Case Report

Extra-gonadal yolk sac tumor of the sigmoid colon: A rare entity

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ABSTRACT

Yolk sac tumors (YSTs) in gastrointestinal tract are rare. YST of metastatic sigmoid colon, managed with surgery and chemotherapy, showed excellent response, contrary to poor prognosis has been reported in literature.

Key words: Extra-gonadal yolk-sac tumor, liver metastases, sigmoid colon

INTRODUCTION

Extra-gonadal germ cell tumors (EG-GCTs) are seldom encountered. The commonly occurring sites mentioned in case reports are the midline structures, namely mediastinum, retroperitoneum, sacrococcygeal region, and pineal gland. Primary EG-GCT of the gastrointestinal system, especially yolk sac tumors (EG-YST), is even rarely reported in literature. Hence, the prognosis and the optimal management strategy for EG-YST are not clearly understood. The case reports describe wide-spread metastases at presentation resulting in a poor overall survival. Considering the success of bleomycin, etoposide, and cisplatin (BEP) chemotherapy regimen in gonadal and metastatic YST, it has been tried in these rare cases too; however, long-term outcome remains unsatisfactory. We describe here a case of primary YST of sigmoid colon with liver metastases in a young female, treated with segmental resection of colon followed by BEP chemotherapy. The patient has been asymptomatic on regular follow-up since last seven years; this makes it all the more unique and worth reporting.

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CASE REPORT

A 28-year-old female from Punjab presented with pain over left hypochondrium and flank in early January, 2008. The pain was moderate in intensity, dull-aching, deep seated, nonradiating, and not related to food intake. The preoperative contrast-enhanced computed tomography (CECT) abdomen performed outside our hospital showed a large tumor - 12.5 cm × 7.5 cm × 5.5 cm arising from sigmoid colon, adherent to mesocolon and mesentery of small bowel and extending up to retroperitoneum. There were four metastases in liver, the largest in the left lobe. There was no intra-abdominal lymphadenopathy or any pelvic deposit. Bilateral ovaries and uterus looked normal.

On February 18, 2008, the patient underwent exploratory laparotomy in the Department of General Surgery of our institute, followed by resection of tumor-bearing segment of sigmoid colon and colo-colic anastomosis. On gross examination, the postoperative specimen showed a 12 cm × 11 cm × 8 cm tumor mass attached to the serosal aspect of the cut intestine. The outer surface of the tumor was smooth and cross-section was mucoid, yellowish pink, showing areas of necrosis and

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hemorrhage. On microscopical examination, lamina propria and muscularis propria were unremarkable. Tumor was seen to arise from serosa and extending into mesocolon. The tumor was arranged in the form of reticular, alveolar–glandular, and focal pseudopapillary pattern. The cells were large, polyhedral, with round vesicular nuclei, conspicuous nucleoli, and moderate to abundant granular cytoplasm, with intracellular and extracellular hyaline globules. Numerous Schiller–Duval bodies were seen. Frequent mitotic figures and few giant cells were noted. Overall features were those of YST [Figure 1a-d]. On immunohistochemical staining, alpha-fetoprotein (AFP) was positive in tumor cells with high background positivity. Beta human chorionic gonadotropin (B-HCG) staining was however negative. Surprisingly, AFP levels were drastically elevated postoperatively (12980 ng/ml from preoperative 210 ng/ml), though B-HCG levels did not change significantly (1.07 mIU/ml from 1.45 mIU/ml) and serum lactate dehydrogenase levels decreased marginally (1019 U/L from 1148 U/L). Postoperative CECT chest and abdomen on March 20, 2008, revealed hepatomegaly with well-defined hypodense lesions in segment II, III, and VII of liver, the largest being 6.5 × 6 × 5.5 in segment III. There was only mild mural thickening at anastomotic site. Rest of abdomen and thorax showed no abnormal findings. CT-guided fine needle aspiration cytology confirmed the hepatic lesions to be metastatic.

The patient received six cycles of three weekly chemotherapy with BEP regime – injection bleomycin 30 U (day 1), injection etoposide 150 mg (day 1–day 5), and injection cisplatin 30 mg (day 1–day 5), which were completed on September, 2008. CECT abdomen on October 11, 2008, showed a significant reduction in the size of hepatic lesions, the largest one measuring 1.1 cm × 1 × 1 cm in segment III [Figure 2]. Rest of the abdomen was normal. The patient was kept on regular follow-up with three monthly tumor markers estimation and yearly positron emission tomography (PET) scan to look for recurrence. The latest PET scan in June, 2015, showed no evidence of residual/recurrence [Figure 3], and tumor markers were within normal limits (AFP-3.43 ng/ml, B-HCG-0.1 mIU/L). The patient has been asymptomatic till date.

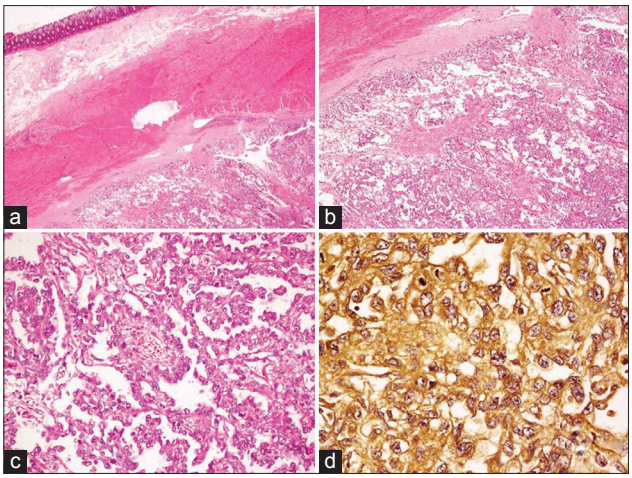

Figure 1: Histopathological details of the tumor (a) depicts unremarkable mucosa, submucosa, and muscularis propria with serosa showing a tumor (H and E, ×20). (b) Tumor is arranged in reticular, alveolar–glandular, and pseudopapillary pattern (H and E, ×40). (c) The tumor shows classical perivascular arrangement (Schiller–Duval bodies) (H and E, ×200). (d) The tumor shows uniform positive staining for alpha-fetoprotein immunostain (×400)

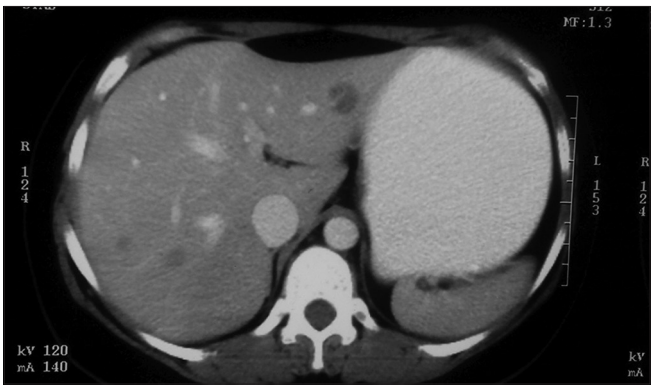

Figure 2: Postchemotherapy contrast-enhanced computed tomography abdomen showed a significant reduction in the size of hepatic lesions

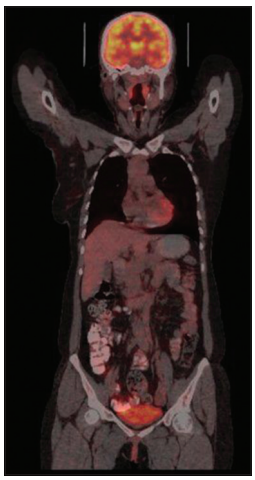

Figure 3: Positron emission tomography scan at last follow-up showed no evidence of residual/recurrence

DISCUSSION

Only in 10–15% of the cases, YST occurs in extra-ovarian locations including mediastinum, sacrococcygeal region, retroperitoneum, vagina, vulva, and pelvis.[1,2] The most popular theories of EG-YST explains that misplacement of the germ cells during the embryonic migration from cranial cavity results in their arrest in the omentum or bowel wall; the other theory holds the aberrant differentiation of multipotential gut mucosa cells responsible for gastro-intestinal, especially gastric YST. Our case can be explained by the first theory.

EG-YST typically occurs in young females, as in our case. So far, only four cases of pure YST (not associated with co-existing adenocarcinoma) of gut have been reported, all in stomach.[3-6] To the best of our knowledge, this is probably the first case of EG-YST in the hindgut. The presence of high levels of AFP and Schiller–Duval bodies is characteristic of YST, as shown in our case. Liver has been reported to be the most common site of metastases at presentation;[6] our case correlates with literature. The traditional treatment of metastatic EG-YST has been optimal surgical debulking, preceded or followed by BEP chemotherapy. Since the sigmoid colon mass was upfront resectable, surgery was done first followed by chemotherapy to handle the metastatic burden in liver. Postoperative serum AFP is a prognostic marker for long-term outcome as well as predicts disease recurrence.[7] The unusually elevated postoperative levels in this case may be due to tumor handling during operation and in part due to liver metastasis, however persistently low trends of AFP levels during the last seven years postchemotherapy, are well correlated to the disease in complete remission. The case series of gastric YSTs, treated with surgery and adjuvant chemotherapy, show a poor prognosis in general with overall survival less than a year.[6] Our case stands truly exceptional in this regard, despite a metastatic presentation to begin with. Whether hindgut YST has a favorable prognosis or any other factor is implicated can only be commented in light of newer evidence yet to be witnessed.

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Conflicts of interest

There are no conflicts of interest.

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