CASE REPORT  OPEN ACCESS

A rare case of brain metastases from appendiceal carcinoma: A case report

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ABSTRACT

Introduction: Primary appendiceal carcinoma is a rare form of gastrointestinal (GI) cancer, accounting for less than 1% of all GI tumors, with few cases involving metastatic disease. Here, we report a case, described infrequently in literature, of a brain mass found to be metastases from appendiceal carcinoma.

Case Report: We report the case of a 76-year-old male with a past medical history (PMH) of chronic lymphocytic leukemia (CLL) and appendiceal carcinoma who was transferred to our hospital after a witnessed seizure. Appendiceal carcinoma was previously diagnosed via colonoscopy for which he had surgical intervention and initially was managed expectantly. He was later noted to have an elevated carcinoembryonic antigen (CEA) with new soft tissue implants on computed tomography (CT). Omental biopsy confirmed presence of adenocarcinoma and systemic chemotherapy was initiated. The patient presented to an outside hospital where he had a pathologic fracture of the left humerus. On day of scheduled surgical repair he had a witnessed seizure. Magnetic resonance imaging (MRI) brain revealed a lesion in the left frontal lobe. The patient underwent craniotomy and resection of the lesion with final pathology revealing metastatic adenocarcinoma.

Conclusion: Although cerebral metastases are seen in approximately 5% of patients with GI cancers, local spread to peritoneal sites is more common. Distal spread is less likely to occur, documented in only 2% of appendiceal carcinoma cases. Due to this, there is no standard treatment regimen. It is important to recognize that although rare, appendiceal carcinoma does have the potential to metastasize to the brain and cause life-threatening sequelae.

Keywords: Appendiceal carcinoma, Brain metastases, GI malignancy, Seizure

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INTRODUCTION

Each year in the United States more than 100,000 patients with cancer develop brain metastases. These metastases are the most common intracranial tumor, accounting for 10–40% of all brain neoplasms [1]. Many patients with newly found brain metastases have no known cancer history, while others have undergone previous resection of their primary cancer.

Primary appendiceal malignancies are a rare form of gastrointestinal (GI) cancer, accounting for less than 1% of all GI tumors [1], with few cases involving metastatic disease. Many cases are incidentally diagnosed when a patient presents with acute appendicitis [2, 3] and definitive diagnosis occurs only after resection and review of pathology specimens. Adenocarcinoma of the appendix is so rare that no National Comprehensive Cancer Network (NCCN) guidelines exist [4–6]. Majority of patients with localized disease undergo debulking
surgery and subsequent right hemicolectomy. Evidence has shown that systemic chemotherapy (fluoropyrimidine based) in patients with advanced and metastatic disease can result in response rates similar to those in advanced colorectal cancers [4–6].

Here, we report a rare case, described very infrequently in literature, of a patient presenting with seizure and brain mass found to be metastases from appendiceal carcinoma.

**CASE REPORT**

A 76-year-old male with a past medical history of CLL stage 0 (diagnosed in 2008) and appendiceal carcinoma (diagnosed in 2016) was transferred to our hospital after a witnessed seizure. Patient’s appendiceal carcinoma was diagnosed via colonoscopy at which time he was found to have polyps, including one near the appendix, which was unable to be removed. Biopsy of these polyps revealed high-grade dysplasia of the soft tissue of the appendix (Figure 1) with corresponding CT showing multiple enlarged peritoneal, mesenteric, and pelvic lymph nodes. The patient subsequently underwent surgical intervention including a laparoscopic appendectomy, partial cecectomy, and eventual right hemicolectomy. Pathology showed moderately differentiated adenocarcinoma, staging at pT3N0. Based on stage, the patient was treated as if he had stage II colon cancer and was therefore monitored expectantly with no adjuvant chemotherapy.

He was later noted to have an elevated CEA of 4.8 and screening CT abdomen and pelvis in May 2019 revealed a new soft tissue mass in congruity with the right lobe of the liver (consistent with peritoneal implants), perirectal lesions (consistent with recurrent neoplasm), two implants superior to the bladder, and a sub-pleural nodule in the right upper lobe (likely metastasis). Resultant omental biopsy confirmed presence of moderately differentiated adenocarcinoma. At this point the decision was made for patient to start systemic chemotherapy for stage IV adenocarcinoma. He received 2 cycles of FOLFOX + Avastin; however, due to significant GI toxicity, treatment was stopped.

He remained stable until presentation to an outside hospital in January 2020 for evaluation of severe left shoulder pain with no recent trauma, injury, or inciting event. There, he was found to have a fracture of the humeral head, with high clinical suspicion of pathologic fracture, and was scheduled for surgical repair. On the day of surgery, the patient had a witnessed seizure. Subsequent MRI brain with gadolinium revealed an enhancing lesion in the left frontal lobe measuring $2.8 \times 2.4 \times 2$ cm. He was started on steroids and transferred to our hospital for further neurosurgical intervention. The patient underwent craniotomy and primary resection of the brain lesion with final pathology revealing metastatic adenocarcinoma staining positive for CDX2 and CK20, negative for CK7 (Figure 2). He tolerated surgery well and has continued to take Keppra with no further seizures. The case was later discussed at neurosurgical tumor board and the decision was made to start stereotactic radiosurgery in February 2020.

**DISCUSSION**

Brain metastases are present in 10–40% of all cancer patients with the most common primary sites including lung, breast, rectum, gastric, and colon cancers [1]. Most frequently, brain metastases are found in the gray-white junction or in watershed areas [1]. Primary appendiceal adenocarcinoma is a rare entity, accounting for approximately 1% of colorectal malignancies with associated cerebral metastases as an even rarer occurrence [2]. Although cerebral metastases are seen in approximately 5% of patient with gastric cancers, local spread to peritoneal sites is more common, whereas lymphatic or hematogenous spread is less likely to occur, documented in only 2% of appendiceal carcinoma cases [2, 7]. Metastatic appendiceal carcinoma is so rare and there is no standard treatment regimen for patients.
Using systemic chemotherapy with a fluoropyrimidine agent was shown to have a median overall survival of 55 months [6]; however, majority of treatment regimens are empiric.

The presence of seizure activity in this patient prompted the workup for intra-cranial malignancy, which otherwise may have been undetected for some time.

**CONCLUSION**

This case report highlights the unusual metastasis of appendiceal carcinoma to the brain. It is important to recognize that, although rare, appendiceal carcinoma does have the potential to invade the central nervous system (CNS) and subsequently cause life-threatening sequelae.

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Authors declare no conflict of interest.

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All relevant data are within the paper and its Supporting Information files.

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