RARE CASE OF LEPTOMENINGEAL METASTASIS FROM APPENDICEAL ADENOCARCINOMA: A CASE REPORT

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ABSTRACT

Leptomeningeal carcinomatosis (LC) is a rare complication of cancer in which the disease spreads to the membranes (meninges) surrounding the brain and spinal cord. LC occurs in approximately 5% of people with cancer and is usually terminal. If left untreated, median survival is 4-6 weeks; if treated, median survival is 2-3 months.[1] The most common malignancies associated with LC are lung and breast cancers, melanoma, and hematologic malignancies.[2] LC from gastrointestinal cancers is generally rare. Primary carcinoma of the appendix is very rare, and its incidence has been estimated to be around 0.12 cases per million annually, and invasive adenocarcinomas account for 4-6% of all appendiceal neoplasms.[3] The most reported locations for metastasis in appendiceal adenocarcinoma are regional lymph nodes, peritoneum, ovary, colon, and liver.[4] To the best of our knowledge, there has been one report of leptomeningeal metastasis due to appendiceal adenocarcinoma in the literature thus far. The present case report describes a 61-year-old female patient with diagnosis of high grade mucinous adenocarcinoma of the appendix stage T4, N1, M1 with mucinous ascites S/P cytoreductive surgery and hyperthermic intraperitoneal chemotherapy (HIPEC) who later developed leptomeningeal carcinomatosis diagnosed by CSF analysis.

KEYWORDS: Leptomeningeal carcinomatosis (LC), appendiceal, hyperthermic intraperitoneal.

CASE REPORT

We report a 61-year old postmenopausal Asian female presented in our unit with the diagnosis of high grade mucinous adenocarcinoma of the appendix stage T4, N1, M1 with mucinous ascites S/P cytoreductive surgery and hyperthermic intraperitoneal chemotherapy (HIPEC) on 02/07/2016, with KRAS and NRAS mutation – wild type by RT-PCR, received First cycle of FOLFOX and ERBITUX based chemotherapy on 06/09/2016, presented few days later in delirious state with h/o headache, vomiting with blurring of vision. On initial evaluation, Electrolytes, ABG, CBC, BSL(R), LFT, RFT and TSH were normal. On further evaluation, MRI brain with contrast and venography showed Idiopathic Intracranial hypertension, not suggestive of neuroparenchymal, leptomeningeal or dural metastases with MR venography showing narrow caliber of dural venous sinuses with no e/o thrombosis.

CSF analysis was done to rule out metastases which revealed glucose level 101 mg/dl, protein level 20 mg/dl, chloride115mmol/L. Cytology was positive for malignant cells and negative for Cryptococcus Ag, Expert MTB and no growth on culture.

The patient now diagnosed as LC due to her appendiceal cancer. She was started on IT methotrexate twice a week starting from 16/09/2016. Till completion of her 2 doses of IT methotrexate, her CSF cytology was positive for malignant cells. On 26/09/2016, CSF cytology was negative for malignant cells. Patient received total 6 doses of IT methotrexate and her CSF is negative for malignant cells.

Patient was on continuous supportive care but later died after 3 month.

DISCUSSION

Primary carcinoma of the appendix constitutes less than 0.5% of gastrointestinal malignancies.[8] Noncarcinoid appendiceal neoplasms consist of 15–65% of all appendiceal tumors, of which mucinous adenocarcinomas usually have a more indolent course compared to more aggressive colonic type adenocarcinomas.[6] Benedix et al. in a multicenter retrospective study of 196 patients with malignant appendiceal tumors showed that the incidence of synchronous metastasis at presentation was highest for appendiceal adenosquamous carcinoma (50%), followed by the mucinous adenocarcinoma type (29%).[7] One of the characteristics of more invasive appendiceal
mucinous adenocarcinomas is the development of peritoneal seeding or pseudomyxoma peritonei. It has been shown that in patients with mucinous ovarian cancer and pseudomyxoma peritonei, the primary tumor actually rises from the appendix and probably even the ovarian tumor is secondary to the appendiceal cancer in those patients.\(^8\) The incidence of LC in cancer patients is increasing, given more prolonged survival rates.\(^9\) Isolation of malignant cells in CSF cytology is the diagnostic gold standard for LC, however the probability of a positive CSF is about 50% in the first LP but the chance increases subsequently in LPs.\(^10\) The overall prognosis for patients with LC is generally poor because most of the time it is accompanied by a more advanced and widespread metastatic disease.\(^11\) It is clear that our patient's primary tumor originated from the appendix as it was evident during surgery. Although the MRI Brain was inconclusive, CSF showed malignant cells.

It is important to keep in mind that LC can occur with any type of cancers including appendiceal adenocarcinoma, and as patients live longer, the clinical prevalence will continue to increase.

REFERENCES