Case Report

Extrapulmonary inflammatory myofibroblastic tumor with synchronous involvement of the liver and appendix

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ABSTRACT

Inflammatory myofibroblastic tumor (IMT) of the liver is a rare disease, often confused with hepatic neoplasm. A 60-year-old man underwent liver resection for suspected hepatocellular carcinoma with concurrent appendicectomy for incidentally detected diseased appendix. Histopathology revealed IMT involving both the liver and appendix. We present the diagnostic challenge and approach in a patient with IMT of the liver with the synchronous incidental involvement of appendix treated successfully by liver resection and appendicectomy.

Key words: Appendix, inflammatory myofibroblastic tumor, liver

INTRODUCTION

Inflammatory myofibroblastic tumor (IMT) is said to have an indeterminate malignant potential. It was initially described in the lung, but it has been known to involve various anatomic locations. Over 250 cases of hepatic IMT have been reported. However, IMT involving the alimentary tract is rare. The spectrum of presentation ranging from those with regressing spontaneously to those with locally aggressive and metastatic behavior have been reported. Although medical management has been described, most patients undergo surgery because of preoperative suspicion of malignancy on imaging or failure of resolution of symptoms.[1-3] We present the diagnostic challenge and approach in a patient with IMT of the liver with the synchronous incidental involvement of appendix treated successfully by liver resection and appendicectomy.

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CASE REPORT

A 60-year-old male presented with pyrexia of unknown origin and weight loss over 10 months. He was detected to have a liver lesion reported to be of suppurative nature on ultrasound and computed tomogram (CT). Serum alpha-fetoprotein was 2.14 ng/mL; the carcinoembryonic antigen was 1.74 ng/mL and CA 19–9 was 0.6 U/mL. Hepatitis B and C viral serologies were negative. He was treated with antibiotics, but symptoms were persistent. After a fine-needle aspiration cytology revealed granuloma, he was advised anti-tubercular drugs. However, at the end of 2 months, there was no relief of symptoms and the lesion increased in size. Thereafter, a percutaneous tru-cut biopsy from the liver lesion was performed, which revealed atypia with suspicion of hepatocellular carcinoma. He was then referred to our unit after a right portal vein embolization (PVE) in view of the anticipated borderline volume of remnant liver for a planned liver resection. One month after PVE, CT revealed a 12 cm × 9 cm × 11 cm hypodense lesion with a thin enhancing rim with extracapsular

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extension, but no extrahepatic or vascular spread. Background liver was normal. With insignificant benefit from PVE, he underwent bisegmentectomy (five and six) and partial resection of adjacent segments seven and four. Intraoperatively, appendix appeared diseased; hence, a concurrent appendicectomy was performed.

Resection margins of the lesion were free of tumor. Microscopy revealed spindle cells in a myxoid stroma with inflammatory infiltrate consisting of plasma cells, lymphocytes, and eosinophils [Figure 1]. On immunohistochemical analysis, anaplastic lymphoma kinase (ALK) and vimentin were focally positive [Figure 2a and b]; smooth muscle actin (SMA) and desmin were negative, supporting the diagnosis of IMT of the liver. Appendix showed similar features. He recovered well and was discharged on the 10th postoperative day and was asymptomatic 3 months, later on, his last follow-up.

DISCUSSION

IMT, also known as inflammatory pseudotumor, most commonly arises from the bronchopulmonary system followed by the mesentery and liver.[1] The first IMT of the liver was reported by Pack and Baker in 1953.[4] IMT involving alimentary tract is rare and to our knowledge, while this is the eleventh reported case of appendicular IMT; synchronous involvement of liver and appendix by IMT has not been reported before in English literature.[5,6]

IMT is a tumor with indeterminate malignant potential. Though the exact etiopathogenesis is unclear, the discovery of ALK translocations in a subset of IMTs indicates that inappropriate activation of the ALK signaling pathway may be a critical step in the neoplastic transformation of myofibroblasts.[7] While usually they present in childhood and young adulthood, it can affect the elderly as well, as seen in our patient.[1,8] Unlike its pulmonary counterpart, in which patients are usually asymptomatic, most reported cases involving liver have one or more systemic symptoms. Fever is the most common presentation. Syndromic fever, malaise, weight loss, anemia, thrombocytosis, polyclonal hyperglobulinemia, and raised erythrocyte sedimentation rate, which reverts after surgical excision, are known.[1] Our patient typically presented with fever, weight loss, weakness, abdominal pain, and leukocytosis. Radiological findings of hepatic IMT are not specific and often difficult to differentiate from malignancy. It is usually hypoechoic on the US and shows delayed enhancement on CT.[9] On percutaneous biopsy, a whorled pattern of fibrosis with a plasma cell component is characteristic. Virtually, all IMTs are positive for vimentin; ALK is positive in 50% cases while positivity of SMA and muscle-specific actin vary.[1]

Spontaneous regression of liver IMTs has been reported in few patients.[10] Patients with biopsy proven IMTs, have been successfully managed with medical treatment including antibiotics, nonsteroidal anti-inflammatory drugs, and steroids.[11] Surgical resection is indicated when malignancy cannot be ruled out; the persistence of symptoms after medical therapy; when lesions increase in size and when lesion causes biliary obstruction and/or portal hypertension.[12] In our case, there was suspicion of malignancy with persistent fever. However, he did not receive steroids as the diagnosis was not possible preoperatively. Resection was feasible in spite of a borderline liver remnant volume as background liver was not cirrhotic and as we resorted to a modified parenchyma-preserving resection.

In the natural history of IMT, recurrence is known in 25–40% cases while metastasis has been reported in <5% of cases. Currently, there is no proven role of chemotherapy or radiotherapy in IMT.[1] While IMT usually involves a single organ system, the progressive involvement of multiple

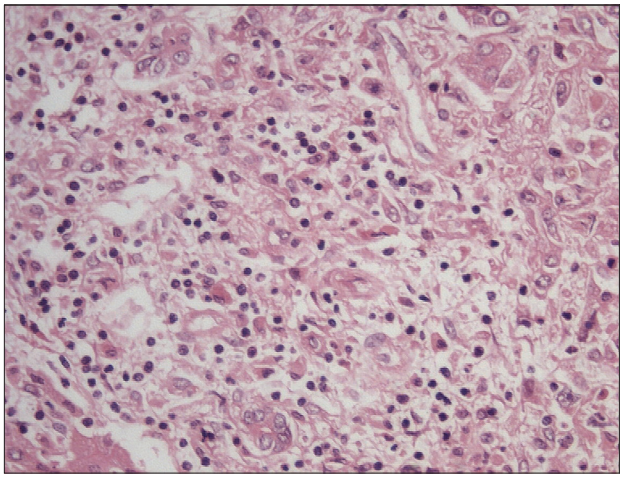

Figure 1: Microscopy of liver (H and E, ×40) showing spindle cells in a myxoid stroma with inflammatory infiltrate

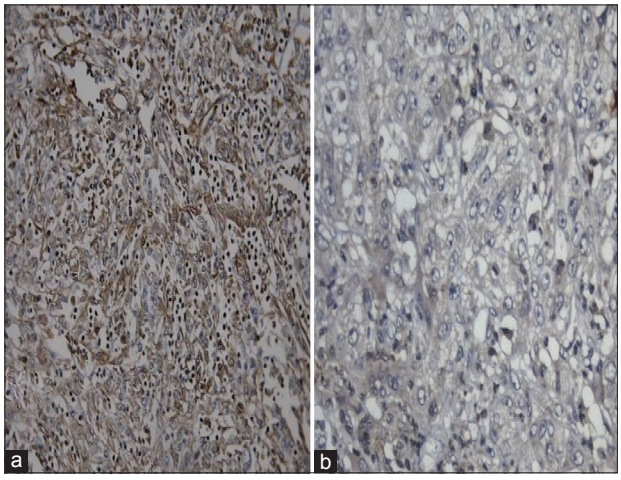

Figure 2: IHC showing vimentin (a) and anaplastic lymphoma kinase (b) positivity

organ systems has been described by Voss *et al*. in IMT involving the hepatopancreaticobiliary system.[13] However, multisystemic involvement in our case was synchronous. Concurrent involvement of omentum along with bone marrow and that of the appendix with omentum has been reported. It is unclear if multifocality represents metastatic disease.[6,14] In our case, it remains to be seen if the multifocality impacts the long-term outcome after a surgical resection resulted in relief from fever.

CONCLUSION

Percutaneous biopsy may help in establishing the diagnosis of hepatic IMT; possibly avoiding surgery if there is a complete response to medical therapy. However, resection not only establishes the diagnosis but also is potentially therapeutic. A careful survey to look for multifocality and multiorgan involvement is necessary. Long-term follow-up is essential to discern if multifocality is true or represents metastatic disease.

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Conflicts of interest
There are no conflicts of interest.

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