Introduction

Intestinal polyposis syndromes, such as familial adenomatous polyposis (FAP), Cowden’s syndrome and Peutz-Jeghers syndrome, are often associated with extraintestinal manifestations though involvement in the head and neck is rare. While many of these manifestations are benign, malignant extraintestinal manifestations do occur. We report an unusual case of a malignant sinonasal mass as the first presentation of an intestinal polyposis syndrome. To our knowledge, this is the first documented case of a patient with an intestinal polyposis syndrome presenting with a non-intestinal type adenocarcinoma of the sinuses.

Case Report

A 36-year-old man presented with a one-year history of an enlarging, destructive left sinonasal mass. Extraocular movements and vision were intact despite proptosis and severe lateral displacement of the left eye. Imaging revealed a large heterogeneous, enhancing destructive mass centered in the left maxilla with extension laterally to the soft tissues of the left cheek and ulceration to the skin surface. Biopsies of the sinonasal mass showed an immunohistochemical staining profile most consistent for primary sinonasal non-intestinal type adenocarcinoma versus a metastatic adenocarcinoma of upper gastrointestinal origin.

Multiple small bowel-small bowel intussusceptions with polypoid and frond-like intraluminal bowel lesions were incidentally noted on imaging. These findings were consistent for a polyposis syndrome, such as Peutz-Jeghers syndrome or Cronkhite-Canada syndrome. Upon further interview, the patient reported a remote history of intestinal polyps for which he had not undergone further work-up. He underwent colonoscopy and subsequent open biopsies of abdominal masses and pathologic appearing lymph nodes without evidence of an abdominal malignancy.

Given the impressive primary size and location, and the uncertainty regarding the status of the multiple gastrointestinal lesions, a primary attempt of surgical control was not attempted. Instead, the patient underwent neoadjuvant chemotherapy with 12 cycles of CAP (cyclophosphamide, adriamycin, cisplatin) with significant reduction in the size of the mass. The patient ultimately underwent a left total maxillectomy, orbital exenteration, subfrontal-craniotomy and partial mandibulectomy with a latissimus dorsi myocutaneous free flap for reconstruction. Final pathology demonstrated well-differentiated adenocarcinoma with negative surgical margins.

At the time of this submission, the patient was scheduled for post-operative radiation therapy with concurrent chemotherapy.

Discussion

Sinonasal adenocarcinomas are rare and diverse malignant neoplasms that present a difficult diagnostic and therapeutic challenge. The WHO classifies malignant glandular neoplasms arising in the nasal cavity and paranasal sinuses into two main groups: salivary gland type and non-salivary gland type adenocarcinomas; the latter group is further subdivided into intestinal-type adenocarcinomas (ITACs) and non-intestinal type adenocarcinomas (non-ITACs). Definitive diagnosis of ITAC and non-ITAC lesions is complicated by the difficulty in distinguishing these lesions from true intestinal neoplasia presenting as lesions metastatic to the sinonasal compartment. These two disease entities not only portend different outcomes, but also require different therapeutic interventions.

Our case demonstrates the first known case of sinonasal adenocarcinoma in the setting of incidentally noted polyposis. Furthermore, due to the similarity in histologic characteristics between non-ITAC and metastatic adenocarcinoma of upper gastrointestinal origin, thorough evaluation with endoscopy and open biopsy was required to ensure absence of malignancy prior to initiation of treatment. Use of preoperative chemotherapy further allowed assessment of the response of all lesions prior to subjecting the patient to surgery.

Conclusions

We present the first known case of a sinonasal non-intestinal type adenocarcinoma as an extracolonic manifestation of an intestinal polyposis syndrome. Clinicians should be aware of this rare entity so that an appropriate treatment plan may be initiated.

References


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