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Case Report Open Access

Lymphatic Malformations: A Rare Case of a Gigantic Mesenteric Lymphatic Malformation in a Paediatric Patient

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Introduction

Lymphatic malformations are considered very rare malformations with their incidence being around 1:4,000 [1]. More commonly they present in the neck historically described as cystic hygromas. Ninety percent of cases are diagnosed by 2 years of age. Presentation of a mesenteric lymphatic malformation has an incidence of 1:100,000 [2].

Case Report

A 14 year old Caucasian boy presented with a 24 hour history of lower abdominal pain with nausea and anorexia.

On examination he was tender in his right iliac fossa with guarding. His bloods were unremarkable except for a slightly raised C reactive protein and urinalysis was clear. A clinical diagnosis of appendicitis was made and the boy underwent a laparoscopy.

Intra-operatively he had a normal looking appendix but a large cystic structure housed within the abdomen.

Post-operatively further investigations were performed and an ultrasound revealed a large intra-abdominal, cystic structure, with multiple septations (Figure 1).

An MRI done showed a large, multi-septated, gigantic lymphatic malformation of unknown origin (Figure 2).

The tumour markers in particular AFP and ALP were normal.

A laparotomy was performed and the lymphatic malformation was removed in toto with its origin located at the third part of the duodenum (Figure 3).

Histopathology reported the lesion as a gigantic mesenteric lymphatic malformation with no evidence of malignancy (Figure 4).

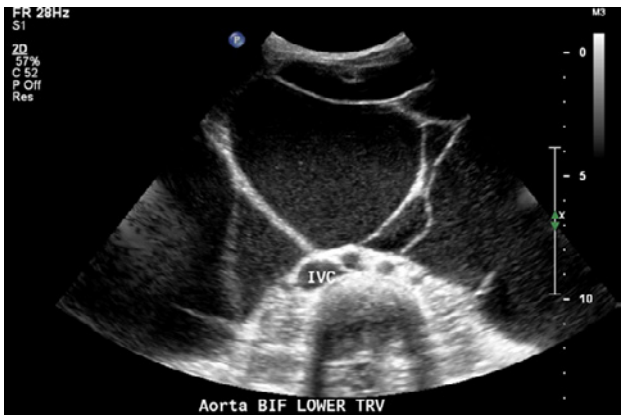

Figure 1: Ultrasound image showing a large cystic lesion in close proximity to vital intra-abdominal vessels.

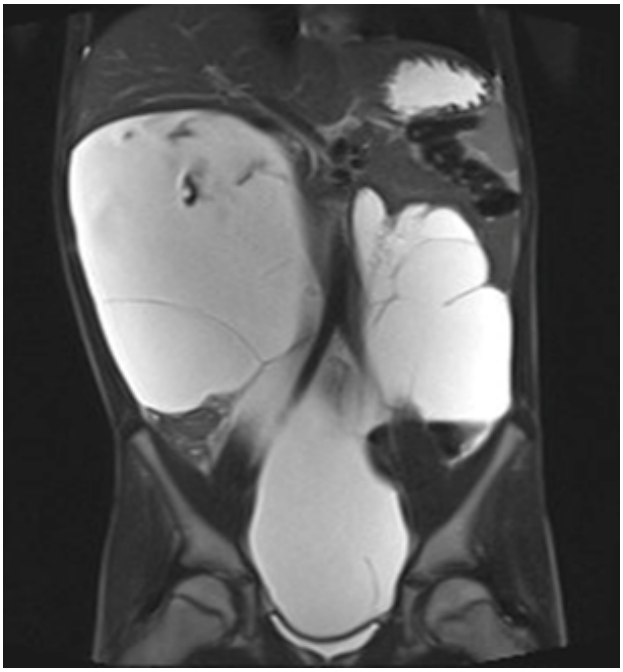

Figure 2: T2 weighted MRI coronal slice showing large multi-septated cystic, gigantic mesenteric lymphatic malformation.

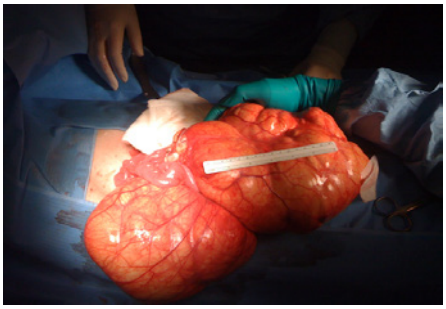

Figure 3: Gigantic mesenteric lymphatic malformation removed in toto still attached to its origin at the third part of the duodenum.

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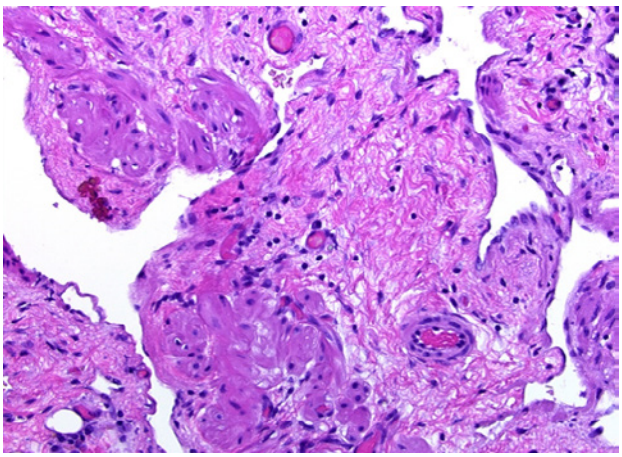

Figure 4: Gigantic mesenteric lymphatic malformation with no evidence of malignancy.

Discussion

Lymphatic malformations are benign, slow growing; vascular malformations that infrequently affect intra-abdominal anatomy. Mesenteric lymphatic malformations are the most commonly found intra-abdominal lymphatic malformations followed by malformations in the omentum, mesocolon and retroperitoneum respectively [2]. They present most commonly in males with a ratio of 3:1. The malformations arise from the disordered embryological development of the lymphatic system. This leads to abnormal or absent communication within the lymphatics. This entity has now been clearly defined in the spectrum of vascular malformations, not as a neoplasm.

Most mesenteric lymphatic malformations present asymptomatically and are found incidentally. Symptomatic patients with mesenteric lymphatic malformations present acutely with complications such as infection, bleeding, bowel obstruction, ascites or secondary to volvulus [3].

Lymphatic malformations have been traditionally classified as simple, cavernous and cystic. However, they are now more commonly classified by their morphogical appearance into macrocystic, microcystic or a combined lesion [4]. Mesenteric lymphatic malformations are thin-walled cystic masses with thin septae dividing the mass into multiple irregular spaces of varying sizes; they have a yellow external surface with large macroscopic interconnecting cysts [5]. The lymphatic spaces are usually filled with proteinaceous eosinophilic fluid. The stroma consists of collagen with lymphatic and lymphoid aggregates. Occasionally, some intra-abdominal lymphatic malformations induce marked inflammatory reactive and inflammatory changes in the surrounding tissues, resulting in the confusing clinical impression of a malignant tumour [6,7].

Diagnosing mesenteric lymphatic malformations can be difficult due to its rare occurrence. Pre-operative ultrasound and CT are suitable tools and will show either a unilocular or multi-locular septae cystic mass [3,8]. However, MRI is more suitable for pre-operative planning as it assesses the lesion extension and origin more proficiently and therefore aids in deciding ones therapeutic approach when excising the lesion [3]. It also differentiates the lesion from a mesenteric cyst as lymphatic malformations lack the specific fat content and fat saturation as seen in MRI demonstration of a dermoid cyst [9].

Total excision of the mesenteric lymphatic malformation is the gold standard treatment however the literature reports varied rates of complete excision with recurrence rates ranging from 0% - 27% [10]. The approach can be trans-abdominal open or a laparoscopic incision however for a gigantic mesenteric lymphatic malformation a formal laparotomy needs to be performed. Total excisional surgery produces risks to vital structures such as intra-abdominal viscera, nerves and arteries. A laparotomy scar can be aesthetically displeasing and result in negative psychological body image issues, especially in the paediatric population [11].

Increasingly, clinicians treating mesenteric lymphatic malformations, which are small in size and found, incidentally, are trending towards monitoring these patients with serial ultrasounds and review as a percentage of these malformations regress spontaneously [12]. The malignant potential if left in situ is 1%; therefore, long-term follow-up is essential in patients with partial excision and recurrence or patients under serial surveillance [13].

In conclusion mesenteric lymphatic malformations are very rare and commonly present incidentally or with secondary complications such as bowel obstruction. Diagnosis can be difficult however MRI is helpful to assess morphology and location. Surgery is the mainstay of treatment, with low recurrences rates; however, serial observation and surveillance can be tailored to certain patients with smaller mesenteric lymphatic malformations.

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