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Case Report

The dual that challenged us to a duel - carcinoma colon with retroperitoneal leiomyosarcoma with liver metastasis - diagnostic and therapeutic dilemma

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ABSTRACT

The presence of multiple primary cancers involving sarcoma in a single patient was first reported as far back as in 1889. We report a case of dual malignancy including carcinoma sigmoid colon and retroperitoneal leiomyosarcoma as a rare case of dual malignancy. The presentation posed a diagnostic and therapeutic dilemma. We proceeded with laparoscopic assisted sigmoid colectomy with retroperitoneal mass excision and post-operative period was uneventful. Patient was started on gemcitabine and docetaxel regimen and on regular follow up. Because of similarity of symptoms/findings between metastatic lesions and second malignancy, it may lead to mismanagement if we are not aware. When in doubt it is better to have pathological diagnosis from every lesion to avoid inappropriate management leading to inferior oncological outcome.

Keywords: Dual malignancy, Sigmoid colon, Retroperitoneal mass

INTRODUCTION

The presence of multiple primary malignant neoplasms (MPMN) in a single patient was first reported more than 100 years back. Since then, this phenomenon has been identified with increasing frequency because of early detection of by screening. There are no standard treatment protocols established to treat these situations. Combination of colonic malignancy with retroperitoneal leiomyosarcoma has not been reported in the literature.

About 10% to 20% of soft tissue sarcomas occur in the retroperitoneum and leiomyosarcoma, liposarcoma and fibrosarcoma are common histological types.[1] Because of similarity of symptoms/findings between metastatic lesions and second malignancy, it may lead to mismanagement if we are not aware.[9]

CASE REPORT

A 40-year-old women was evaluated for left iliac fossa pain for 2 months. Magnetic resonance imaging (MRI) showed 7.1×4 cm heterogenous lesion in left adnexa with 2.4 cm lesion in right lobe of liver in segment VII and another lesion 2.3 cm in the inferior aspect of left lobe of liver suggestive of metastasis. Positron emission tomography–computed tomography (PET-CT) showed fluorodeoxyglucose (FDG) avid lesion of 9 cm in the left iliac fossa. FDG avid hypodense lesion of 2.5 cm in segment VII of liver and 2.9 cm in segment III of liver. FDG avid wall thickening for length 1cm in sigmoid colon (Figure 1).

Colonoscopy showed a proliferative growth at 23 cm from anal verge and biopsy was reported as moderately differentiated adenocarcinoma. Image guided biopsy of

left adnexal mass was suggestive of spindle cell neoplasm with probability of sex cord stromal tumor or metastasis to ovary. Tumor markers were CEA – 7.2, CA 125 – 10.7, AFP – 1.7 and B-hCG – 1.2. On examination patient had a mass a size 8×7 cm occupying left iliac fossa and left lumbar region. Due to the uncertainty in the definitive diagnosis of having carcinoma sigmoid colon with ovarian and liver metastasis or double primary with metastasis, liver biopsy was taken and report came out as – leiomyosarcoma with IHC - SMA positive, DESMIN and H-CALDESMON – positive, Ki67 – 21% and S-100, CD-117, and PAN CK – negative. Patient was planned for diagnostic laparoscopy and intra operatively a mass of size 8×8 cm noted in retroperitoneal region adjacent to left ovary in the left infundibulo pelvic ligament (Figure 2). Since there was no peritoneal disease, ascites and liver surface was normal we proceeded with laparoscopic assisted sigmoid colectomy with retroperitoneal mass excision and post-operative period was uneventful.

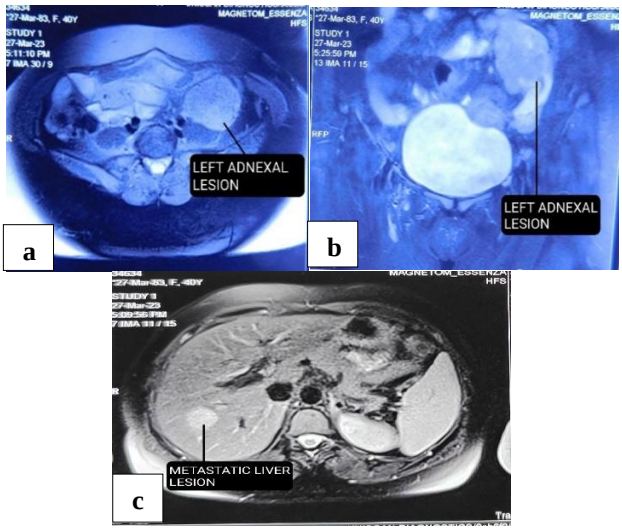

Figure 1: PET – CT showing (a) and (b) adnexal lesion and (c) liver lesion.

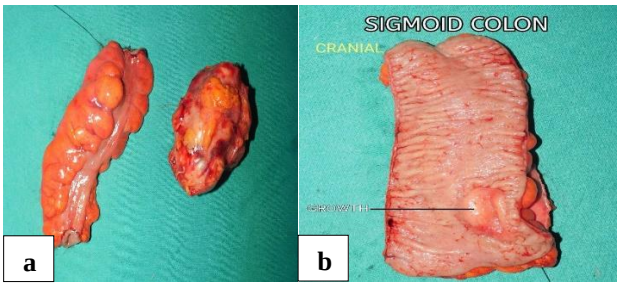

Figure 2: Specimen image (a) cut opened sigmoid colon showing growth and (b) resected sigmoid colon with retroperitoneal mass.

Post-operative histopathological examination was moderately differentiated adenocarcinoma in sigmoid colon of stage pT3N1a (Figure 3). Retroperitoneal tumor was reported as leiomyosarcoma with IHC – SMA diffuse positivity, DESMIN – diffuse positivity, Ki67 – positive and C-KIT – negative (Figure 4). Liver biopsy showed malignant spindle cell neoplasm, likely from leiomyosarcoma (Figure 5).

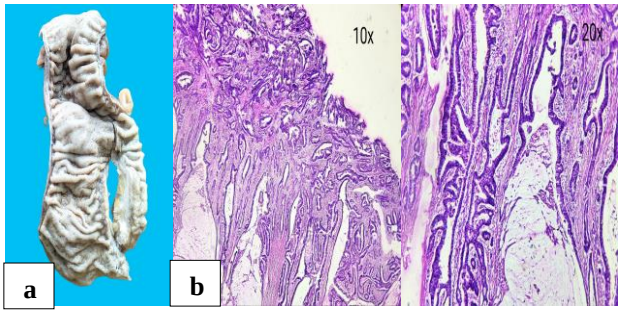

Figure 3: (a) Formalin fixed sigmoid colon, and (b) colonic mucosa exhibiting dysplasia with an adjacent infiltrating malignant neoplasm arranged in closely packed tubular glands in cribriform pattern with foci of extracellular pools of mucin - adenocarcinoma grade 2.

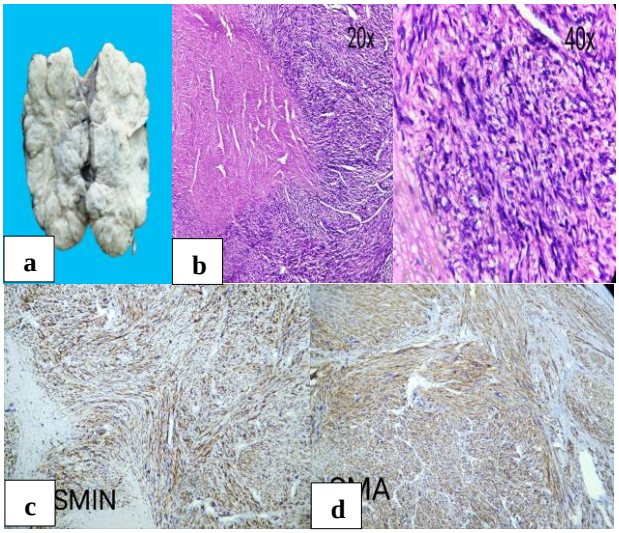

Figure 4: (a) Formalin fixed specimen of retroperitoneal tumor, (b) sections from retroperitoneal tumor showed a fairly circumscribed malignant neoplasm arranged in intersecting fascicles, vague storiform pattern and composed of spindle cells showing brightly eosinophilic cytoplasm, nuclei exhibiting moderate degree of pleomorphism, atypical mitosis of 20/10 HPF noted, (c) and (d) IHC Desmin and SMA positivity – leiomyosarcoma.

In view of metastatic disease patient was planned to start chemotherapy for leiomyosarcoma. Patient was started on gemcitabine and docetaxel regimen and on regular follow up with a plan of reassessment of liver metastasis after chemotherapy and resection if feasible.

Review of literature

In the literature one case of colonic malignancy with leiomyosarcoma has been reported where in

leiomyosarcoma of left upper limb had occurred as a second metachronous malignancy following the treatment of colonic adenocarcinoma.[2] Another case of dual primary including urinary bladder malignancy with left thigh leiomyosarcoma has been reported as possibly first case of this kind.[3] Thereby we report a case of dual malignancy including carcinoma sigmoid colon and retroperitoneal leiomyosarcoma as a rare case of dual malignancy and probably the first case in the literature to be reported. Moreover, the presentation posed a diagnostic and therapeutic dilemma. Retroperitoneal leiomyosarcoma is the second most common soft tissue sarcoma (28%) after liposarcoma and has better prognosis. The recently published METASARC observational study, among 2225 patients with metastatic STS, found a positive association of overall survival with combination chemotherapy (anthracycline based combination regimens) but recently more encouraging results are obtained by chemoembolization with polyvinyl alcohol sponge mixed with cisplatin followed by intra hepatic arterial infusion of vinblastine. With this therapy, Mavligit et al reported a 70% tumor response rate (50% regression) and a median duration of regression of 12 months.[4] Studies suggests that liver resection for hepatic metastases from leiomyosarcoma is favourable in comparison with all treatment alternatives.

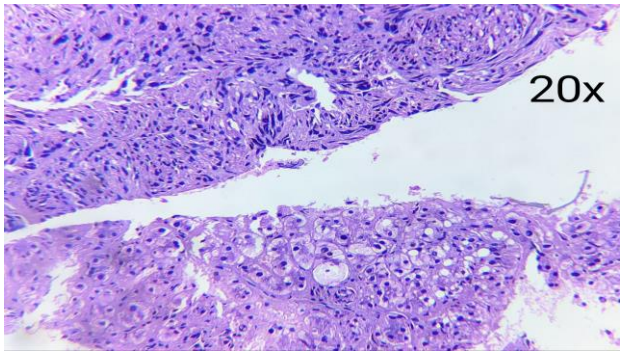

Figure 5: Malignant neoplasm arranged in storiform pattern and composed of spindle cells.

DISCUSSION

Recognition of multiple primary malignant neoplasms goes as far back as 1889, when initial criterion for its diagnosis was described by Billroth and further established by Warren and Gate as: each tumor must have a clear picture and histological confirmation of malignancy; each tumor must be topographically distinct and separated by healthy mucosa; and the lesions must not be metastases of each other. So, early diagnosis of multiple primary malignant neoplasms should be considered in patients not only during treatment of malignancy but also during long term follow-up.[5]

Multiple primary malignancies in a single patient were first described in 1879 by Billroth. The neoplasms may be limited to a single organ and single cancer type or, as in our case, involve multiple and anatomically separate organs and types. The North American Association of Central Cancer Registries (NAACCR) classifies multiple primary tumors into two categories: synchronous, in which the cancers occur at the same time (the Surveillance Epidemiology and End Results Program (SEER) definition is within two months) and; metachronous, in which the cancers follow in sequence, that is, more than two months apart. Metachronous primary malignancies are becoming increasingly common because of an increase in the number of elderly cancer survivors, greater awareness, and improved diagnostic modalities.[6] The pathophysiology behind the occurrence of multiple primary malignancies has been theorized to be common carcinogen induced multiple cancers in an exposed epithelial surface, called "field-cancerization" as seen in head-neck tumors, as a late side effect of treatment used to treat the first tumor, and a genetic predisposition to neoplasia. Other possible causal factors include persistent carcinogen exposure from environment, effects of ionizing radiation, increased use of organ transplant, and the increasing use of newer treatment modalities like hormonal manipulation, target therapies, genetic manipulation.[6] Second primary malignancies may also occur in the absence of recognizable etiologic or triggering factors. This rare coexistence of colonic malignancy with retroperitoneal sarcoma may therefore be due to coincidence, which prompted us to report this case and it lead to diagnostic and therapeutic dilemma also. If we have not evaluated the pathology of liver lesion and undertaken the surgery, the treatment might have been inappropriate.

Leiomyosarcoma

Leiomyosarcoma is generally classified into three categories: leiomyosarcoma in the retroperitoneum or peritoneal cavity, which is the most common type, of which 1/2 to 2/3 occurs in the retroperitoneum; skin and subcutaneous leiomyosarcoma, with the best prognosis among all the three; and prototype vascular smooth muscle sarcoma, which is the rarest type. Primary retroperitoneal leiomyosarcoma is mostly seen in people aged 40–70 years, and is more prevalent in women than in men (ratio of 2:1).

Retroperitoneal leiomyosarcoma may occur anywhere in retroperitoneum and outside of pelvic retroperitoneum, originating from retroperitoneal smooth muscle tissue, such as blood vessels, spermatic cord, embryonic mesonephric duct, and paramesonephric duct remnant. Retroperitoneal leiomyosarcoma exhibits three major growth patterns: completely extravascular(extraluminal) (62%), completely intravascular (intraluminal) (5%), and both extra- and intraluminal (33%). A majority of retroperitoneal leiomyosarcoma metastasizes through hematogenous dissemination, more frequently than gastrointestinal leiomyosarcoma. Autopsy findings suggest metastatic involvement of the lung (80%), bone (40%), liver (39%), peritoneum (19%), and brain (16%). These tumors can occasionally involve regional lymph nodes.[7]

Diagnosis

Computed tomography (CT) is the most important imaging method for evaluating tumor features and invasion of surrounding structures. Magnetic resonance imaging (MRI) has the advantage in determining involved blood vessels because of multi-axis planar imaging capacity, intrinsic difference in strength of signals, flow-air interface and flow-enhanced technology. The diagnostic value of PET-CT remains unclear; however, it may be an alternative choice for clarifying recurrence and metastasis.[7]

Treatment

Surgery

Surgical resection is currently the primary means of treating retroperitoneal leiomyosarcoma. Leiomyosarcoma located in the retroperitoneum is mostly >10 cm (maximum diameter) in size. The large tumor size and important organs/structure adjacent to the tumor make surgery extremely challenging. If tumor directly invades major blood vessels and involves partial resection and reconstruction of the inferior vena cava and renal vein. In fact, leiomyosarcoma is usually encapsulated, growing expansively rather than directly invading the surrounding structures. During surgery, the tumor body is more easily separated from the surrounding tissue. Even after complete resection of the primary tumor, 50% of patients may experience a relapse. Similar to the first operation, reoperation is indicated for patients without identifiable distant metastasis. Some patients with local relapse may still obtain a long term disease-free survival after tumor resection.[7]

Chemotherapy

Leiomyosarcoma is relatively resistant to chemotherapy. Although it cannot cure these patients, chemotherapy may slow down tumor progression. Doxorubicin, epirubicin, ifosfamide, or dacarbazine alone, as well as anthracycline-based combinatory regimen, are common palliative protocols for metastatic soft tissue sarcoma. In phase II clinical trial of gemcitabine plus docetaxel, the progression-free survival (PFS) and overall survival (OS) were 6.2 months and 17.9 months, respectively.[7] Preoperative chemotherapies, although not common, is an acceptable alternative, but it is considered a category 2B recommendation of the National Comprehensive Cancer Network (NCCN) guidelines 2022.[8]

Radiotherapy

Radiotherapy plays a very limited role in the treatment of leiomyosarcoma. The potential benefit of neoadjuvant radiation includes decreasing tumor size, improving resectability and improving local control. Some studies reported that when neoadjuvant radiotherapy was given the 5-year disease-free survival was 37% with an overall survival of 56% in patients treated with a dose of 50.4 Gy. Preoperative radiation is a category 2B recommendation of the NCCN, for tumors at high risk for local recurrence. In patients undergoing an R0 resection, post-operative radio-therapy should be given to patients with high-grade tumors, and those with close margins. After an R1 resection, the NCCN recommends postoperative radiotherapy if neoadjuvant therapy was not given and a boost of 10–16Gy if preoperative radiotherapy was given.[8]

Leiomyosarcoma with liver metastasis

The most common sites of metastases are the liver and lungs. The liver is a common site of recurrence after curative resection of the primary tumor. So far, the most common treatment for liver metastases from leiomyosarcoma has been chemotherapy (ifosfamide and doxorubicin regimens). The reported tumor response rates are poor, with a duration of response of only a few weeks or months. Recently, more encouraging results have been obtained by chemoembolization with polyvinyl alcohol sponge particles mixed with cisplatin powder, followed by intrahepatic arterial infusion of vinblastine. But recent studies suggest that resection of the hepatic metastasis gives better outcome in terms of overall survival among all the available treatment for hepatic metastasis. In the literature a single centre study conducted over a period of 15 years with 34 liver resections for hepatic metastasis concludes that median survival was 32 months after R0 resection and 20.5 months after R1/2 resection. The 5-year survival rate was 13% for all patients and 20% after R0 resection. In 10 patients with extrahepatic tumor at the time of the first liver resection, 6 R0 and 4 R2 resections were achieved. After R0 resection, the median survival was 40 months (range 5–84 months), with a 5-year survival rate of 33%. After repeat liver resection, the median survival was 31 months (range 5–51 months); after R0 resection and after R1/2 resection it was 28 months. Thus, the long-term outcome after liver resection for hepatic metastases from leiomyosarcoma is superior to that after chemotherapy and chemoembolization. The presence of extrahepatic tumor should not be considered a contraindication to liver resection if complete removal of all tumor masses appears possible. In selected cases of intrahepatic tumor recurrence, even repeated liver resection might be worthwhile.[4] And also a case has been reported who underwent repeated resections of recurrent metastases with three liver resections, and a survival benefit of 29 years was achieved.[1]

CONCLUSION

When we find multiple lesions, particularly in body cavities like chest and abdomen, we need to be cautious while evaluating. When in doubt it is better to have pathological diagnosis from every lesion which we did in our patient. Otherwise the diagnosis and treatment may be inappropriate leading to inferior oncological outcome. In our case proactive approach solved the dilemma leading to proper management.

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