CASE REPORT

Paraduodenal inflammatory pseudotumor masquerading as malignancy

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SUMMARY

A 30-year-old woman presented with severe abdominal pain and abdominal distension. CT demonstrated two intra-abdominal masses, one involving the left lateral segment of the liver and the other adjacent to the duodenum. Initial biopsies were consistent with focal nodular hyperplasia of the liver and non-specific lymphocytic infiltrate in the paraduodenal mass. Due to persistent symptoms, the patient underwent laparoscopic resection of the paraduodenal mass. Final pathology was consistent with an inflammatory pseudotumour and the patient's symptoms subsequently resolved.

BACKGROUND

Inflammatory pseudotumours (IPTs) are rare entities that originate most commonly in the lungs but can also present in intra-abdominal organs and mimic solid tumours. They can appear hypervascular on contrast-enhanced imaging and can be mistaken for neuroendocrine tumours (NETs), hepatic haemangiomas, carcinomas, gastrointestinal stromal tumours (GISTs) and other neoplastic lesions. Due to the inability to differentiate pseudotumours from neoplasms by imaging and biopsy, and presence of symptoms at diagnosis, resection is often indicated. For otherwise healthy patients, a laparoscopic approach is appropriate and can minimise morbidity and expedite recovery.

CASE PRESENTATION

The patient was a 30-year-old woman with no medical or surgical history who initially presented with severe lower abdominal pain, distension and constipation, requiring a visit to the emergency department. She denied nausea, vomiting, fever, weight loss, haematochezia or any antecedent trauma. CT demonstrated a 5.3×9.5 cm mass of the liver in segments II and III and a 5.0×3.8 cm paraduodenal mass (figure 1A-C).

The patient was treated for constipation and irritable bowel syndrome with laxatives, analgesics, probiotics and lubiprostone, which improved symptoms without complete resolution. She had no significant medical or surgical history and her only medication was an oral contraceptive, which was discontinued at the onset of symptoms. She denied smoking, alcohol use and family history of malignancy.

INVESTIGATIONS

The patient was initially evaluated and underwent workup at an outside institution. She underwent

upper endoscopy, which did not reveal an intraluminal lesion. She underwent a percutaneous biopsy of the liver mass, which was consistent with focal nodular hyperplasia (FNH). She underwent percutaneous biopsy of the paraduodenal mass, which revealed an atypical lymphocytic infiltrate. She was recommended to undergo resection of both the liver and paraduodenal masses at the outside institution.

The patient presented to our care for a second opinion regarding the need for surgical resection. Physical examination revealed no abdominal tenderness or palpable mass. She underwent routine bloodwork including a complete metabolic panel, complete blood count and coagulation panel, which revealed no abnormalities. Testing for IgG subclasses were normal including an IgG₄ level of 14.0 mg/dL (normal: 4–86 mg/dL). Internal pathological review of prior biopsies was performed. The liver biopsy was consistent with non-specific lobular inflammation. The paraduodenal lesion demonstrated fibrous tissue with dense lymphocytic infiltrate and increased IgG₄.

She underwent an endoscopic ultrasound which demonstrated mild extrinsic compression of the duodenal bulb by a homogeneous, hypoechoic, and well-defined 4.3×4.0 cm mass. A fine-needle aspiration was performed and showed benign intestinal epithelium.

DIFFERENTIAL DIAGNOSIS

The initial diagnosis for the patient's liver lesion was FNH based on imaging and biopsy. The paraduodenal lesion was thought to be a neuroendocrine neoplasm based on well-demarcated nature and degree of contrast enhancement on CT. The patient continued to have gastrointestinal symptoms, and she was recommended to undergo laparoscopic resection of the paraduodenal mass with possible open exploration if required from a technical or oncological standpoint. The liver mass would be visualised at the time of surgery, and a biopsy was planned only if the appearance was malignant or discordant with the percutaneous biopsy result.

TREATMENT

The abdomen was entered using direct cutdown technique at the infra-umbilical position and a 10 mm blunt-tipped trocar was placed. Two 5-mm trocars were introduced in the right upper and mid-abdomen for use by the surgeon. An additional 5-mm trocar was placed in the left abdomen for the assistant. The mass was fleshy and vascular, and

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Figure 1 Preoperative CT images demonstrating enhancing lesions of (A) the left lateral segment of the liver, (B) paraduodenal tumour and (C) both tumours in coronal section.

appeared to be anterior to the duodenum and arose from the gastrocolic omentum laterally (figure 2A,B). The liver lesion was visualised (figure 2C) and no biopsy or resection was attempted due to the benign appearance. The paraduodenal tumour was elevated off the underlying duodenum and pancreas. Attachments to the adjacent structures were divided circumferentially using a combination of blunt dissection and the Maryland jaw Ligasure device (Covidien, Minneapolis, MN, USA). Dissection was performed close to the mass and the resection bed is shown in figure 2D. The specimen was extracted via the infraumbilical incision using an Endo Catch retrieval bag (Covidien).

The tumour was sent for frozen section examination, which demonstrated an extensive small, round, blue cell tumour, which raised the possibility of a well-differentiated NET or lymphoproliferative process.

The patient recovered uneventfully postoperatively, her diet was advanced, and she was discharged on postoperative day 2.

OUTCOME AND FOLLOW-UP

Final pathology was consistent with an IPT. On gross examination, the lesion was firm and somewhat fibrotic in appearance. Figure 3 demonstrates a representative histological section and immunohistochemical (IHC) stains. Histologically, the lesion

Figure 2 Intraoperative images of (A) and (B) paraduodenal tumour, (C) liver lesion and (D) surgical site after resection.

Figure 3 H&E stain and immunohistochemical stains of the lesion. The lesion consists of a lymphoplasmacytic infiltrate with areas surrounded by fibrosis. There are prominent nodules of CD20-positive B cells with admixed CD3 and CD43 T cells. Definitive germinal centres were absent as highlighted by lack of CD10 and BCL6 and lack of CD21 follicular dendritic cell network. The BCL2 highlights the B and T cells but no aberrant expression of LEF1 or Cyclin D1 was seen. The lesion had a low proliferation rate as highlighted by the Ki67. There was focal increase in IgG₄ expressing lymphocytes.

was composed mainly of inflammatory infiltrate in a fibrotic background. The inflammatory infiltrate was composed of CD20 B cells admixed with CD3-positive T cells. The B cells were arranged in nodules and were weakly BCL2 positive. The B-cell networks had a follicular dendritic cell network as demonstrated by CD21, but germinal markers BCL-6, CD10 and LMO2 were negative. There was no aberrant coexpression of LEF1, Cyclin D1 or CD43 in the B cells. CD138 staining highlighted plasma cells, which were seen surrounding the nodules of B cells. Kappa and lambda in situ hybridization, as well as IHC stains demonstrated a polyclonal population of plasma cells. Additionally, IHC stains for CD30, CD15, and pan keratin AE1 and AE3 were negative. Ki-67 highlighted primarily cells within the T-cell areas and labelled less than 5% of those cells. Congo red stain was negative for amyloid deposition. An IHC stain for IgG₄ highlighted a pocket of plasma cells with IgG₄ expression up to 53 IgG₄ expressing plasma cells per high-power field. B-cell clonality studies failed to demonstrate a clonal population. No neuroendocrine differentiation was seen in the final sections.

Overall, the lesion demonstrated a dense lymphocytic infiltrate without evidence of clonality or atypical antigen expression by B cells or T cells. There was an increase in IgG₄ but serum IgG₄ levels were not elevated. The lesion was therefore best classified as IPT.

The patient experienced complete resolution of symptoms and remained symptom free after 18 months of surgery.

DISCUSSION

IPTs have been reported most commonly in the lungs and orbit but can appear in other locations, including the liver, spleen, mesentery, omentum, pancreas, stomach and small bowel.^{1 2} These lesions can behave aggressively with local invasion and can recur after resection, mimicking true malignancies.¹ They have been reported by a variety of names, including inflammatory myofibroblastic tumour, histiocytoma, xanthoma and plasma cell granuloma.²⁻⁴

The clinical presentation can be variable and can include a systemic inflammatory syndrome with fever, weight loss, malaise, non-specific symptoms, including abdominal pain and

anaemia, and symptoms due to mass effect or ulceration. Workup includes cross-sectional imaging with intravenous contrast. CT of IPTs can demonstrate highly variable appearance with hypo-, iso- or hyper-attenuating lesions, and contrast administration can demonstrate peripheral, homogeneous or heterogeneous enhancement.^{1 5} MRI can be used to further characterise these lesions, which are often hypointense on T1-weighted images and hyperintense on T2-weighted images.¹ The age range of presentation is also variable, but these lesions appear most commonly in children and young adults, with a female predominance in one series of extrapulmonary IPTs.⁴

The radiographic appearance and biological behaviour of IPTs can mimic that of neoplastic lesions. The pathogenesis of IPTs and whether they represent true neoplasm is under debate. It has been suggested that these lesions develop in response to trauma, surgery, malignancy or infection^{1 5} although our patient did not have these risk factors. Local recurrence is common and is reported in the literature in 18%–40% of cases,^{2 4} related to incomplete resection or rapid proliferation of grossly occult residual disease. Metastatic recurrence is rare but has been reported.⁵ Long-term outcomes for patients with IPT appear favourable. For patients with at least 3 months of follow-up in one series,⁴ 33 of 49 patients who underwent resection had no recurrence, 11 had recurrence and 7 were disease free

after re-resection. The likelihood for recurrence was noted to be related to completeness of resection. Another four patients who had biopsy without resection were all alive at follow-up with spontaneous regression in three patients. A small number of lesions have been reported to undergo sarcomatous degeneration.^{4 5} Due to the locally aggressive behaviour, surveillance after resection is recommended.^{2 5}

Surgical resection is the mainstay of therapy, even in recurrent disease, and chemotherapy and radiation have no defined role in management.^{4 6} Surgical resection should be complete and can be approached laparoscopically if negative margins and complete resection can be accomplished. Laparoscopic management is commonly undertaken for splenic IPTs⁷ and has been reported for other locations including the liver.⁸ We find that laparoscopy, even for oncological resections, can expedite patient recovery. However, if the resection is technically difficult or intraoperative appearance is concerning for malignancy, the surgeon should not hesitate to perform a laparotomy for a full oncological resection.

On gross examination, these lesions are firm and fleshy.⁴ Coffin *et al*⁴ described three histological patterns with (1) spindle cells in an oedematous and myxoid background, (2) spindle cell proliferation and (3) collagen formation similar to that seen in fibromatosis. The differential for these cases is typically broad and include amyloid deposition, well-differentiated

Patient's perspective

While on a business trip in October 2016, I was admitted to the emergency room with severe abdominal pain. The doctor advised for a CT scan and informed me that he found two unidentified masses in my stomach and liver. I was very concerned but more focused on getting my pain under control so I could fly back to Ohio. After a very painful flight and 3 days in bed, I was able to schedule an appointment with a liver specialist. The liver specialist did not believe the tumours were related to my pain and believed that I had developed bad irritable bowel syndrome (IBS). I was put on medicine to aid in digestion which did alleviate some of the pain I was feeling.

Throughout November and December, I had multiple MRIs, a biopsy and several appointments with the liver specialist, physician assistants and others. The liver biopsy was very stressful because I was not informed about the procedure. I was surprised by the length of the needle, requirement for being awake and shocked by the amount of blood. I wished I had been mentally prepared.

Overall, I had a terrible time getting results for my tests or speaking with my doctor or nurse to get more information. I had a hard time focusing at work and felt exhausted by trying to figure out answers. Eventually, I was told the biopsy came back 'good', but details were vague. The medical team believed the mass in my liver was probably FNH, and the second mass might be a swollen lymph node.

In mid-January, the doctor referred me to a surgeon for next steps. The surgeon recommended that I have both masses removed by open surgery. I was very nervous about such an invasive procedure—particularly because I was previously told FNH tumours typically are not necessary to be removed.

I met with a second surgeon for a second opinion in February. He took time to review the CT scan with me and my family and I could tell he was intently listening to my symptoms and concerns. He answered our questions and advised additional tests. My family and I quickly agreed with this surgeon for my procedure, particularly because he was confident the procedure could be done laparoscopically and only one of the tumours needed to be removed. The FNH tumour could stay and be monitored in the future. While I was still worried about the masses, I felt like I was finally moving forward to recovery and I was so thrilled that the procedure could be done with a minimally invasive procedure that would be a much easier recovery and limit the scarring.

I was scheduled for surgery in March. The days leading up to surgery I was excited—I was looking forward to feeling better and moving on with my life. However, once I was in pre-op I was terrified. I finally realised the full risk of having surgery and potential outcomes of the pathology.

Days following surgery I could not comprehend results of the initial frozen pathology. My husband mentioned the doctors thought the tumour that was removed could be NET, but it did not sink in. Once I was off pain medication, I finally digested the results. I was upset, to say the least. One of the most interesting symptoms about these masses was my pain would come and go. Before surgery, there were days I could barely stand up straight, and days I felt pretty healthy and was able to go running and exercise. I could not imagine I had cancer but was able to do those things.

The following 4 weeks felt like months. I had initially been told the pathology results would take around 1 week. Each day I hoped to get news, I was so anxious to know the results and felt like I was in Limbo for almost a month. My surgeon and his nurse were very helpful and kind during this time. While they were waiting for pathology, they still took time to speak with me and let me know they had not heard anything. This gave me confidence in the process and relieved some of my worry.

The day I went back to work my surgeon called and informed me the tumour was not cancerous. I was so happy it was hard for me to not cry at my desk. In addition, my pain is completely gone and I stopped taking all digestive medicine instantly. I am so thankful for the care and compassion shown by the medical team.

NET, lymphoma, myeloid sarcoma, as well as infectious aetiologies, such as mycobacterial pseudotumor and histoplasmosis. In our patient, other aetiologies such as GIST, leiomyoma or schwannoma were considerations. Workup includes exclusion of lymphoproliferative disorders. IHC staining for vimentin and smooth muscle actin is almost always positive (99% and 92%, respectively, in a series of 75 patients), with the majority also positive for desmin (69%).⁴ Cytokeratin staining is positive in 36% of cases and was negative in our case. IHC staining for IgG₄ will often reveal increased IgG₄ expressing plasma cells associated with the inflammation. Additionally, stains for CD117 and DOG1 are necessary to exclude a GIST and S100 can help exclude a schwannoma.

The pathogenesis of IPT is thought to be mediated by cytokines, such as interleukin-1 (IL-1) and IL-6,⁴ resulting in many of the systemic signs and symptoms that accompany IPT.⁵ There is a subset of IPTs that are associated with IgG₄-related disease,⁹ which are responsive to corticosteroid therapy but the histopathology was not suggestive in our patient, and her serum IgG₄ was within normal limits.

IPTs have been reported across a spectrum of patient demographics and anatomic locations. Preoperative tissue diagnosis is difficult due to radiographic appearance that can mimic neoplastic lesions and biopsies that are negative for malignancy,

which leads to the conclusion of a sampling error. Operative resection should be the treatment of choice if the diagnosis is in question and complete resection appears feasible. Surveillance after surgery should be undertaken due to the high rate of early local recurrence.^{3 5 6} There are no specific guidelines to dictate the frequency of surveillance; our approach is a history and physical exam every 6–12 months for 2 years, supplemented with cross-sectional imaging if there is suspicion of recurrence.

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Learning points

- Inflammatory pseudotumour can mimic a neoplastic process and should be in the differential of a solitary intra-abdominal mass
- Laparoscopy is an acceptable approach for a lesion with unclear diagnosis with potential benefits for more rapid postoperative recovery
- Pathological diagnosis of inflammatory pseudotumour requires exclusion of lymphoproliferative disorders and systemic IgG₄-related disease

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