Synchronous malignancy of adenocarcinoma of small intestine and mesenteric malignant fibrous histiocytoma-an unusual case

Dear Editor:

Synchronous double malignancies involving different organs are relatively rare and uncommon findings except in the skin and colon. Synchronous primary malignancies are becoming increasingly frequent because of an increase in number of elderly patients and improvement in diagnostic techniques. Only few cases of double malignancy have been reported in literature. The incidence of multiple primary cancers is reported to be between 0.3 to 4.3% [1]. The authors regard this case worthy of publication because this is the unusual reported case of adenocarcinoma of small intestine and mesenteric malignant Fibrous Histiocytoma (MFH). The occurrence of another malignancy of different organs in patients with known malignant tumor is known as double malignancy and is categorized into: a) synchronous: in which the cancer occurs at the same time or within six months and b) metachronous: in which cancer follows in sequence (more than six months apart).

Certain criteria have been laid down to diagnose synchronous malignancy, such as: each tumor should be separate from the other, each should be malignant and neither should be a metastasis from the other i.e. the microscopic and morphologic features of the two tumors must be entirely different [2]. There is no reliable clinical means of distinguishing second primary from metastasis and diagnosis must be made by exploration and biopsy [3]. A 60 years old female admitted with mass in the right lower abdomen with abdominal pain since 1 month with history of constipation, loss of appetite and weight. Per abdominal examination revealed an intra-abdominal lump of 7 X 6 cm in right lower abdomen with minimal transverse mobility.

CT scan revealed 7.2 X 6.7cm well-defined mass arising from the small bowel loop and another mass partially adherent to adjacent parietal wall muscle with a diagnosis of Adenocarcinoma and Malignant histiocytoma. On laparotomy, an ulcerative growth was seen arising from small bowel nearly about 30 cm from ileo-cecal junction not infiltrating the surrounding structures and a separate partially cystic and partially solid mass arising from the mesentery. No suspicious lymphadenopathy or metastases were seen in the liver and peritoneum.

Figure-1: Gross photograph showing ileal loop with ulcerative lesion

Figure-2: Gross photograph showing grey-white nodular mesenteric mass, partly cystic and solid
Grossly, the resected small bowel was 8.0 cm in length with the cut section showing ulcerative growth in the mucosa measuring about 0.8 X 1.0 cms, (Figure-1) and a separate grey-white nodular mesenteric mass measuring 5.0 X 4.0 cms with irregular margins, foci of hemorrhage, necrosis and cystic change (Figure-2).

Microscopically, sections from resected small bowel showed intestinal glands arranged in back-to-back pattern lined by epithelial cells with hyperchromatic nuclei, scanty cytoplasm forming multi layers and papillae with features of moderately differentiated adenocarcinoma (Figure-3). Sections from mesenteric mass showed tumor arranged in storiform pattern consisting of a mixed population of spindle cells and histiocytes with hyperchromatic nuclei and clear cytoplasm suggestive of MFH (Figure-4). The sections subjected later for immunohistochemistry were vimentin positive, confirming the diagnosis of MFH.

MFH is a pleomorphic sarcoma initially described by O’Brien and Stout. MFH has a peak incidence in the seventh decade of life [4]. It usually arises 50% in the lower limb, 24% in the upper limbs, 16% in the trunk and 9% in retroperitoneum [5]. It uncommonly arises primarily from the intra-peritoneal cavity, the head and neck region, dura mater, brain, lung, heart, aorta, pancreas, liver, spleen, breast, intestine and mesentery [3- 4]. Various theories of origin have been proposed, most notably those of a fibroblastic or dual fibroblastic-histiocytic tumor or origin from primitive mesenchymal cells capable of showing variable differentiation but none have been universally accepted [6]. MFH of the mesentery is an extremely rare highly malignant neoplasm with early metastatic spread. Malignant small intestine tumors are very rare accounting for 0.1-0.3% of all malignancies. Adenocarcinoma, the most common subtype of small intestinal malignancies accounts for 31.2% [7]. In our case, palliative resection and anastomoses of small bowel was done by resecting 10 cm of small intestine from both sides along with excision of the mesentery. The reasons why some patients develop multiple cancers remain obscure. The concept of “field cancerization” explains the role of same external carcinogens with an intrinsic susceptibility and its exposure to different tissues in the same individual play a role in the development of synchronous malignancies [1, 8]. The prevalent coincidence of micro satellite instability suggests that the genetic defect of mismatch repair deficiency along with germ line mutation of p53 tumor suppressor gene for development of synchronous malignancies [8]. The prognosis of a synchronous tumor is significantly lower when compared to malignancies of a metachronous nature, despite some encouraging individual results. We investigated a case of synchronous malignancy of adenocarcinoma of small intestine and mesenteric Malignant Fibrous Histiocytoma in abdominal region, which is not only rare but also unusual presentation. A therapeutic dilemma exists in deciding the course of treatment in patients with synchronous malignancies. More cases should be reported in literature so as to formulate a definite line of management in such difficult scenarios.
References


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