CASE REPORT

Mantle cell lymphoma: a rare cause of a solitary duodenal mass

Ghita Moussaide,¹ Ali Kazemi,² Ricardo Mitre,² Marcia C Mitre³

¹Department of Internal Medicine Residency Program, Allegheny Health Network, Pittsburgh, Pennsylvania, USA

²Department of Gastroenterology, Allegheny Health Network, Pittsburgh, Pennsylvania, USA

³West Penn Allegheny Health System, Pittsburgh, Pennsylvania, USA

Correspondence to

Dr Marcia C Mitre, mmitre@WPAHS.ORG

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SUMMARY

Mantle cell lymphoma is a very aggressive lymphoma with a very poor prognosis. It commonly involves the gastrointestinal tract but rarely presents as primary gastrointestinal lymphoma. The most notable cases of primary gastrointestinal mantle cell lymphomas have been described as multiple lymphomatous polyposis and have a very poor prognosis. We report a case of primary gastrointestinal mantle cell lymphoma that was discovered by endoscopic biopsy of a single duodenal polyp in a 70-year-old woman who was previously treated for *Helicobacter pylori* gastritis. She presented with a 6-month history of indigestion, heartburn and abdominal bloating. A subsequent workup revealed one extranodal site of involvement, lymphatic involvement below the diaphragm and a normal bone marrow biopsy. We followed a wait-and-watch approach including serial CT scans and blood tests. Two years later, her symptoms have not progressed and her disease has remained stable.

Figure 1 Solitary duodenal bulb mass.

BACKGROUND

Primary gastrointestinal mantle cell lymphoma is rare and very aggressive with a poor survival rate even with potent chemotherapy. Our patient presented with mild gastrointestinal symptoms and her lymphoma only consisted of a single polyp with one extranodal involvement. Her disease did not take the expected aggressive course, and it remained stable 2 years after diagnosis even without chemotherapy. This has enabled her to maintain a quality of life that may not have been possible with the side effects of chemotherapy.

CASE PRESENTATION

A 70-year-old woman with a history of Prinzmetal's angina, hypertension, hyperlipidaemia and previously treated *Helicobacter pylori* gastritis presented with a 6-month history of indigestion, heartburn and abdominal bloating. Three of her brothers had colon cancer, her mother had breast cancer and her father had prostate cancer.

INVESTIGATIONS

A physical examination revealed a well-nourished, average-built woman in no acute distress. Her vital signs were stable. She had no palpable cervical, axillary, supraclavicular or inguinal lymphadenopathy. Her abdomen was soft and non-tender. Routine laboratory tests including a complete blood count, basic metabolic panel and lipase levels were within normal limits.

On endoscopic evaluation, the oesophageal mucosa and stomach mucosa appeared normal. A 1.2-cm duodenal bulb polyp was seen (figure 1). Gastric biopsies showed chronic gastritis and were positive for *H pylori*. Histological evaluation of the biopsy taken from the duodenal polyp revealed a diffuse atypical lymphoid infiltrate of the duodenal mucosa. The lymphoma cells were consistent with mantle cell lymphoma and were positive for CD20, CD5, BCL-2 and Cyclin D and were negative for CD3, CD10, BCL-6 and CD23 (figures 2 and 3). MIB stain showed a proliferative activity of 15–20%. A colonoscopy revealed normal mucosa.

For relief of her symptomatic non-ulcer dyspepsia, *H pylori* gastritis was treated with amoxicillin, clarithromycin and lansoprazole. Oncological evaluation showed an Eastern Cooperative Oncology Group (ECOG) performance status of 1. She also underwent a CT/positron emission tomography (CT/PET) for staging which showed an enlarged left inguinal lymph node and a non-enlarged left external-iliac lymph node (figure 4). There was no sign of bone or central nervous system involvement, and her bone marrow biopsy was also normal.

DIFFERENTIAL DIAGNOSIS

A formal diagnosis of mantle cell non-Hodgkin's lymphoma was made with one extranodal site of involvement and lymphatic involvement below the diaphragm. Her disease was categorised into an intermediate group for aggressiveness.

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Figure 2 Immunohistochemistry for Cyclin D1, showing strong nuclear staining, as a consequence of the t(11;14)(q13;32).

TREATMENT

Two oncologists reviewed her case and felt that due to the indolent nature and low-to-intermediate risk prognostic index of her disease, it was best to hold off aggressive chemotherapy. Instead they recommended a wait-and-watch approach with close monitoring of her symptoms, blood tests for anaemia and lactate dehydrogenase (LDH) levels monthly, and surveillance CT/PETs every 3 months.

OUTCOME AND FOLLOW-UP

Her symptoms of dyspepsia resolved after completing treatment for *H pylori*. Repeat endoscopy and endoscopic ultrasound showed no significant change in the polyp size. Over the past 2 years, her disease has remained stable.

DISCUSSION

Mantle cell lymphoma is known to be an aggressive, non-curable type of non-Hodgkin's lymphoma with short remission periods and frequent relapses. It represents 4% of all lymphomas in the USA. Patients are typically men in their sixth or seventh decade of life who present with stage III/IV disease

Figure 3 H&E x40. Monomorphous small lymphocytes with irregular nuclei, condensed chromatin, inconspicuous nucleoli and spare cytoplasm. Intermixed singly distributed macrophages are also present.

Figure 4 CT/positron emission tomography showing avid fludeoxyglucose uptake along the duodenum.

including involvement of the lymph nodes, spleen and bone marrow. Extranodal involvement also includes the skin, lacrimal glands and gastrointestinal tract. Mantle cell lymphoma is associated with a t(11;14) translocation which leads to over-expression of Cyclin D1. Cyclin D1 is present in 98% of cases and is often used for detection of mantle cell lymphoma.¹

Risk stratification is based on the mantle cell lymphoma international prognostics index (MIPI) which incorporates age, ECOG score, LDH and white cell count (WCC). The ECOG score depends on the patient's symptoms and performance status. According to the American Journal of Hematology's 2012 Annual Clinical Updates in Hematological Malignancies, the median overall survival (OS) for low-risk groups was not reached, the OS for the intermediate-risk group was 51 months and that of the high-risk group was 29 months.¹

While approximately 84% mantle cell lymphoma cases have some degree of microscopic gastrointestinal involvement regardless of the presence of gastrointestinal symptoms,² only about 4–9% of all primary gastrointestinal B cell lymphomas are caused by primary mantle cell lymphoma.³ To date, multiple lymphomatous polyposis is usually seen with gastrointestinal mantle cell lymphoma.^{4–6} Mantle cell lymphoma presenting as multiple lymphomatous polyposis is incurable and has a mean survival rate of 3 years even with therapy.⁶

The definition of primary gastrointestinal lymphoma is inconsistent in the literature. Some studies use Dawson's criteria to classify gastric lymphomas as primary when the stomach is predominantly involved, and if present, intra-abdominal lymphadenopathy corresponds to the expected lymphatic drainage of the stomach.⁷ Other reports define primary gastrointestinal involvement when the initial symptoms of the disease are located in the abdomen or present as a disturbance of the gastrointestinal function.⁸ Other studies consider non-Hodgkin's lymphoma to be of primary gastrointestinal involvement if it presents with: (1) no palpable superficial lymphadenopathy, (2) no enlarged mediastinal lymph-nodes on chest radiographs, (3) normal WCC count and differential, (4) predominating bowel lesion at laparotomy, where the only lymph nodes involved are in the immediate neighbourhood of the mass and (5) tumour-free liver and spleen.⁹

Our patient's lymphoma was categorised as primary gastrointestinal, as it presented with abdominal symptoms and involvement of only two lymph nodes below the diaphragm

with no involvement of any other organs. She had no palpable lymph nodes and her blood work was within normal limits.

Instead of a common presentation of multiple lymphomatous polyposis, our patient's tumour consisted of only one polyp, and her symptoms were mild and never included B symptoms such as fever, night sweats or weight loss. Furthermore, while multiple lymphomatous polyposis behaves in an aggressive manner, mantle cell lymphoma presenting as a single duodenal lesion has been more indolent. More than a year after her initial diagnosis, our patient's disease has remained stable.

Therapy for mantle cell lymphoma is based on the individual patient's risk. A simple wait-and-watch approach in the begin-

ning is considered appropriate therapy for elderly patients or patients with a low MIPI. Chemotherapy can be started once patients develop symptoms. Initial aggressive chemotherapy +/- bone marrow transplant is recommended in young symptomatic patients. The median duration of remission is 1.5–3 years.¹

Owing to the indolent nature of our patient's lymphoma, it was decided to follow the wait-and-watch approach. She continues to be monitored for the development of symptoms or changes in her laboratory tests or imaging studies. A wait-and-watch approach in her case was a safe option with preservation of a good quality of life and avoidance of side effects from early initiation of cytotoxic and biological therapy.

Contributors MCM and RM were involved in the care of the patient and review of the manuscript. GM and AK wrote the manuscript.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Learning points

- ▶ Mantle cell lymphoma (MCL) is a type of non-Hodgkin's lymphoma that commonly involves the gastrointestinal tract in advanced disease stage with regional lymph node involvement.
- ▶ Primary gastrointestinal MCL is rare. Multiple lymphomatous polyposis was one of the first reported primary gastrointestinal MCLs. It was described as the gastrointestinal counterpart of MCL and is just as aggressive.
- ▶ Primary gastrointestinal MCLs have since been reported to present as a bleeding ulcer or ulcerating mass, all requiring chemotherapy.
- ▶ We found that primary gastrointestinal MCL can also present as a single polyp and can have a very indolent course, not requiring the usual potent chemotherapy reserved for MCL.
- ▶ Although MCL is known as being a highly aggressive disease, some presentations, such as our patient's, may be appropriate for the adoption of wait-and-watch approach which allows the preservation of a good quality of life and avoidance of side effects from early initiation of cytotoxic and biological therapy.

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