Primary Malignant Fibrous Histiocytoma of the Cecum

by Patrick C Bonasso, Jared Feyko, Nezar Jrebi

We report the case of a 70 year-old female with a palpable abdominal mass and anemia. After colonoscopy was inconclusive for diagnosis, resection via laparoscopic right hemicolectomy revealed malignant fibrous histiocytoma of the cecum. This case discusses the pathology and treatment of this rare disease.

CASE PRESENTATION

A 70-year-old female presented with abdominal pain, fatigue, anorexia and weight loss. On exam, she had a palpable abdominal mass in the right lower quadrant. Her initial laboratory values revealed severe microcytic, hypochromic anemia with a hemoglobin of 7.6 g/dL. Her white blood cell count (WBC) was 5.4 thou/mcL, platelets 350 thou/mcL, creatinine 0.5 mg/dL and carcinoembryonic antigen (CEA) less than 0.2 ng/mL. Computed tomography (CT) of the abdomen and pelvis showed a mixed density lobular cecal mass measuring 7.5 x 10.2 x 7.9 cm with several enlarged lower right mesenteric lymph nodes (Figure 1).

The patient was admitted and underwent colonoscopy with biopsy (Figure 2). Pathology showed ulceration with inflammation and scattered atypical cells suspicious for malignancy. She was taken to the operating room and underwent laparoscopic right hemicoloectomy (Figure 3). She progressed through hospitalization without any complications. She tolerated a regular diet and was discharged on postoperative day three.

Final pathology revealed a neoplasm composed of large, pleomorphic giant cells and spindle cells, arranged in fascicular and storiform pattern mixed with inflammatory cells infiltration, including lymphocytes, plasma cells, polymorphonuclear leukoocytes (PMNs) and multinucleated giant cells. Immunohistochemical (IHC) staining showed positive reaction of tumor cells with α_1 antitrypsin, Vimentin and focal smooth muscle actin (SMA). The tumor was Grade III with 9 out of 21 lymph nodes positive for malignant disease.

Discussion

Malignant fibrous histiocytoma (MFH) is the most common soft tissue sarcoma in adults.[1] Common locations of this sarcoma include the lower and upper extremities (most common, 46%), retroperitoneum, abdominal cavity, head and neck.

Primary MFH in the alimentary tract is rare. There have only been 25 reported cases in the colorectal literature,[2] most commonly in the right colon. Often

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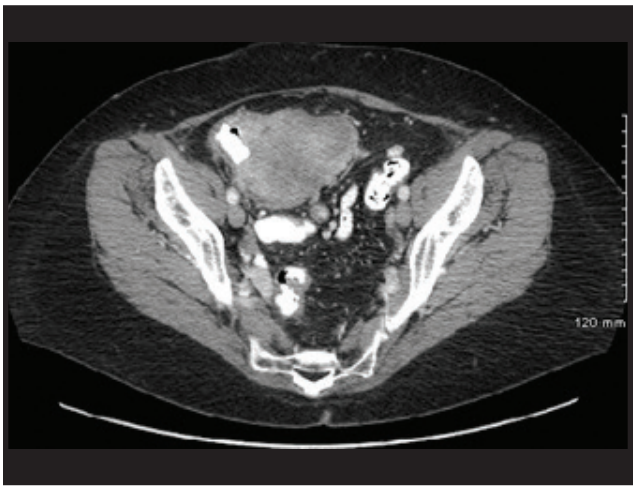

Figure 1. CT Scan

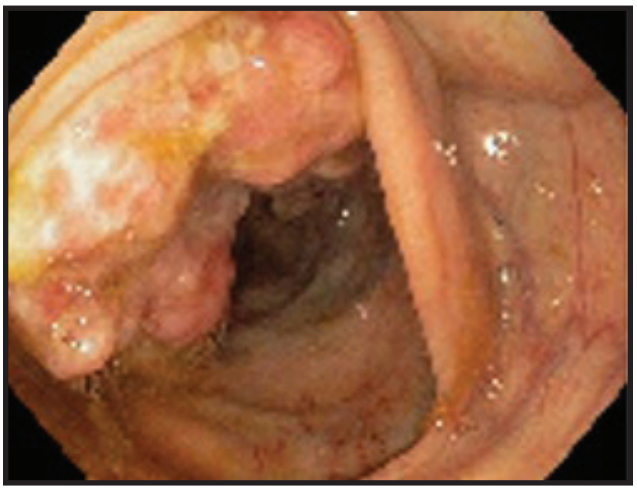

Figure 2. Endoscopic Image

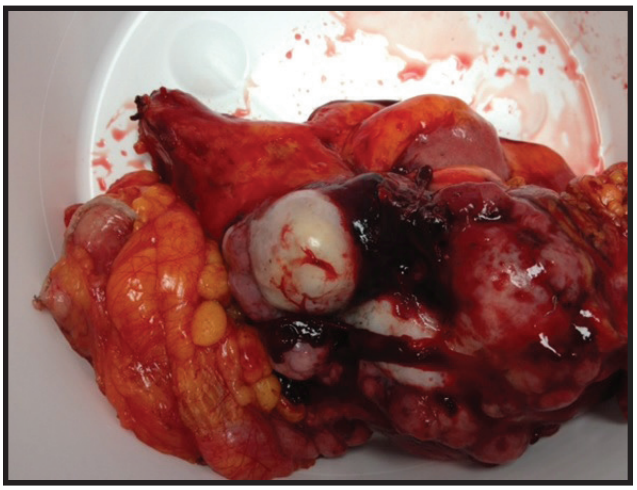

Figure 3. Surgical image after resection

these tumors are large with an average size of 8.1 cm and a range of 2-19 cm. Review of previously reported cases show a male to female ratio of 2.3:1 with an average age of 55.7 years. Most of the tumors were solitary, but some were multiple.

Common presenting symptoms include abdominal pain, fever, palpable mass, bloody stools, diarrhea and anorexia secondary to compression from the large mass. Laboratory values commonly include leukocytosis, elevated erythrocyte sedimentation rate (ESR) and elevated C-reactive protein (CRP). Predisposing factors include genetics, exposure to radiation or chemotherapy, chemical carcinogens, chronic irritation and lymphedema.

Treatment options include radical surgical resection, chemotherapy with cyclophosphamide, vincristine, adriamycin, dimethyl triazenoinidazole carboxamide (CYVADIC) or radiotherapy (5000 to 5400 cGy). The prognosis is unclear with most reports suggesting that outcomes are poor. The overall survival at two and five years is 60% and 47%, respectively.[2]

The tumor arises from primitive mesenchymal cells that retain histiocytic and fibroblastic potential. Pathology of the sarcoma shows both histiocytic and fibrous elements accompanied by pleomorphic giant cells, xanthoma cells and inflammatory cells. Pleomorphic fibroblastic cells are commonly arranged in a storiform pattern and positive for vimentin and α 1-antichymotrypsin. MFH is further classified into four types: storiform-pleomorphic, inflammatory, myxoid and giant cell. The most common type is storiform-pleomorphic.[3]

CONCLUSION

We report a rare case of malignant fibrous histiocytoma (undifferentiated pleomorphic sarcoma) of the alimentary tract treated with colon resection. ■

References

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