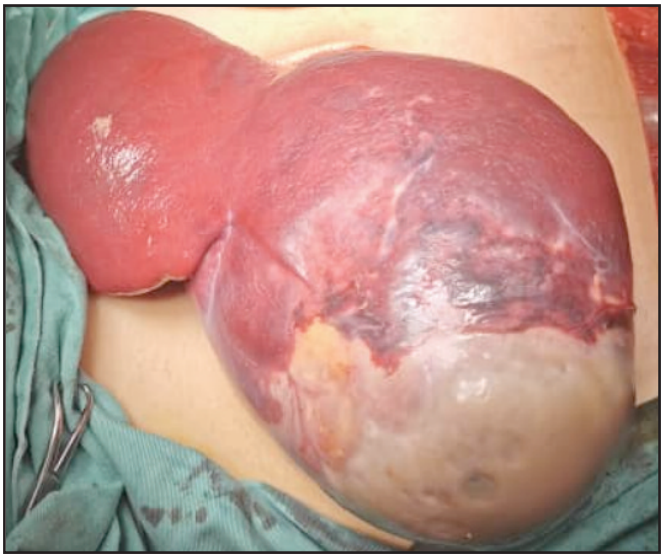

Fig. 1: Gross pathological examination shows uneven splenomegaly.

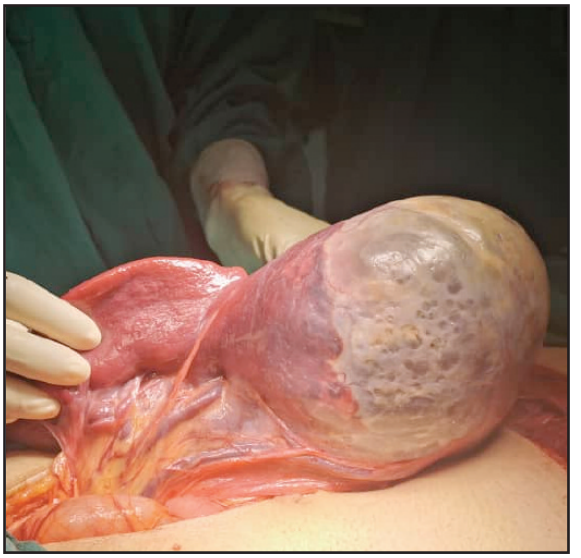

Fig. 2: Large splenic tumour occupying inferior pole of the spleen.

specimen showed a normal appearance of the superior pole and a huge tumour from the inferior splenic pole. Thus, leading to negative Traube's space during the examination finding.

Therefore, the case was referred by a primary health care team to evaluate the ovarian mass. The patient was seen first by our gynaecologist and transabdominal ultrasound were done twice. Based on the Royal College of Obstetrician and Gynaecologists Green-top guideline, a pelvic ultrasound is the single most effective way of evaluating an ovarian mass with transvaginal ultrasonography are preferred due to its higher sensitivity over transabdominal ultrasound. However, considering that the patient is a virgin adolescent, transvaginal ultrasonography was not performed. In addition, at present, the routine use of Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) for the assessment of ovarian masses does not improve the sensitivity or specificity obtained by transvaginal ultrasonography in the detection of ovarian malignancy.[4] Nevertheless, in our case, a decision for cystectomy kept in view salphingo-oophorectomy was made for symptomatic relieve and need for tissue histopathological examination evaluation to confirm diagnosis.

On ultrasound, the findings for LCA or splenic tumours vary widely diversified, from heterogeneous echotexture without specific nodules to hyperechogenic, hypoechogenic or isoechogenic appearing lesions. However, the spleen has traditionally been regarded as an orphan organ.[5] In our case, ultrasound was misleading, suggesting a huge ovarian mass with uniloculated ovarian cyst.

Nonetheless, from the general surgery point of view, in most cases involving an abdominal mass, CT and MRI are adequate to establish a confident diagnosis. MRI is the best radiological imaging method to differentiate between LCA and other angiomatous vascular lesions [5]. Ultrasonography quality is operator dependent and subjective to interpretive error thus we must also recognize the clinically relevant limitations and pitfalls associated with the use of ultrasound. A definitive diagnosis of LCA requires histological examination. The differential diagnosis of lesions that can mimic LCA on imaging includes lymphangioma, hamartoma, lymphoma, Kaposi's sarcoma, and haemangioma. On gross examination at histopathology, an LCA will typically reveal splenomegaly. Macroscopically, splenic involvement is characterized by multiple, spongy, cystic blood-filled, circumscribed nodules. The nodules are often multifocal similar in size and are well delineated from the surrounding splenic tissue but do not have a surrounding capsule. It is characterised histologically by anastomosing vascular channels lined by tall or flat endothelial cells, which may anastomose with normal splenic sinuses at the periphery.[5]

However, our case showed atypical splenomegaly and a focal solitary mass measuring 17×18 cm of the anterior splenic capsule with clear margins. To the best of our knowledge, this shows an unusual presentation of splenomegaly that can mislead a diagnosis. Bhatt S et al. also stated that, less commonly, these lesions can be solitary or completely replace the splenic parenchyma. Therefore, a definitive diagnosis can only be obtained pathologically.[5]

Although this is a benign tumour, surgical removal is always indicated. Treatment for symptomatic LCA is splenectomy, but currently this is not recommended for asymptomatic LCA. Due to the association and reported cases of malignant transformation, the option for splenectomy should be considered. In our case, a decision was made for splenectomy made on table during surgery due to incidental findings by the gynaecology team.

CONCLUSION

Thus far, this is the first case of LCA presented as an incidental finding splenic tumour disguised as a huge

ovarian cyst reported in Malaysia. We believe that further imaging workup and multidisciplinary approach might be helpful to build up a harmonious environment and better healthcare service. A definite diagnosis is difficult to make preoperatively because histological examination is the only accurate means of making this diagnosis.

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