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Ruptured Giant Leiomyoma of the Appendix: A Case Report

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Abstract

We present a case of a 79-year-old female who was admitted with acute abdominal pain and haemoperitoneum. Pathological findings demonstrated a spindled-cell lesion arising from the appendix. The lesion consisted of smooth muscle bundles arranged in fascicles with a whorled appearance. Appendicular leiomyoma is a rare lesion with only a few case reports previously described. We discuss the possible pathological differential diagnosis of such lesions, including GIST, leiomyosarcoma and schwannoma. We present a discussion of the utility of immunohistochemistry in such cases in order to diagnose such tumours.

Keywords: Appendix; Leiomyoma; Tumour; Immunohistochemistry

Introduction

Leiomyomas are rare soft-tissue tumours of the appendix. A review of soft-tissue tumours of the large bowel found a total of 23 appendiceal leiomyomas published in the literature between 1875-1996[1]. A single-centre case series of 101 appendiceal tumours between 1949 and 1972 included only two leiomyomas[2]. The most common presenting symptoms include pain, palpable abdominal mass and haemorrhage. Appendiceal leiomyosarcomas are rarer than leiomyomas and more commonly present with haemorrhage. There was no significant difference between the size of leiomyomas and leiomyosarcomas reported, with the majority of appendiceal lesions measuring less than 5cm maximal length [1]. Case reports demonstrate these lesions can be very large, with previously reported giant leiomyomas weighing up to 500 grams and 15cm maximal length [2-4].

Case Presentation

A 79-year-old female presented to Queen's Medical Centre Emergency Department, Nottingham, UK with severe sudden onset right-sided flank pain. This pain had started suddenly the previous day and worsened overnight with one episode of vomiting. Whilst the patient described that she had generally been feeling well previously, she noted abdominal bloating in the previous two weeks. She was otherwise of minimal co-morbidity and able to live alone independently. She had a past medical history of hypertension, previous ectopic pregnancy, and previous hysterectomy. She did not take any regular medications. Blood tests demonstrated: Hb 109, from baseline of 135 in the previous year; Lactate was 1.6; WCC 7.61; Neutrophils 5.86; CRP 7. The patient was apyrexic, haemodynamically stable and observations were normal.

CT abdomen-pelvis with contrast demonstrated a 95mm rounded mixed density mass within the lower abdomen and pelvis to the right of the midline and contiguous with the appendix. Free fluid was present in the abdomen with predominantly high-density fluid in the pelvis consistent with haemoperitoneum. The radiological differential diagnosis was of a ruptured Gastrointestinal Stromal Tumour or torted fibroid (Figure 1).

The patient underwent laparotomy and appendicectomy the day after admission. Management options of angio-embolization and open surgery were considered. The patient was a Jehovah's Witness and refused to receive exogenous blood products. This, along with the radiological suspicion of active bleeding, resulted in the decision to proceed to emergency surgery to achieve a definitive solution as promptly as possible. Although the patient had agreed to receive cell-saver autologous transfusion, the

surgical team were hesitant to use this owing to the perceived risk of tumour seeding by recycling blood arising from the tumour bed. At laparotomy there was haemoperitoneum and active bleeding from the lesion. The patient had an uneventful recovery and was discharged day 5 after surgery.

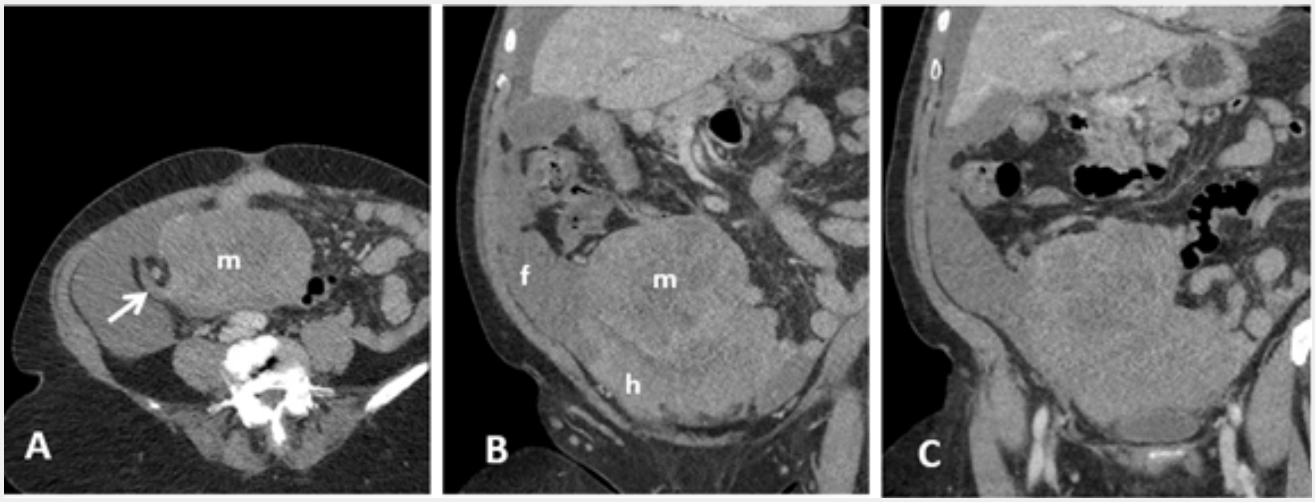

Figure 1: Radiographic images from a CT scan of the abdomen and pelvis with intravenous contrast material. A) Axial section demonstrating a soft tissue mass with heterogeneous attenuation (m) in the pelvis to the right of the midline attached to the appendix (white arrow) B+C) Coronal sections demonstrating proximity to surrounding structures including the uterus. There is free intraperitoneal fluid (f) and evidence of high-density fluid (h) consistent with haemorrhage.

The resected specimen demonstrated a well-circumscribed lesion measuring 110 x 105 x 85mm arising in the distal appendix. Its gross weight was 450 grams. On grossing, the cut surface had a whorled appearance with focal yellow degenerate areas. Histopathological examination demonstrated a spindled-cell lesion arising from the body and distal portion of the appendix. The lesion consisted of smooth muscle bundles arranged in fascicles with a whorled appearance. The stroma of the tumour showed collections of lymphoid aggregate with focal areas of necrosis. However, cytologically the lesion was bland with mild pleomorphism and mitotic activity was very low with 1 mitosis per 50 high-power fields (Figure 2A).

Figure 2: Macroscopic images A) Gross specimen demonstrated a well-circumscribed mass at the tail of the appendix B) There were ectatic vessels close to the subserosa with evidence of haemorrhage C) The cut-surface demonstrated a whorled appearance.

At the periphery of the lesion there are large ectatic vessels lying close to the subserosa. In one of the sections there was evidence of bleeding into the subserosa, probably representing the site of haemorrhage. Immunohistochemical staining revealed: Desmin (+), SMA (+), Caldesmon (+), DOG1 (-), CD34 (-), CD117 (-), S100 (-). Ki67 staining was very low. The above histological and immunohistochemical features confirmed a diagnosis of appendiceal leiomyoma (Figure 3).

Discussion

A PubMed (1950-2020) search was conducted using the following keywords: appendiceal leiomyoma OR appendix +

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leiomyoma OR appendicular leiomyoma OR veriform leiomyoma OR appendix + fibroid OR appendiceal fibroid OR vermiform fibroid. Six English-language case reports were found in this search [3-8]. A review of 50,000 appendicies included 830 leiomyomas, accounting for 1.66% of the tumours identified in the case series, and 632 malignant tumours [9]. The rarity of leiomyomas of the appendix compared to the rest of the large bowel is largely accounted for by the small volume of the appendix, reducing the probability of neoplastic mutation within this structure. Appendectomies, often as a consequence of acute appendicitis, decrease the percentage of the adult population with an intact appendix, thus further decreasing the incidence of neoplasms arising from this structure in the adult population.

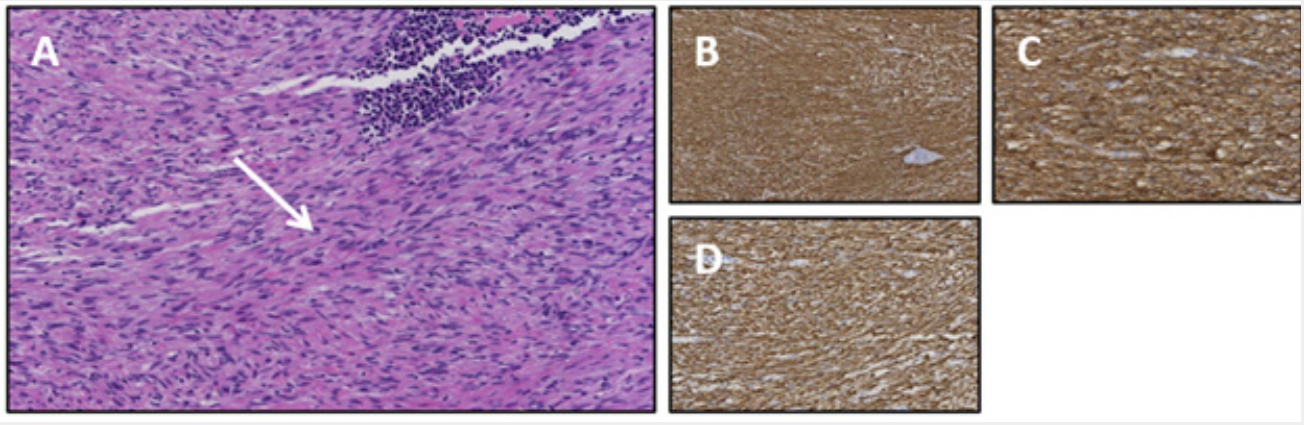

Figure 3: Microscopic images A) Haematoxylin & Eosin sections demonstrated a whorled spindle cell-lesion (white arrow). Immunohistochemistry demonstrated strong positivity for Caldesmon (B), SMA (C) and Desmin (D).

Whilst appendicular leiomyomas presenting with haemoperitoneum have been reported, such a presentation is uncommon and many leiomyomas are often asymptomatic[1]. As previous reports have shown[3,4], these leiomyomas can reach a very large size. In the present case, this caused mechanical compression and dilatation of the overlying subserosal vessels, leading to haemorrhage (Figure 2B).

Table 1: Review of immunohistochemistry in the diagnosis of soft-tissue tumors of the appendix.

	Immunohistochemistry							
Tumour	**CD117**	**DOG1**	**CD34**	**S100**	**SMA**	**Desmin**	**Caldesmon**	**KI67**
Leiomyoma	-	-	-	-	+	+	+	Low
Leiomyosarcoma	-	-	-	-	+	+	+	High
GIST	+	+	+	-	-	-	-	Variable
Schwannoma	-	-	-	+	-	-	-	Variable

To our knowledge, no previous reports discuss the utility of immunohistochemistry in such cases. Indeed, many of the published cases precede the widespread availability of this technique[1]. However, immunohistochemistry is of use in such rare cases to exclude other tumours (Table 1). The main differentials in this case are leiomyosarcoma, gastrointestinal stromal tumour (GIST) and schwannoma. Leiomyosarcoma of the appendix is less common than leiomyoma but carries a far worse prognosis[1]. Criteria to distinguish these tumours include degree of nuclear atypia and mitotic count. As such, ki67 may help illustrate regions of proliferation and help determine the malignant potential of the tumour. Both leiomyosarcoma and leiomyoma are positive for the smooth muscle markers SMA, desmin and caldesmon. Gastrointestinal stromal tumour (GIST) is the most common malignant mesenchymal tumour of the gastrointestinal tract [10]. Cytologically they demonstrate spindle or epithelioid cells arranged in a fascicular pattern. They characteristically express CD117, DOG1 and CD34. S100 and SMA are typically negative. Schwannomas of the appendix are very rare, with only a handful of cases reported in the literature [11]. Microscopic appearances are of interlaced spindle cells and with features of nuclear palisading, hylanised vessels and Verocay bodies. S100 staining is positive on immunohistochemistry with CD34, CD117, DOG1 and desmin typically negative.

In conclusion, we present a case of appendiceal leiomyoma presenting with acute haemorrhage. This case illustrates the need to consider soft tissue tumours of the appendix in the differential

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diagnosis of acute abdomen pain. Furthermore, we demonstrate the importance of immunohistochemistry in subtyping soft-tissue tumours of the appendix, as within this group of tumours there are considerably different prognoses.

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