Unusual association of diseases/symptoms

Acute onset of generalised pruritus as presenting symptom of Klatskin tumour

Patrick Tobbia,¹ Leslie A Norris,² Nodira Karimova¹¹Department of Internal Medicine, Cone Health, Greensboro, North Carolina, USA²Campbell University, Buies Creek, North Carolina, USA

Correspondence to Dr Patrick Tobbia, tobbia@gmail.com

Summary

A 60-year-old Korean woman presented with generalised pruritus and abdominal pain. She was initially treated with oral antihistamine therapy, but upon progression of symptoms and jaundice involvement, she was found to have a Klatskin tumour. The patient was not a candidate for surgical resection and is currently undergoing palliative care.

BACKGROUND

Cholangiocarcinoma is a tumour type that arises from the epithelial cells of the intrahepatic and extrahepatic bile ducts. This cancer is rare, but highly lethal because most patients present at a locally advanced stage and do not respond well to current therapy.¹ Risk factors for cholangiocarcinoma include chronic infestation with parasitic liver flukes, primary sclerosing cholangitis, ulcerative colitis, developmental abnormalities of the biliary tract, hepatolithiasis, hepatitis B and C infection and thorotrast exposure.^{1 2}

There are two types of cholangiocarcinoma, intrahepatic and extrahepatic. The most common type of cholangiocarcinoma is the Klatskin tumour, which is extrahepatic and is located at the bifurcation of the common bile duct. Presenting symptoms depend on the location of the tumour. Klatskin tumours typically present with jaundice and pale stools. The reported incidence in England and Wales is two cases per 100 000, with the majority of cases affecting the elderly.³ For unclear reasons, the incidence of intrahepatic cholangiocarcinoma has been rising over the past two decades in Europe, North America, Asia, Japan and Australia, while rates of extrahepatic cholangiocarcinoma are declining internationally.^{1 4–7}

CASE PRESENTATION

A 60-year-old Korean woman initially presented to our clinic with a 3-day history of mild upper abdominal discomfort, nausea and generalised pruritus. The patient reported that her air conditioning had broken down 4 days prior and that since that time she had been experiencing these symptoms. She reported that her symptoms were similar to the pruritus she experienced previously from poison ivy exposure. She denied contact with poison ivy, but did endorse recent outdoor activity in cleaning her yard. She also complained of reflux sensation. Her medications included ibuprofen, naproxen and aspirin. She was sent home on oral antihistamine therapy and a steroid taper, which was the same regimen used to treat her previous poison ivy exposure. The patient had a history of

gastroesophageal reflux disease (GERD) and was complaining of worsening symptoms and abdominal discomfort that were unrelieved by antacid therapy. She was started on esomeprazole therapy and was counselled on lifestyle modifications. No labs were drawn at this visit. Given the patient's ethnicity, jaundice was not recognised by the examining physician at her initial visit.

She returned to the clinic after several days with unresolved symptoms and new onset jaundice. Liver function tests showed elevated levels of bilirubin (15.7 mg/dl; normal 0.3–1.2 mg/dl), alkaline phosphatase (214 U/l; normal 39–117 U/l) and transaminases in the 200–300 range. Other lab results were unremarkable.

She was subsequently admitted to the hospital for further workup and underwent an abdominal ultrasound, which revealed a prominent common bile duct and intrahepatic duct. Abdominal CT showed evidence of Klatskin tumour with marked bilateral intrahepatic biliary ductal dilatation. There was no definitive evidence of metastatic disease. MRI showed a 2 cm mass in the liver hilum causing severe intrahepatic biliary dilatation in both the right and left lobes, which was consistent with CT findings. Gastroenterology was consulted and recommended percutaneous transhepatic cholangiography (PTC) and CA 19-9, which is a tumour antigen that is often increased in the presence of Klatskin tumours. CA 19-9 was elevated to 483.1 U/ml (normal <35.1 U/ml). She had positive hepatitis B core antibody, but negative hepatitis B IgM core antibody, suggesting remote infection with hepatitis B (figure 1).

PTC resulted in successful placement of bilateral 10-French biliary drainage catheters for symptomatic relief given the severity of the patient's pruritus. A brush biopsy was also obtained, but was not enough for a definitive diagnosis. The patient was evaluated by a pancreaticoduodenal surgeon and was found to have an unresectable tumour. At her most recent follow-up, the patient's pruritus had significantly improved after placement of intrabiliary stents to relieve compression caused by the tumour.

Figure 1 Abdominal CT consistent with a central cholangiocarcinoma centred in the left lobe of the liver. Obstruction of both the right and left hepatic ducts with circumferential narrowing and involvement of the left portal vein.

OUTCOME AND FOLLOW-UP

Currently, she is being followed up by palliative care and is receiving radiation therapy along with 5-fluorouracil therapy.

DISCUSSION

Although rare, cholangiocarcinomas account for approximately 3% of all gastrointestinal malignancies, with a prevalence in autopsy studies of 0.01–0.46%.¹ The increasing incidence of this cancer is concerning due to the poor response and advanced presentation of the tumour, which often rules out resection. Surgical resection remains the only potentially curative treatment with a 5-year survival rate ranging from 0% to 22% for hilar cholangiocarcinoma.⁷ Resection is possible in less than one-third of patients at the time of diagnosis.⁷ For those patients not eligible for resection, the preferred palliative option is biliary drainage with metal stents.⁷ There is no evidence to support the effect of chemotherapy on survival. The chemotherapy agents that have been studied include 5-fluorouracil monotherapy or in combination with leucovorin, methotrexate, cisplatin, mitomycin C or interferon- α .⁸ There are no randomised controlled trials that demonstrate therapeutic efficacy of radiation therapy.⁸

A common presenting symptom of cholangiocarcinoma is pruritus. Pruritus is a very common symptom, and is often overlooked or treated without further investigation, yet it is important to remember that 10–50% of cases with generalised itching have systemic disease.⁹ Causes of pruritus include dermatological, medication-related,

cholestatic, haematological, and endocrine aetiologies. Through this case we want to emphasise the broad differential that one should keep in mind while evaluating patients with pruritus, especially in those patients with no underlying skin disorder prior to empiric antihistamine treatment. In our case, the patient presented with similar symptoms to a prior episode of contact dermatitis from poison ivy exposure. She was initially treated with the same combination of steroid and antihistamine therapy without resolution of symptoms. When she returned to clinic with symptom progression, further workup led to the diagnosis of Klatskin tumour. The diagnosis was complicated by the patient's Asian ethnicity as jaundice was not initially recognised.

Learning points

- ▶ Patients presenting with generalised pruritus with no underlying skin disorder should be thoroughly evaluated for all possible pruritic aetiologies.
- ▶ Jaundice may be less apparent in patients of Asiatic ethnicity, so physicians should be more diligent in patients presenting with pruritus.
- ▶ As the only curative therapy is surgical resection, early detection is important. Patients who present with more advanced disease are rarely surgical candidates.
- ▶ For those patients who are not candidates for surgical resection, palliative options include intrabiliary stenting to alleviate symptoms.

Competing interests None.

Patient consent Obtained.

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