

Case report-Olgu sunumu

Granular cell tumor of the esophagus

Özofagus yerleşimli granüler hücreli tümör

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Abstract

Granular cell tumors are rare lesions and only 4-6% of cases were located in gastrointestinal system. Approximately one third of these cases occur in the esophagus. They are usually small and asymptomatic lesions and malignancy rate is approximately 2-4%. They are usually detected incidentally, however may be cause some nonspecific symptoms in patients. Although endoscopic follow up is sufficient for small and asymptomatic tumors, surgical or endoscopic resection is recommended in symptomatic patients and lesions with malign potential. We report here in, a case of granular cell tumor, wich was detected in distal esophagus on upper gastrointestinal tract endoscopy and resected by endoscopic polypectomy, in a patient with nonspecific dyspeptic complaints.

Keywords: Granular cell tumor, esophagus, endoscopic polypectomy

Özet

Granüler hücreli tümörler nadir lezyonlardır ve vakaların sadece % 4-6'sı gastrointestinal sistemde lokalize olur. Bunlarında yaklaşık üçte biri özofagusta ortaya çıkar. Genellikle küçük ve asemptomatik lezyonlardır ve malignite oranları yaklaşık %2-4 arasındadır. Genellikle tesadüfen saptanırlar ancak bazı nonspesifik semptomalara da sebep olabilirler. Küçük ve asemptomatik lezyonlarda endoskopik izlem yeterli olabilirken semptomatik hastalarda ve malignite potansiyeli olan lezyonlarda cerrahi veya endoskopik rezeksiyon önerilmektedir. Bu yazıda, nonspesifik dispeptik yakınmalar nedeniyle tetkik edilen, üst gastrointestinal sistem endoskopisinde distal özofagusta submukozal polipoid lezyon tespit edilerek endoskopik polipektomi ile rezeksiyon uygulanan ve granüler hücreli tümör 5 tanısı konulan bir vaka bildiriyoruz.

Anahtar sözcükler: Granüler hücreli tümör, özofagus, endoskopik polipektomi

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Introduction

Granular cell tumors (GCT) were initially described by Abrikossoff in 1926 [1]. Most of these neoplasms are generally located in tongue, oral mucosa, skin and subcutaneous tissue, however, they can be detected in different parts of body such as respiratory tract, biliary tract, breast, external genitalia, cranial and peripheral nerves. They are generally benign and asymptomatic lesions and malignancy rate is approximately 2-4%. The tumor has been rarely seen in gastrointestinal (GI) tract and approximately one third of these cases occur in the esophagus. They are usually located in the distal part of esophagus [2-4]. Nonspecific symptoms such as epigastric pain, dyspepsia, nausea and vomiting can be observed in some patients [5, 6].

We report here in, a case of granular cell tumor of esophagus with complaints of epigastric pain, nausea and vomiting, wich was detected in distal esophagus on upper gastrointestinal tract endoscopy and resected by endoscopic polypectomy.

Case report

Thirty-four year old women applied to gastroenterology clinic with nausea, vomiting and epigastric pain for four months. She has no any diseases in her history and she has been used esomeprazole 40 mg once in a day for dyspeptic complaints. Her physical examination and laboratory findings were normal. Upper GI endoscopy was performed to evaluate these complaints. Yellowish-white polypoid lesion in distal esophagus at the 37th cm from incisors, covered by normal mucosa and approximately 6-7 mm in diameter, antral polyp and antral gastritis were detected in upper GI endoscopy (Figure 1). Endoscopic polypectomy was performed to polypoid lesions in the esophagus and antrum. Histopathology of antral polyp was reported as hyperplastic polyp. Tumoral lesion, which was regular and growth with nodular pattern under the stratified squamous epithelium was detected on the histologic examination of esophageal polypoid lesion. Tumoral cells seen as uniform, with normochromatic nucleus and wide granular pink cytoplasm. Tumor cells showed periodic acid-Schiff [PAS] positivity in their cytoplasm and immunoreactivity for S100 protein and vimentin [Figure 2, 3]. C-Kit, CD34, smooth muscle actin (SMA), desmin and HM45 were negative on immunohistochemical assessment. Due to this findings, lesion was reported as granular cell tumor. Two months after the excision of lesion, upper GI endoscopy and endoscopic ultrasonography (EUS) were performed and not detected recurrences or any other pathology. Granular cell tumors have been usually seen at head and neck regions and only 4-6% of cases were located in GI system [7]. Approximately one third of these cases occur in the esophagus [2, 4]. GCTs generally shows a female predominance, however, esophageal GCTs are frequently observed in men [7, 8]. Distal esophageal lesions may be cause of symptoms such as regurgitation, epigastric pain and discomfort due to affect on esophageal motility. GCTs are also may cause nonspecific symptoms such as nausea, vomiting, epigastric pain and dyspepsia as in our case, however, they are usually detected incidentally. [5, 6, 9, 10]. The endoscopic appearance of GCT is usually a yellowish submucosal lesion covered by normal appearing mucosa and less than 20 mm as in our case. In Kaplan's report, endoscopic appearance of tumor was described as aphthous lesion [10]. GCTs are usually solitary lesion localized in distal esophagus, however, multifocal GCTs have been reported as well [9, 11]. The most characteristic histologic feature of these lesions is innumerable fine cytoplasmic granules and diffuse PAS positivity of the cytoplasm. They show immunoreactivity for S-100 protein, vimentin, neuron-specific enolase [NSE], CD68 and CD57 [12-15]. PAS, S-100 and vimentin positivity were reported in our case. GCTs are generally benign lesions and malignancy rate is reported as approximately 2-4% [12, 16]. The proposed histologic criteria of malignancy of GCTs include tumor necrosis, tumor cell spindling, large nucleoli, increased mitotic activity, high nuclear to cytoplasmic ratio, and pleomorphism. Tumors fulfilling at least 3 of these criteria are classified as malignant [17]. Local recurrence following treatment, rapid and recent growth exceeding 4 cm and infiltrative growth pattern should be considered as possibility of malignancy [18, 19]. Periodic endoscopic monitoring is sufficient for small and asymptomatic lesion. Surgical and endoscopic resection are recommended for lesions which was causing dysphagia, greater than 1 cm, showing rapid growth, transmural infiltration and suspicion of malignancy [5, 6, 20, 21]. EUS is helpful for detection of localisation of tumor, involvement of lymph-node and therapeutic evaluation. Lesions which was limited in the submucosa without involvement of muscularis propria can be resected by endoscopic polypectomy [22]. In a study by Yasuda et al. [23] reported that, criteria for endoscopic resection are small tumor size (< 20 mm), tumor unattached to the muscularis propria and absence of malignancy criteria. Excision by biopsy forceps or endoscopic polypectomy is more available procedure for small lesions and has a low cost [15, 22]. Laser and diatherapy loop are new therapeutic options, however, laser treatment is expensive and not always available and diatherapy loop has a high risk of perforation [23, 24]. In our case, lesion was resected by endoscopic polypectomy without pre-evaluation with EUS or biopsy, because it was firstly considered as a polyp, and no any

complication was observed. It was reported that, recurrence has not been observed after the surgical or endoscopic excision in a follow-up period ranging from 6 months to 5 years [3, 20, 22, 24, 26]. There was no recurrence in our case, on the control upper GI endoscopy which was performed 2 months after the excision of lesion. In addition, EUS was performed to evaluate submucosal pathology, infiltration and lymph-nodes, but no any pathology was detected in the esophageal wall or lymph-nodes. Due to this findings lesion was considered as benign and endoscopic monitoring was planned. The coexistence of benign GCT with esophageal squamous carcinoma or, bronchial, gastric and mamarian adenocarcinomas have been reported, although there is no any research which was evaluated real possible association and contributing factors [15]. Laboratory findings, abdominal ultrasonography, colonoscopy and chest X-ray were normal in our patient.

In conclusion, GCT should be taken into consideration in the differential diagnosis of polypoid or small submucosal nodular lesion which was detected in the esophagus. Small and asymptomatic lesions can be safely follow-up periodically with endoscopy, however lesions with rapid and infiltrative pattern of growth and local recurrens should be evaluated carefully because of malign potential. Patients should be assessed about coexistence with other malign dieases.

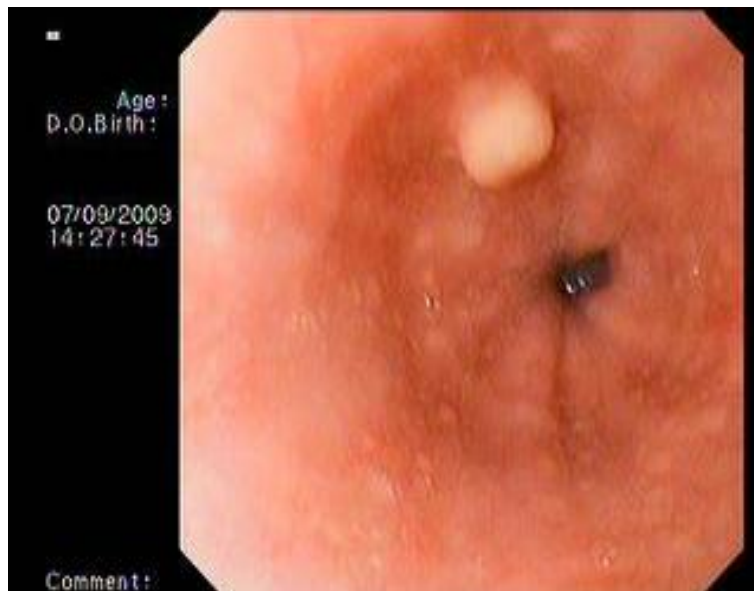


Figure 1. Upper GI endoscopy shows approximately 6-7 mm in diameter, yellowish, nodular/polypoid lesion located in the distal esophagus, impressed into the submucosa.

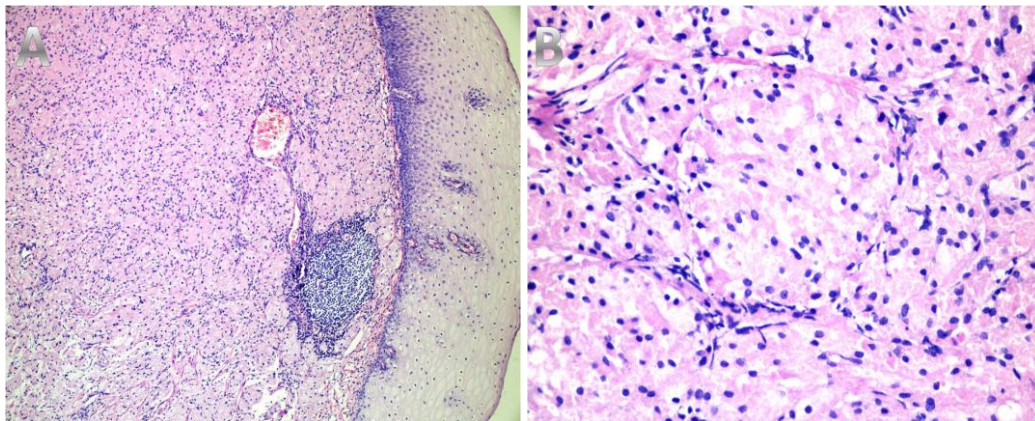


Figure 2. Granular cell tumor. Low power view of the tumor with overlying squamous epithelium, H&E x100 [A]. High power view of the tumor showing innumerable fine cytoplasmic granules, H&E x400 [B].

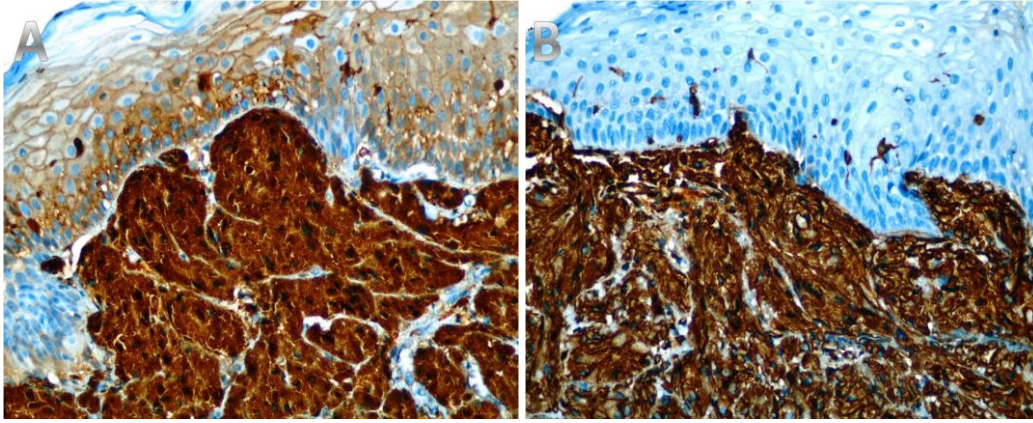


Figure 3. Immunohistochemical S-100 [A] and Vimentin [B] positivity of the neoplastic cells in granular cell tumor, x400 [DAB].

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