Educational Case Report

G. Niraj* and Cathy J. Richards

Leiomyosarcoma of the small intestine presenting as abdominal myofascial pain syndrome (AMPS): case report

https://doi.org/10.1515/sjpain-2020-0099
Received June 21, 2020; accepted July 9, 2020; published online September 25, 2020

Abstract

Objective: To highlight an extremely unusual presentation of an aggressive, rare small bowel malignancy presenting as abdominal myofascial pain syndrome.
Case presentation: The report is presented from a tertiary pain medicine unit at a university teaching hospital. A female patient presenting with chronic abdominal pain was initially diagnosed as abdominal myofascial pain syndrome. The report details the possible facilitation of the diagnosis of a rare, highly aggressive small bowel tumour by interventional treatment for abdominal myofascial pain syndrome.
Conclusion: This case highlights a rare and aggressive malignancy of the small intestine presenting clinically as abdominal myofascial pain syndrome.

Keywords: abdominal myofascial pain syndrome; chronic abdominal pain; leiomyosarcoma; trigger point injection.

Introduction

Leiomyosarcoma (LMS) of the gastrointestinal tract is a rare and aggressive soft tissue tumour. Patients present with non-specific symptoms including abdominal pain that are often not taken seriously [1]. As a result, at the time of diagnosis, visceral metastasis is common and the prognosis is poor [2].

Abdominal myofascial pain syndrome (AMPS) is a common cause of chronic abdominal pain and is often misdiagnosed as visceral abdominal pain syndrome or functional abdominal pain syndrome [3]. The author presents a unique case of undiagnosed LMS of the small intestine presenting with definitive features of AMPS.

The patient provided written consent for the use of de-identified data for analysis and publication in a peer-reviewed journal.

Case report

A 45 year old female was referred by a gastroenterologist with five-month history of left sided upper abdominal pain predominantly on the left side. An upper gastrointestinal (UGI) endoscopy and a computer tomographic (CT) scan of the abdomen did not reveal a visceral cause. The referral stated possible abdominal wall as the pain generator. The onset of pain was gradual and the patient appeared in distress as the pain had a significant effect on her mood, sleep and quality of life. The pain was described as a constant dull ache with intermittent sharp, stabbing flare-ups. Flare-ups were triggered on movement and after eating. There was increased sensitivity to touch over the left upper abdomen. Bowel function was unaffected. There was no history of any trauma. Comorbidities included a past history of gastritis and iron deficiency anaemia.

On examination, the abdomen was soft and there was no organomegaly. There was allodynia to touch over the left upper quadrant. There were trigger points in the upper rectus abdominis muscle (RAM), predominantly on the left side. Tenderness on examination increased when the abdominal wall was tensed (positive Carnett's sign) [4]. The initial diagnosis was abdominal myofascial pain syndrome (AMPS) secondary to gastritis (viscerosomatic convergence).

The patient underwent ultrasound guided abdominal trigger point injection with a mixture of 0.25% levobupivacaine and depot methylprednisolone. Needle sign identified 12 trigger points in the upper RAM (eight on the left and four on the right side) in zones 1–3 [3]. Trigger points are usually observed in three areas (zones 1–3) in the RAM

***Corresponding author: Dr. G. Niraj,** Consultant in Anaesthesia & Pain Medicine, Clinical Research Unit in Pain Medicine, University Hospitals of Leicester NHS Trust, Gwendolen Road, Leicester, UK, Fax: +44 1162584661, E-mail: Niraj.g@nihr.ac.uk
Cathy J. Richards: University Hospitals of Leicester NHS Trust, Gwendolen Road, Leicester, UK

on ultrasound guided needling. Zone 1 corresponds to medial end of RAM; zone 2 is the middle third while zone 3 is located in the lateral end of RAM [3]. Pathognomonic myofascial twitch was observed in seven trigger points on the left upper rectus [3]. The diagnosis of AMPS was confirmed on observing myofascial twitch response in multiple trigger points.

The patient reported significant post-procedural flare-up for 10 days requiring oral morphine (40–60 mg/day). Although opioids are reported to be ineffective in somatic musculoskeletal pain, a short course of oral morphine was able to reduce the associated distress, anxiety and the intensity of post-procedural pain in the patient.

Two weeks after the intervention, the patient developed non-stop vomiting, diarrhoea and increased pain in the upper abdomen. The patient required emergency admission after five days from the onset of vomiting and diarrhoea. A CT of the abdomen revealed high-grade small bowel obstruction. She underwent an emergency laparotomy. Surgery revealed a polypoidal tumour in the small bowel causing ileal intussusception. A small bowel resection and primary anastomosis was performed. The patient had an uneventful recovery and was discharged on day 5. Histology of the small bowel tumour revealed a malignant polypoidal spindle cell tumour with high mitotic activity (34 mitoses per 10 high power fields) showing strong positive staining for smooth muscle actin (SMA) with focal desmin, strong oestrogen receptor (ER) and focal progesterone receptor (PR). The features were those of a high-grade (Trojani grade 3) leiomyosarcoma, completely excised.

Discussion

The novelty of the report is the unique presentation. Leiomyosarcoma of the small intestine is a rare neoplasm to the extent that the WHO is unable to provide meaningful data on demographic or clinical features [5, 6]. Leiomyosarcoma is an aggressive, slow growing malignancy and is often diagnosed at a late stage when curative treatment is futile [7]. The tumour is not clearly visualised on CT scan of the abdomen. Delayed diagnosis is common and is usually as a result of non-specific signs of vomiting and abdominal pain [1].

Chronic abdominal pain (CAP) can be broadly classified into three types based on aetiology [8]. They include chronic visceral abdominal pain syndrome (VAPS), Chronic abdominal wall pain (CAWP) and functional abdominal pain syndrome (FAPS). CAWP can be subdivided into abdominal myofascial pain syndrome (AMPS) and anterior cutaneous nerve entrapment syndrome (ACNES). AMPS is the commonest type of CAWP and is primarily caused by underlying visceral inflammation (viscero-somatic convergence) and less commonly, by trauma [3]. AMPS is diagnosed by clinical presentation, absence of on-going visceral inflammation and Carnett's sign on examination. However, the pathognomonic sign of AMPS is the elicitation of myofascial twitch when the needle enters the affected muscle. This can be visualised on ultrasound scanning [3].

The patient was initially seen by a gastroenterologist with a special interest in visceral abdominal pain. Thereafter, the patient was reviewed by a pain medicine specialist with an extensive interest in AMPS [3, 9]. The patient displayed symptoms and signs highly suggestive of AMPS in the absence of visceral inflammation (negative UGI endoscopy, CT scan of the abdomen). Interventional treatment for AMPS appeared to confirm the diagnosis when the pathognomonic sign of myofascial twitch was visualised in seven trigger points. The author was wholly convinced of the diagnosis and this was conveyed to the patient.

The eventual diagnosis of a highly malignant but resectable tumour may have been fortuitous. However, there may have been another possibility. Opioid use in chronic abdominal pain is common and is often a major concern for physicians managing these patients [9]. In this case, the high dose opioid consumption may have played a role in facilitating the diagnosis. A combination of rectus abdominis spasms (from the flare-up) and opioid use by an opioid naïve individual may have triggered the intussusception leading to small bowel obstruction (opioid induced gastrointestinal dysfunction, OIGID) requiring emergency surgery and the subsequent diagnosis of an underlying high grade, rare malignancy [10].

Leiomyosarcoma can occur in the small bowel, colon and rectum, often as an obstructing polypoid intraluminal mass. Histological and immunohistochemical features are similar to those of leiomyosarcoma at other sites. ER and PR positive tumour can be seen in an extra-uterine leiomyosarcoma and therefore, it is essential to exclude metastasis from a gynaecological primary.

Presence of tender trigger points in the rectus abdominis muscle and myofascial twitch on ultrasound guided needling suggest the possibility of viscero-somatic convergence initiated by the tumour [3].

In conclusion, this case highlights a rare and aggressive malignancy of the small intestine presenting clinically as abdominal myofascial pain syndrome.

Research funding: None declared.

Author contributions: All authors have accepted responsibility for the entire content of this manuscript and approved its submission.

Conflict of interest: The authors' state no conflict of interest.

Informed Consent: The authors' state that written informed consent was obtained from the patient.

References

1. Paski SC, Semrad C. Small bowel tumours. Gastrointest Endoscopy Clin N Am 2009;19:461–79.
2. Chunwei X, Hongyan H, Jingjing W, Zhang B, Shao Y, Zhang L, et al. Diagnosis value of CD117 and PDGFRA, alone or in combination DOG1, as biomarkers for gastrointestinal stromal tumors. Ann Transl Med 2015;3:308.
3. Niraj G. Pathophysiology and management of abdominal myofascial pain syndrome (AMPS): a three-year prospective audit of a management pathway in 120 patients. Pain Med 2018;19: 2256–66.
4. Thomson H, Francis DM. Abdominal wall tenderness: a useful sign in the acute abdomen. Lancet 1977;2:1053–4.
5. Yamamoto H, Handa M, Tobo T, Setsu N, Fujita K, Oshiro Y. Clinicopathological features of primary leiomyosarcoma of the gastrointestinal tract following recognition of gastrointestinal stromal tumours. Histopathology 2013;63:194–207.
6. Aggarwal G, Sharma S, Zheng M, Reid MD, Crosby JH, Chamberlain SM, et al. Primary leiomyosarcomas of the gastrointestinal tract in the post-gastrointestinal stromal tumor era. Ann Diagn Pathol 2012;16:532–40.
7. Guzel T, Mech K, Mazurkiewicz M, Dąbrowski B, Lech G, Chaber A, et al. A very rare case of a small bowel leiomyosarcoma leading to ileocaecal intussusception treated with a laparoscopic resection: a case report and a literature review. World J Surg Oncol 2016;14:48.
8. Niraj G, Chaudhri S. Prospective audit of a pathway for in-patient pain management of chronic abdominal pain: a novel and cost-effective strategy. Pain Med 2018;19:589–97.
9. Alva S, Niraj G. Opioid reduction and long-term outcomes in abdominal myofascial pain syndrome (AMPS): a 6-year longitudinal prospective audit of 207 patients. Pain Physician 2020 in press;2020. .
10. Drewes AM, Munkholm P, Simren M, Breivik H, Kongsgaard UE, Hatlebakk JG, et al. Definition, diagnosis and treatment strategies for opioid-induced bowel dysfunction-recommendations of the Nordic working group. Scand J Pain 2016;11:111–2.