Clinics in Oncology

Case Report
Published: 08 Sep, 2016

Lymphoepithelial-Like Carcinoma Involving a Rectal Tonsil

Quatrino G[2], Kampagianni O[1], Boudreaux CW[1], Laurini JA[1] and Grimm L[2]*

[1]Department of Pathology, University of South Alabama, USA

[2]Department of Surgery, University of South Alabama, USA

Abstract

Lymphoepithelial-like carcinoma (LELC) of the colon and rectum is an exceedingly rare diagnosis. We report a case of the disease in a 45-year-old female who first presented with rectal bleeding and a submucosal, mobile rectal mass. After workup, she underwent transanal excision of the mass. Pathology results confirmed LELC arising from a rectal tonsil. A rectal tonsil is defined as a prominent localized area of lymphoid hyperplasia, itself a rare finding. The uniqueness of the disease does not lend itself to standard treatment protocols. After multidisciplinary discussion, our patient was treated with adjuvant chemoradiation similar to a squamous cell carcinoma of the anus. She received treatment with 54Gy of radiation, 5-fluorouracil, and mitomycin C. She has recovered well from surgery and chemoradiation therapy without any signs of recurrence at 6 months. She continues to undergo surveillance with anorectal exams, endoscopic ultrasound, and CT scans.

Introduction

Lymphoepithelial-like carcinoma (LELC) is well described in the literature in certain locations in the body such as the esophagus, stomach, salivary glands, lung, thymus, bile duct, skin, uterine cervix, oral cavity, urinary bladder, mammary gland, vagina, trachea, and larynx [1]. However, it is only described in the colon and rectum seven times [2]. We describe the eighth such case and the first such case in a rectal tonsil. Several associations with LELC have been described including Epstein-Barr virus, hereditary nonpolyposis colorectal cancer, and ulcerative colitis but none within the rectal tonsil [1,3,4].

Case Presentation

A 45-year-old Caucasian woman presented to surgery clinic complaining of rectal bleeding and a rectal mass for two years that the patient presumed was hemorrhoidal disease. She was referred by her gynecologist who felt the mass by digital rectal exam, which triggered a workup by a local gastroenterologist who performed a colonoscopy and rectal endoscopic ultrasound. The colonoscopy found a single tubular adenoma in the sigmoid colon and an approximately 2cm submucosal nodule in the distal rectum without communication with anal structures (Figures 1 and 2). The EUS confirmed a 1.38cm submucosal mass without invasion of the muscular is in the rectum with concern for rectal carcinoid tumor. The patient had a history of type II diabetes mellitus and hypertension for which she was taking metformin and bisoprolol/HCTZ. Previous surgical history included transabdominal hysterectomy with bilateral salpingo-oophorectomy, appendectomy, and cholecystectomy. Family history was relevant for cardiovascular disease and absent of colorectal disease or cancer. Review of systems was positive for fatigue, nose bleeds, rhinorrhea, abdominal pain, hematuria, back pain, and headaches. She is married, works as a hair stylist, quit smoking one year prior, and denies alcohol use. On exam, she had a BMI of 32.1. Digital rectal and anoscopic exam revealed a palpable, smooth, firm but mobile 1-1.5cm mass in the posterior midline approximately 4 cm from the anal verge, just proximal to the dentate line. She was a suitable candidate for diagnostic and possibly therapeutic transanal excision which she underwent successfully (Figure 3).

Histologic analysis of the specimen demonstrated aggregates of lymphoid tissue. Malignant cells with pleomorphic nucleoli were interspersed in the lymphoid tissue confirming LELC (Figures 4-6). Tissue margins were negative for neoplasia. Cell staining for p16 was greater than 95% suggesting a human papillomavirus link. However, further testing for HPV types 6, 11, 16, and 18 were negative. EBV testing was performed but also negative.

During her first postoperative clinic visit, she complained of swelling in her right submandibular

OPEN ACCESS

Correspondence:
Gregory Quatrino, Departments of Surgery, University of South Alabama, USA, Tel: 813-758-5458; E-mail: GQuatrino@health.southalabama.edu

Received Date: *27 Jul 2016*
Accepted Date: *17 Aug 2016*
Published Date: *08 Sep 2016*

Citation:
Quatrino G, Kampagianni O, Boudreaux CW, Laurini JA, Grimm L. Lymphoepithelial-Like Carcinoma Involving a Rectal Tonsil. Clin Oncol. 2016; 1: 1078.

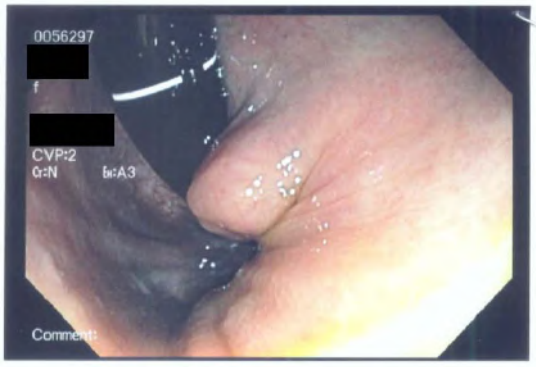

Figure 1: Retroflexed view of submucosal mass.

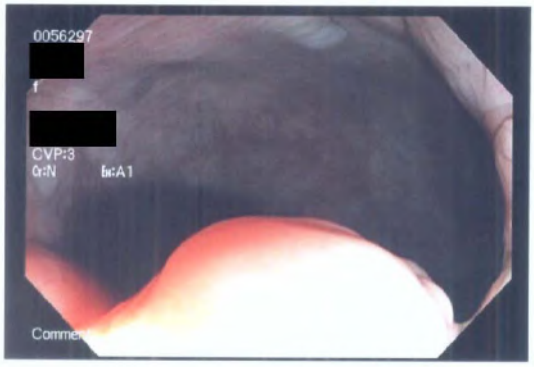

Figure 2: Submucosal rectal mass.

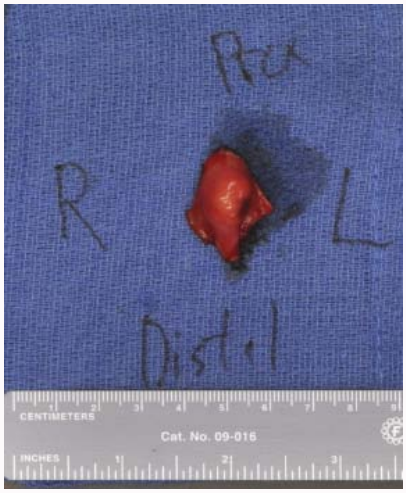

Figure 3: Gross pathology of rectal lesion.

region. She was subsequently sent to a local otolaryngologist for exam with biopsies of the nasopharynx and right submandibular gland. Pathology proved benign. Pelvic exam by gynecologic oncology with vaginal cuff biopsies was also negative for malignant or dysplastic disease. Metastatic workup including PET-CT, CEA, and LFTs was negative. CEA level was 1.2. After a multidisciplinary discussion, she was referred to medical oncology for adjuvant chemoradiation. Chemoradiation therapy consisted of 54 Gy of radiation, 5-fluorouracil, and mitomycin C over 61 days starting on post-operative day 70. During her fourth week of treatment, she developed fatigue, nausea, and desquamation of her perianal region requiring

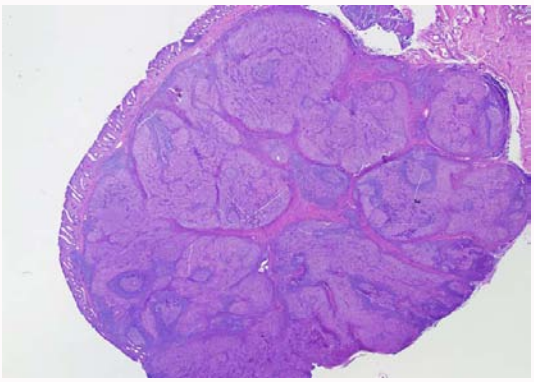

Figure 4: Syncytial sheets of malignant cells with eosinophilic cytoplasm and interspersed germinal centers.

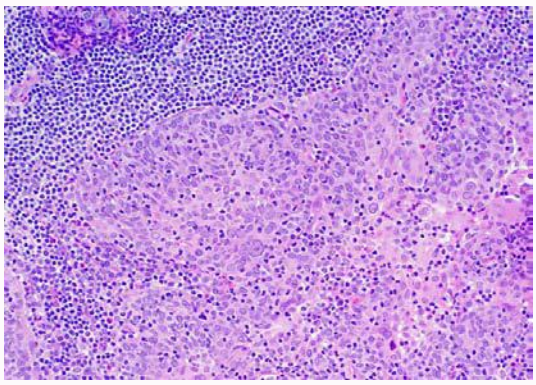

Figure 5: Malignant cells infiltrated by lymphocytes.

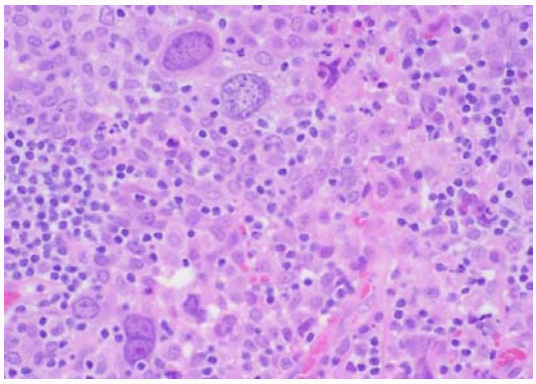

Figure 6: Moderate pleomorphism and cytologicatsypia of malignant cells.

a nearly three week break in treatment. She recovered from this well and completed therapy without further setbacks. She is now over six months removed from surgery without any signs of recurrence. For surveillance, she will undergo clinical anorectal exams every 3 months for 2 years, then every 6 months for 3 years; rectal EUS every 6 months for two years, then annually for 3 years; and annual computed tomography scan of the chest/abdomen/pelvis for 5 years.

Discussion

Lymphoepithelioma-like carcinoma of the colon and rectum is a very rare disease. There have been previous case reports with LELC of the colon and rectum associated with EBV, HNPCC, and UC. However, none have ever been described in association with the rectal tonsil. Previous case reports have described treatment modalities such

as endoscopic resection and colectomy. No further recommendations have been made for surveillance. In our case, the suspected diagnosis at the time of operation was a neuroendocrine tumor. It was not until final pathology resulted, that we discovered the LELC.

There are no standard treatments for LELC of the colon or rectum. Since tumor margins were negative and without muscularis invasion and metastatic workup was also negative, we felt she received adequate oncologic resection. In consultation with medical and radiation oncology, we felt the patient would benefit from adjuvant chemoradiation similar to squamous cell carcinoma of the anus. LELC is a radiosensitive tumor [5]. Chemoradiation has certainly been used in LELC in other locations in the body, but the result of which therapy is best and when has still to be firmly established [6-9]. We decided on a treatment regimen proven in treating anal cancer and another rare cancer of the rectum, squamous cell, administering radiation, 5-FU, and mitomycin C [10-11].

Lymphoid follicles are found widely distributed throughout the large bowel with the greatest concentration in the rectum [12]. We describe what we believe is a first for LELC, its presence within the rectal tonsil. The rectal tonsil is a dense collection of lymphoid tissue found most commonly proximal to the dentate line that will often, if present, be a cause of rectal bleeding [13]. The rectal tonsil is a rarely describe identity in the literature with fewer than 18 cases reported since 2006 [14]. Due to the paucity of both of these entities, finding the two linked is the first time noted in the literature to our knowledge.

As this is likely a first-of-its-kind cancer, there are no surveillance guidelines. We decided on our surveillance of clinic anorectal exams every 3 months for 2 years, then every 6 months for 3 years; rectal EUS every 6 months for two years, then annually for 3 years; and annual computed tomography scan of the chest/abdomen/pelvis for 5 years based on NCCN guidelines for rectal cancer [15]. We decided to include rectal ultrasound as submucosal or mesorectal lymphatic recurrence may go undetected by physical exam. It will also allow for examination of lymph nodes with the opportunity for fine needle aspiration of suspicious nodes.

Conclusion

Our patient had a unique rectal cancer. LELC of the colon and rectum is such a rare disease that developing appropriate treatment and surveillance protocols would be near impossible, but we feel that our adjuvant therapy and surveillance decisions could serve as a guide for future instances of this quite rare entity. We believe she received proper surgical and medical treatment to cure her disease based on guidelines for anal/rectal cancer and case series of LELC at other locations.

References

1. Kojima Y, Mogaki M, Takagawa R, Ota I, Sugita M, Natori S, et al. A case of lymphoepithelioma-like carcinoma of the colon with ulcerative colitis. J Gastroenterol. 2007; 42: 181-185.
2. Mori Y, Akagi K, Yano M, Sashiyama H, Tsutsumi O, Hamahata Y, et al. Lymphoepithelioma-like carcinoma of the colon. Case Rep Gastroenterol. 2013; 7: 127-133.
3. Kon S, Kasai K, Tsuzuki N, Nishibe M, Kitagawa T, Nishibe T, et al. Lymphoepithelioma-like carcinoma of the rectum: possible relation with EBV. Pathol Res Pract. 2001; 197: 577-582.
4. De Petris G, Lev R, Quirk DM, Ferbend PR, Butmarc JR, Elenitoba-Johnson K. Lymphoepithelioma-like carcinoma of the colon in a patient with hereditary nonpolyposis colorectal cancer. Arch Pathol Lab Med. 1999; 123: 720-724.
5. Ewing J. Lymphoepithelioma. Am J Pathol. 1929; 5: 99-108.
6. Liang Y, Wang L, Zhu Y, Lin Y, Liu H, Rao H, et al. Primary pulmonary lymphoeithelioma-like carcinoma: fifty-two patients with long-term follow-up. Cancer. 2012; 118: 4748-4758.
7. Suzuki I, Chakkabat P, Goicochea L, Campassi C, Chumsri S. Lyphoepithelioma-like carcinoma of the breast presenting as breast abscess. World J Clin Oncol. 2014; 5: 1107-1112.
8. Gille T. Lymphoepithelioma-like carcinoma of the skin treated with wide local excision and chemradiation therapy: a case report and review of literature. Case Rep Oncological Medicine. 2012; 2012: 241816.
9. Kessler ER, Amini A, Wilson SS, Anp-C KB, Raben D, La Rosa FG, Crawford ED. Lymphoepithelioma-like carcinoma of the urinary bladder. Oncology. 2015; 29: 462.
10. Musio D, De Felice F, Manfrida S, Balducci M, Meldolesi E, Gravina GL, et al. Squamous cell carcinoma of the rectum. The treatment paradigm. Eur J Sur Oncol. 2015; 421: 1054-1058.
11. NCCN guidelines for anal cancer. National Comprehensive Cancer Network Website.
12. Langman JM and Rowland R, The number and distribution of lymphoid follicles in the human large intestine. J Anat. 1986; 197: 189-194.
13. Farris A, Lauwers GY, Ferry JA, Zukerberg LR. The rectal tonsil a reactive lymphoid proliferation that may mimic lymphoma. Am J Surgi Pathol. 2009; 33: 1075-1079.
14. Hong JB, Kim HW, Kang DH, Choi CW, Park SB, Kim DJ, et al. Rectal tonsil: a case report and literature review. World J Gastroenterol. 2015; 21: 2563-2567.
15. NCCN guidelines for rectal cancer. National Comprehensive Cancer Network Website.