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ONLINE CASE REPORT

Asymptomatic Ileal Schwannoma presenting as a Mesenteric Tumour

Case report and review of literature

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أورام شوان اللفائفيّة غير المصحوبة بأعراض تعرض كورم مساريقي

تقرير حالة ومراجعة الأدبيات

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الملخص: ورم شوان اللفائفي هو ورم حميد ينشأ من خلايا شوان في الجهاز العصبي المركزي أو الطرفي. المواقع الأكثرشيوعا تشمل الرأس والأطراف. من النادر أن ينشأ هذا الورم من الضفيرة العصبية للقناة الهضمية. ومن الأندر أن يكون موقع المنشأ هو دقاق الأمعاء الدقيقة. نقدم في هذا التقرير حالة مريض لديه كتلة مركزية في البطن كان قد تم تشخيصها قبل الجراحة كورم مساريقي. ومع ذلك، أثبت تشخيص الكِيمْياءُ النسيجية المَناعِيّة للعينة المستأصلة جراحيا بأن الورم هو ورم شوان اللفائفي.

مفتاح الكلمات: خلايا شوان؛ حالات شاذة؛ ورم غمد الليف العصبي؛ كيس مساريقي؛ الأورام اللفائفية؛ البطن تقرير حالة؛ عمان.

ABSTRACT: A schwannoma is a benign tumour which arises from the schwann cells of the central or peripheral nervous system. Common sites include the head and limbs; it is rare that this tumour arises from the gastrointestinal tract's neural plexus. It is even rarer to find the ileum as the site of origin. We report a patient who presented with a central abdominal mass which was preoperatively diagnosed as a mesenteric tumour. However, immunohistochemistry of the surgically-removed specimen proved it to be a benign ileal schwannoma.

Keywords: Schwann Cells, abnormalities; Neurilemmoma; Mesenteric Cyst; Ileal Neoplasms; Abdomen; Case Report; Oman.

A schwannoma is usually a benign, encapsulated neoplasm which arises from the Schwann cells of the peripheral, autonomic or cranial nerves. The most common site for presentation is the 8th cranial nerve.[1] Schwannomas of the gastrointestinal (GI) tract are rare; even if present within the abdomen, the most common site is the stomach.[2] Only a few cases of benign ileal schwannomma have been reported in the literature.[3–14] We report a case of ileal schwannoma where the patient presented with an abdominal mass and discomfort.

Case Report

A 37-year-old male presented with an abdominal mass which had arisen the previous week. There were no obstructive or other associated symptoms. His past medical and surgical history included a laparoscopic hernia repair 7 years previously; his family history was unremarkable. An abdominal examination revealed an 18 x 16 cm non-tender mass in the umbilical region, firm in consistency and dull on percussion, with no visceromegaly or ascites. The external genitalia, per rectal examination and the rest of the systemic examination were normal. A mesenteric or a retroperitoneal tumour was considered clinically as the initial diagnosis.

The patient's haemoglobin level was 14.4 g/dl. He had a normal coagulation profile and a total leukocyte count. His blood sugar level was 4.5 mmol/L, blood urea was 3.1 mmol/L, serum creatinine was 79 µmol/L and his liver function tests were normal (ALT [alanine aminotransferase] 48 IU/L, AST [aspartate aminotransferase] 24 u/L, total bilirubin 4 µmol/L). An ultrasound scan showed a large heterogeneous mass of unidentified origin with internal vascularity. A computed tomography (CT) scan of the abdomen with intravenous and oral contrast showed a mesenteric

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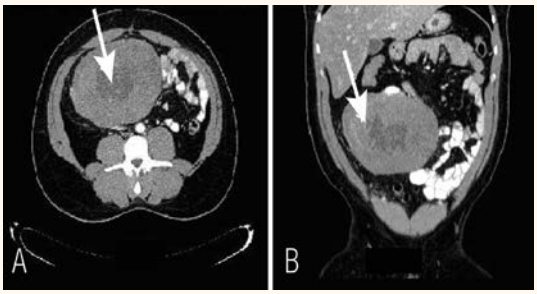

Figure 1 A&B: Computerised tomography scans, from the axial (**A**) and coronal (**B**) view, of the mass in the ileocaecal region without obstruction.

mass of 15 x 14 x 11 cm, with faint enhancement, and central necrosis with no calcification [Figure 1]. The mass was abutting the ascending colon and the hepatic flexure. There was no vascular enhancement or lymph node involvement. An ultrasound-guided true-cut biopsy was performed and showed a low-grade spindle cell lesion suggestive of fibromatosis, myofibromatosis, a solitary fibrous tumour or a benign nerve sheath tumour.

Exploratory laparotomy findings revealed a well-encapsulated 2 Kg tumour in the mesentery of the ileum, with an adherent ileocaecal junction and the appendix stretched over the tumour [Figure 2]. No retroperitoneal infiltration or mesenteric lymph adenopathy was seen. The liver, spleen and the rest of the viscera were normal. A resection of the tumour, along with a limited right hemicolectomy and an end-to-end ileocolic anastomosis, were performed. His recovery was uneventful. Histopathological examination showed a segment of the small bowel with an un-encapsulated neoplasm present within the *muscularis propria* of the ileum, and extending outwards into the serosa. The neoplasm was composed of bland spindle cells with tapered elongated nuclei, and a scattering of plumper nuclei with pale chromatin and small distinct nucleoli [Figure 3]. Immunohistochemical markers showed a diffuse positivity of S-100, with variable intensity, ranging from moderate to focally strong, highlighting the areas of neural differentiation. The cells were also strongly positive for neurospecific enolase (NSE). The diffuse positivity of S-100 favoured the diagnosis of schwannoma. The recovery of the patient was uneventful, and he was discharged on the 7th postoperative day. The one year follow-up showed no recurrence.

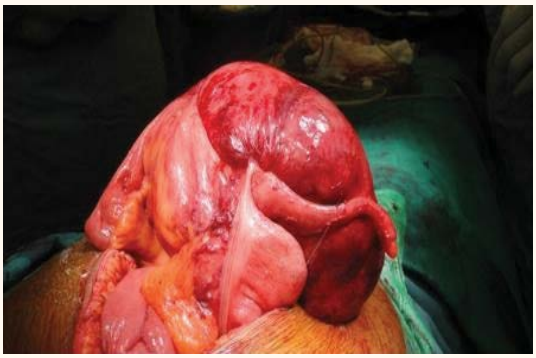

Figure 2: The mass in the ileocaecal region.

Discussion

Ileal schwannomas usually present with abdominal pain,[11] but may also present with other complications; melena,[9,11] degeneration,[4,6] and intussusception[5,10,13] having also been reported [Table 1]. This case presented with an abdominal mass without obstructive symptoms. A review of the limited available literature showed that it was the first case to present asymptomatically. A preoperative diagnosis is quite difficult.[11] An ultrasound (US) may show a mass of variable echogenicity. There is no consensus on the CT findings of ileal schwannomas, as, in reported cases, CT was non-diagnostic and showed only a mass.[11] Magnetic resonance imaging (MRI), as reported by Nagi,[11] may show a submucosal tumour; however, the findings were again nonspecific. In cases

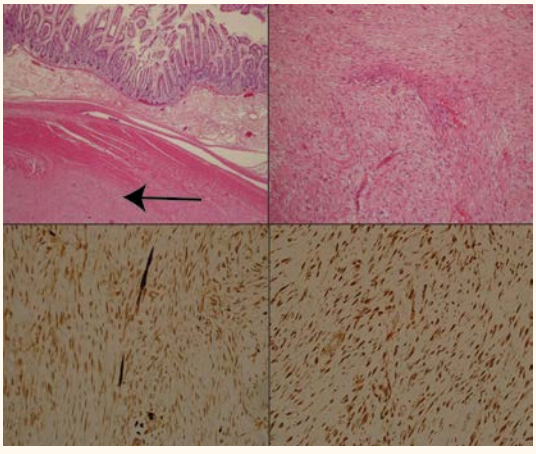

Figure 3: The top images show an unencapsulated spindle cell neoplasm involving the *muscularis propria* of the bowel wall (arrow). Under high magnification, it is formed of fascicles of bland spindle cells. The tumour cells are positive for S-100 (lower left image) with darker staining nerve bundles highlighted. Neurospecific enolase is also positive (lower right image).

Table 1: A review of the literature on schwannomas

Name of Study	Year of publication	Origin of study	No. of cases reported/reviewed	Presentation
Cornette De St-Cyr M, *et al.***	1958	France	1	-
Poulat R, *et al.***	1963	France	1	Degeneration Intraperitoneal bleeding
Maison E.**	1969	France	1	Ileo-ileal invagination
Filimon C, *et al.***	1974	Romania	1	Degeneration
Ilieve H.**	1977	Croatia	1	-
Gourtsoyiannis NC, *et al.**	1993	Greece	1	-
Jadhav RN, *et al.**	1996	Mumbai (India)	1	Bleeding per rectum
Insegno W, *et al.*	1996	Genova	1	Intussception in pregnancy
Nagal T, *et al.*	2003	Beppu (Japan)	5	Adbominal Pain = 3 Melena = 2
Rangiah DS, *et al.**	2004	Australia	1	-
Hirasaki S, *et al.*	2008	Japan	1	Ileocolic intussception
Târcoveanu E, *et al.**	2011	Romania	1	-

** = abstract only; ** = no abstract.*

presenting with melena blood, a scintigraphy[11] or angiography may be of help. In these cases, upper or lower GI endoscopies may rule out other causes of GI haemorrhage. In the present case, an US-guided biopsy was supposed to assist in making a decision; however, the results were inconclusive. Nevertheless, an US-guided biopsy may rule out lymphomas, in which case surgery could be avoided. Spilling, a rupture of the mass, or seeding of the malignant tumour are a few of the theoretical risks associated with a US-guided biopsy of any intra-abdominal mass. The preoperative differentiation between a benign or malignant schwannoma by a flourodeoxyglucose positron emission tomography (FDG-PET) scan[15] has been proposed by some authors, but its application in diagnosing an ileal schwannoma has not yet been established. The gold standard is a histopathological examination of the surgically-removed specimen. With electron microscopy, spindle-shaped cells are characteristic, with areas of hyper- and pauci-cellularity). Immunohistochemistry confirms a schwannoma if the staining for S-100 protein is positive.

The mainstay of treatment is a surgical resection and an end-to-end anastomosis. The extent of the resection will depend upon the location of the tumour. If, after the ileal resection, a sufficient length of the dista ileum is available for ileo-ileal anastomosis, continuity should be restored by anastomosing the two ends. In our case, location was so distal in the ileum that we had to proceed with a limited right hemicolectomy. Histopathological analysis has a major role in the further management of such cases. In cases of benign ileal schwannomas, no recurrence has yet been reported;[11] however, lifelong surveillance will be required in cases of malignant ileal schwannomas.

Conclusion

The ileal schwannoma is a rare entity with variable presentation, and arriving at a preoperative diagnosis is usually challenging. In cases presenting without complication, a resection and an anastomosis of the ileal is the treatment of choice.

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