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CASE REPORT

Hepatic Angiosarcoma with Metastasis to Small Intestine

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ABSTRACT

Hepatic angiosarcomas are rare tumours with poor prognosis, with patients usually dying within 6 months. Metastases mainly occur in lymph nodes, spleen, lungs, bones and adrenals. Metastasis to small intestine is even rarer. Similarly, primary or metastatic angiosarcomas in small intestine are extremely rare, often present with recurrent gastrointestinal bleeding and anemia, and have an extremely poor prognosis. Both primary or metastatic intestinal angiosarcomas may exhibit epithelioid morphology. It may be very difficult to differentiate between primary and secondary cases in intestine and especially when the tumour exhibits epithelioid morphology.

Key words: *Hepatic angiosarcoma. Jejunal metastasis. Immunohistochemical markers.*

INTRODUCTION

Hepatic angiosarcoma is an extremely rare neoplasm with a rapidly fatal course. Patients usually die within 6 months of diagnosis. Metastases mainly occur in lymph nodes, spleen, lungs, bones and adrenals.[1] However, there are reports of hepatic angiosarcomas also metastasizing to the gastrointestinal tract.[2-4] Angiosarcoma occurs very rarely in the intestinal tract as either a primary or metastatic tumour and can present great diagnostic difficulty.[5]

We present a case of a 38 years old man with primary hepatic angiosarcoma involving both lobes of the liver, with metastasis to the jejunum.

CASE REPORT

A 38-years-old male presented to the gastroenterology clinic with complaints of abdominal pain and unexplained weight loss. He had been having these complaints for one year. On examination, he was a lean individual. Abdominal examination revealed tenderness in lower abdomen and mild hepatomegaly. Rest of the systemic examination was unremarkable. Laboratory tests were performed and were all unremarkable. CT scan of abdomen revealed multiple hypodense lesions in both lobes of the liver along with jejunal thickening. A possibility of lymphoma or tuberculosis was raised.

Upper G.I. endoscopy was performed. Esophagus showed whitish patches, and a hyperaemic patch at Lower Esophageal Sphincter (LES). Biopsies from stomach and duodenum were unremarkable. A small hyperaemic, thickened area was noted in the jejunum. The findings in the esophagus were indicative of candidiasis.

Histopathological examination of esophageal biopsies showed acute and chronic inflammatory infiltrate along with a number of *Candida* organisms. Histology of biopsies from gastric antrum and duodenum was unremarkable.

Histological examination of the multiple biopsies taken from the thickened, hyperaemic area in the jejunum revealed intact villous architecture. A neoplastic lesion was seen, which was composed of nests of plump cells in the lamina propria. These cells had abundant eosinophilic cytoplasm and indistinct cell boundaries giving an epithelioid appearance (Figure 1). The nuclei were round to oval with clumped chromatin and small nucleoli. The initial differential diagnoses on histology included adenocarcinoma, neuroendocrine tumour, Gastrointestinal Stromal Tumour (GIST), malignant melanoma etc. However, appropriate immunohistochemical

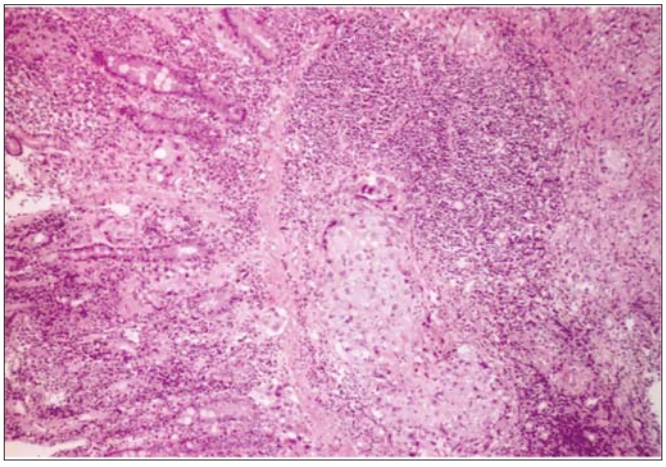

Figure 1: Jejunal biopsy showing nests of plump epithelioid cells in the lamina propria. (H&Ex20).

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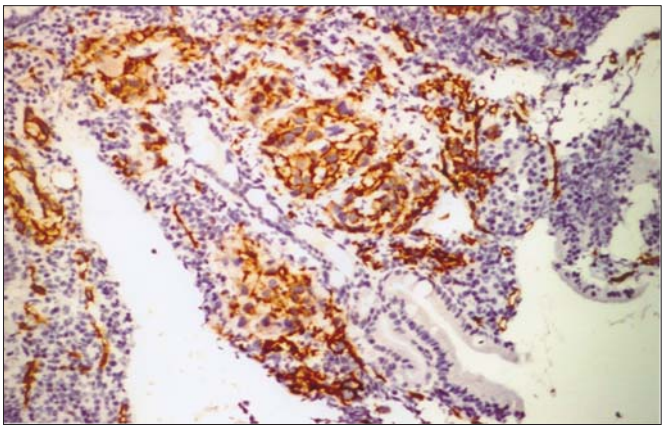

Figure 2: Nests of tumour cells showing positivity for vascular marker CD34. (Jejunal biopsy) (x20).

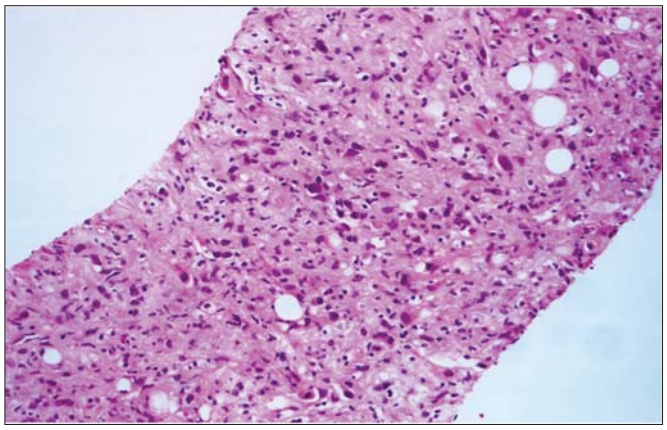

Figure 3: Liver core infiltrated by highly atypical cells lining vascular channels. (H&Ex20) .

workup excluded all these possibilities. A possibility of a vascular neoplasm was then considered and appropriate vascular immunohistochemical markers (CD31, CD34) were performed which were both positive (Figure 2). After extensive discussion of the case in the Departmental Consultation Conference, the consensus diagnosis among all the consultants was malignant vascular epithelioid neoplasm. Since the patient had multiple hypodense lesions in the liver, a liver biopsy was strongly advised.

Core biopsy of the liver on histopathological examination showed liver tissue infiltrated by a neoplastic lesion composed of freely anastomosing vascular channels. Mitotic figures were identified. Highly atypical cells exhibiting marked nuclear pleomorphism and hyperchromasia were seen lining these channels (Figure 3), and some of these cells showed small central lumina. On immunohistochemistry, the atypical cells showed positivity for vascular markers (CD31, CD34) and vimentin, and were negative for cytokeratin AE1/AE3, CD30 and LCA (leucocyte common antigen). The liver biopsy was also discussed in the Departmental Consultation Conference, and since the liver tumour exhibited greater cellular atypia and less epithelioid morphology, the consensus diagnosis was that this patient had a primary angiosarcoma of the liver with metastases to the jejunum.

DISCUSSION

Primary hepatic angiosarcoma appears as spongy, greyish white or hemorrhagic nodules involving the whole of the liver.[1] Similar was the situation in this case with multiple nodules involving the entire liver. Metastases of angiosarcoma to intestine from primaries in various organs other than liver have been described although these are very rare. These include intestinal metastases from primaries in breast, spleen, thyroid, aorta, retroperitoneum etc.[5-7]

Primary angiosarcomas of the small intestine are even rarer with a very poor prognosis.[8] Diagnosis of primary or secondary cases in intestine is very difficult, especially when the tumour exhibits epithelioid morphology.[5] There are few reports of primary small intestinal angiosarcomas with epithelioid morphology. Multifocality is seen in some of these.[8] cases may present as recurrent gastrointestinal bleeding.[9] In one report, small intestinal angiosarcoma led to perforation of bowel wall and acute abdomen.[10] Similarly, metastatic angiosarcomas in the intestine can present with gastrointestinal bleeding, anemia and abdominal pain,[5-7] have very poor prognosis and may have an epithelioid morphology.[2,5]

As mentioned in the case report section, this case showed epithelioid morphology in the liver as well as the jejunum. As discussed above, it is very difficult in such cases to distinguish between primary and secondary. However, for this case, the departmental consultation consensus was that the angiosarcoma was primarily in the liver, with the lesion in the jejunum representing metastases from the liver primary. The concensus finally proved to be correct. It is recommended that the approach of correction of coexistent lesions be adopted in such puzzling cases.

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