CASE REPORTS

Multiple Synchronous Granular Cell Tumors Involving the Colon, Appendix and Mesentery: a Case Report and Review of the Literature

Husain Saleh1,2, Mohammed El-Fakharany1, Maurice Frankle2

1) Wayne State University; 2) Detroit Medical Center, Detroit, MI, USA

Abstract

A granular cell tumor (GCT) is typically a benign neural tumor of Schwann cell origin that occurs in the 4th to 6th decade of life usually as a solitary painless nodule in the dermis or subcutis. It can also be found in internal organs including the larynx, bronchus and gastrointestinal (GI) tract. Within the GI tract, it is most common in the esophagus followed by colon. Colonic GCT is mostly found incidentally during colonoscopy or surgical resection as a solitary submucosal sessile nodule, although, some may cause rectal bleeding. In this report, we describe a case of a 62 year-old woman who was found to have submucosal rectosigmoid mass at screening sigmoidoscopy. Full colonoscopy and CT-scan studies revealed multiple GCTs of the colon, appendix and the mesentery, raising the suspicion of malignant metastatic disease. However, surgical resection of all the masses in an exploratory laparatomy proved them to be benign GCTs. This case emphasizes the need to consider GCTs of the GI tract when multiple asymptomatic lesions are found incidentally in the colon before any aggressive surgical intervention is undertaken. It is also the first case of GCT involving the mesentery. A literature review of GCT of the GI tract is also provided.

Keywords


Introduction

A granular cell tumor (GCT) is a rare, usually benign, soft tissue neoplasm. GCT has been encountered most frequently in skin or subcutaneous tissues of the chest and upper extremities, tongue, breast and female genital region [1]. GCT is rarely found in the gastrointestinal tract, with the esophagus representing the preferred location [2]. Colonic GCT is an exceedingly rare neoplasm that is usually found incidentally on colorectal examinations [3-7].

Most studies represent case reports describing GCT as a solitary mass lesion. In this article, we report a case of a woman who was found, during routine exam and screening colonoscopy, to have multiple GCTs involving the rectosigmoid colon, appendix, cecum and mesentery, all of which were successfully treated by surgical resection. On reviewing the literature, this appears to represent the first case of multiple GCTs of the colon extending to the mesentery.

Case report

The patient was an otherwise healthy 62 year old woman who was found to have a rectal mass on a routine annual examination by rigid sigmoidoscopy. The patient had a history of anemia, gastroesophageal reflux disease and hysterectomy for uterine leiomyomata. Subsequent flexible colonoscopy revealed a 3.0 cm intramural mass at the rectosigmoid junction and cecal, appendiceal and mesenteric masses. Further evaluation by cystoscopy disclosed urethral stricture and stenosis. Based on these clinical findings and on gastroenterologist consultation, carcinoid tumors probably metastatic to mesenteric lymph nodes were suspected. The decision was made to perform surgical resection and exploration. Examination of the patient showed no evidence of skin or subcutaneous lesions.

On exploratory laparotomy, the rectosigmoid mass was palpable, intramural and freely mobile but it could not be easily defined for excision. On further exploration, at least three distinct additional nodules were found in the appendix, cecum and mesentery. Few additional minute nodules less than 0.5 cm were noted in the rectosigmoid colon. The appendiceal, cecal and mesenteric nodules were excised and sent for histologic examination.
Pathologic examination: grossly, the lesions were firm, poorly defined unencapsulated with grey dense cut surfaces. The masses ranged in size from 0.5 cm to 3.0 cm, and the largest one involved the rectosigmoid. Microscopically, all the resected lesions were composed of solid nests and ribbons of round to polygonal cells separated by slender fibrous septae (Figs. 2, 3). The cells had small nuclei and mostly inconspicuous nucleoli, and the cytoplasm was abundant eosinophilic and finely granular (Fig. 4). Only rare mitoses were seen. The great majority of cells showed both cytoplasmic and nuclear reactivity for S-100 immunostain (Fig. 5), which confirmed our diagnosis of GCTs.

**Fig 1.** Colonoscopic view of the rectosigmoid submucosal mass with apparently rounded nodule and intact mucosa.

**Fig 2.** A low power magnification showing a submucosal nodule with intact mucosa. There are sheets of infiltrating tumor cells (H&E, X100).

**Fig 3.** The mesenteric nodule shows nests of tumor cells and surrounding fibrous septae, within fatty tissue (H&E, X100).

**Fig 4.** The tumor cells are large round-polygonal with abundant eosinophilic granular cytoplasm. The nuclei are oval to round, uniform with small nucleoli (H&E, X100).

**Fig 5.** The tumor cells show nuclear and cytoplasmic reactivity with S100 protein immunostain (S100 protein, X400).

**Discussion**

Granular cell tumor is believed to originate from the Schwann cells. A close association of GCTs with peripheral nerves has been documented [8], and reactivity to S-100 immunostain, both nuclear and cytoplasmic, confirmed its origin of Schwann cells. These tumors have been subsequently found to be positive for immunostains that label myelin proteins and myelin associated glycoproteins, which further confirms this origin [9].

In the gastrointestinal tract, GCTs are rare and mostly found in the esophagus, followed by the colon [3-5]. They are usually detected incidentally during endoscopic examinations as solitary small nodules, mostly less than 2 cm, covered by normal appearing mucosa [3, 4, 8, 10]. In this sense, the clinical impression is usually either a submucosal lipoma or a polyp. Very rarely, however, multiple gastrointestinal
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GCTs can be seen in the same patient [11-14]. Interestingly, Eriksen et al [15] reported the presence of tubulovillous adenoma in the vicinity of GCT.

Because it is virtually impossible to recognize a colonic GCT by gross appearance during endoscopy, the final diagnosis of GCT depends on pathological findings. The tumor is characteristically composed of plump histiocytelike, bland-looking neoplastic cells with abundant granular eosinophilic cytoplasm containing acidophilic, PAS-positive, diastase-resistant granules. The nuclei are small, uniform with virtually absent mitotic figures. By immunostaining, neural markers, including S-100 protein or NSE are expressed uniformly [16, 17] .

Colon GCTs are extremely rare lesions, most of which present as solitary nodules. Endo et al [16] reported 33 cases of colorectal GCT in Japan, and Rossi et al [10] found 55 patients diagnosed with GCTs of the colon in reviewing the literature in 2000. Most of the reported colonic GCTs have been typically found in the cecum, rectum, anal canal and ascending colon.

Although mostly benign, malignant GCTs have been reported. The most important indicator of malignancy is the tumor size, with more than 60% of metastatic GCTs being larger than 4 cm in diameter [7, 18, 19]. On the other hand, in most colonic GCTs, the tumor size is less than 2 cm and the tumor is well separated from the muscularis propria. Since this tumor is considered to be usually benign, endoscopic removal has been the most appropriate choice of treatment for colonic GCT [3, 10, 16, 20].

A minority of GCTs in the colon were reported as multiple masses rather than solitary nodules. Yamada et al [21] reported a case of eight GCTs all of which were in the ascending colon. Melo et al [18] reported a case of 52 GCTs, spanning the entire colon from cecum to sigmoid colon. He suggested that in such cases, a long period of observation with repeated colonoscopy may be more appropriate than an immediate aggressive approach.

Malignant GCTs are extremely rare and we could find only few reports of such cases involving the GI tract. Yoshizawa et al described a case of malignant GCT including the esophagus in a 71-year old man who died from a metastatic tumor to the liver and pleural fluid [22].

During a thorough review of the literature, we failed to find any colonic GCT that also involved the mesentery. Hence, our case appears to be the first report of colonic multiple GCTs that also involved the mesentery. All the tumor nodules in our case had similar histomorphologic features of benign GCT including bland tumor cells, very rare mitosis and a maximum size of 3.0 cm or less. The presence of GCT nodules in the mesentery is somewhat perplexing and difficult to explain. It may be a result of a direct extension of the tumor from the muscle propria into the mesentery, or a synchronous development of a tumor from the Schwann cells present in the mesentery.

To summarise, we report a case of asymptomatic multiple GCTs involving the sigmoid colon, cecum, appendix and mesentery discovered incidentally during screening colonoscopic examination in an otherwise healthy woman. Gastroenterologists, surgeons and pathologists need to include this possibility in their differential when encountering submucosal or intramural solitary or multiple nodules of the GI tract.

References


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