Togetherness of the Multiple Diverticulosis of the First Part of the Duodenum with Neurofibromatosis Type I: A Case Report

Nörofibromatozis Tip 1 ile Duodenum Birinci Kısımda Multiple Divertikülozis Birlikteliği: Olgu Sunumu

Nörofibromatozis Tip 1 ve Duodenal Divertikülozis / Neurofibromatosis Type 1 and Duodenal Diverticulosis

Ozgur Turk, Hasan Polat
Sivrihisar State Hospital, General Surgery Department, Eskisehir, Turkey

Özet

Duodenal divertikülozis klinik pratikte sıktır. Duodenal divertiküllerin büyük çoğunluğu duodenum ikinci kısımda yerleşmektedir; sıklığı %67 olarak bildirilmiştir. Üçüncü ve dördüncü kısımın sıklığı ise %20 dir. Duodenum birinci kısım yerleşimli multiple divertikülozis nadir karşılaşılan bir vakadır. Nörofibromatozis Tip 1 ile duodenal divertikülozisin birlikteliği ile ilgili bir literatür bilgisi yoktur. Biz burada tesadüfen duodenum birinci kısım divertikülosisi tanısı alan ciltte multiple nörofibromatozis nodülleri olan bir hastayı sunduk. Nörofibromatozis bir çok organı etkileyebilen bir hastalıktır. Sindirim sisteminde karsinoid tümorler ve nöroendokrin tümörler sık karşılaşılmaktadır. Eşlik eden gastrointestinal hastalığın tanısı için endoskopik inceleme ve abdominal görüntüleme yöntemleri faydalıdır. Sonuç olarak Nörofibromatozis Tip 1 bir çok sistemi etkileyebilmesinden dolayı dikkatli değerlendirilmesi gereken bir hastalıktır.

Anahtar Kelimeler

Duodenum; Divertikül; Nörofibromatozis

Abstract

Duedonal diverticulosis is common in clinical practice. Most of duedonal diverticulosis is located at the second part of duodenum; incidence is reported as 67%, incidence of third and forth part is %20. Multiple diverticulosis of the first part of the duodenum is a rare case. There is not any knowledge togetherness of neurofibromatosis type 1 and duedonal diverticulosis. Here we reported incidentally diagnosed multiple diverticulosis of duodenum that is located in the first part in a neurofibromatosis type 1 patient with multiple neurofibromatosis nodules. Neurofibromatosis could affect various organs. Along the digestive system carcinoid tumors and neuroendocrine tumors are more encountered diseases. Endoscopic examinations and abdominal imaging may be useful for the diagnosis of gastrointestinal coexisting disease in neurofibromatosis type 1 . In conclusion neurofibromatosis type 1 could affect multiple systems and it should evaluate carefully.

Keywords

Duodenum; Diverticulum; Neurofibromatosis

DOI: 10.4328/JCAM.3112 Received: 27.11.2014 Accepted: 16.12.2014 Printed: 01.04.2014 J Clin Anal Med 2014;5(suppl 2): 197-9
Corresponding Author: Ozgur Turk, Sivrihisar State Hospital General Surgery Department 26040 Eskisehir,Turkey.
GSM: +905054403377 F.: +90 2227112002

Introduction

Diverticulosis can be seen along the all digestive tract. Duedonal location is the second most common site following colon and jejunum and ileum than other sites. Duedonal diverticulosis (DD) is common in clinical practice. Most of DD is located at the second part of duodenum; incidence is reported as 67%, incidence of third and forth part is %20 [1]. DD located in first part of duodenum is uncommon. 90% of DD is asymptomatic and diagnosed incidentally during clinical investigation and radiological imaging. When it is symptomatic, the most common symptoms are abdominal pain, nausea ,vomiting and rarely lack of appetite. DD is usually diagnosed during endoscopy procedure including Endoscopic Retrograde Cholangiopancreatography (ERCP) and computed tomography [2]. DD can be single or multiple; up to five and more DD reported in the literature. Multiple diverticulosis of the first part of the duodenum is a rare case. Neurofibromatosis type 1 (NF1) is a multisystem disease affect skin, skeletal system, cardiovascular system and digestive system. Gastrointestinal system symptoms are associated with constipation and gastrointestinal stromal tumors [3]. Diagnosis of NF1 depends on seven cardinal diagnostic criteria; two of them must be for diagnosis. There is not any knowledge togetherness of NF1 and duedonal diverticulosis. Here we reported incidentally diagnosed multiple diverticulosis of duodenum that is located in the first part in a NF1 patient with multiple neurofibromatosis nodules. We thought that this case is interesting because of location and size of diverticulum and accompanying NF1.

Case Report

A 49-year-old man admitted to hospital with abdominal pain, vomiting and nausea. His compliant was ongoing for one year. Medical history of patient and family history were not significant. His first physical examination revealed us multiple cutaneous and subcutaneous lesions disseminated on abdominal skin (Figure 1a). These lesions were various size and different locations. Cutaneous lesions evaluated as neurofibromas (Figure 1b). There was a Café-au-lait macule at left chest wall (Figure 1c). Skin fold freckling (Crowe's sign) observed in axilla and evaluated as cardinal diagnostic criteria in further examination (Figure 1d). There was increase of pigmentation all over body skin. Abdominal ultrasound revealed normal and blood samples taken. Hematological parameters and biochemical study resulted in normal ranges. Upper gastrointestinal system endoscopy showed us multiple duodenal diverticulosis located at the first part of duodenum (Figure 2). Three adjacent diverticula were located at the posterior wall of first part of duedonal bulb. Duodenal mucosa was normal, no diverticulitis sign determined. Minimal non specific gastritis detected in antrum. Multiple biopsies were taken from antrum and corpus. There was not any other intraluminal pathology. Biopsy revealed chronic gastritis and helicobacter pylori was negative in giemsa staining. Lansoprozole treatment at 30 mg dose used for one month and patients complaints regressed. Further diagnosis of NF1 and duodenal diverticulosis advised to the patient but he refused treatment and examination. Because of the patient's negative consent and lack of symptoms of diverticulosis no further treatment applied. The patient is still followed in our clinic.

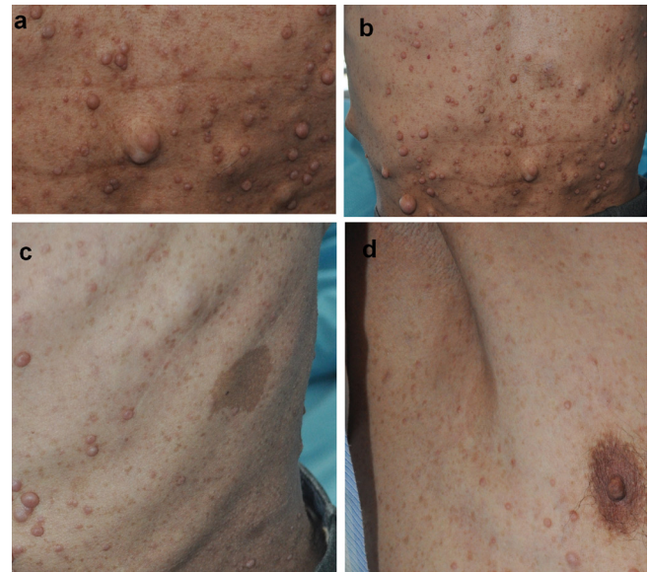

Figure 1. Multiple cutaneous and subcutaneous lesions disseminated on the abdominal skin(a), Cutaneous lesions evaluated as neurofibromas(b), Café-au-lait macule at left chest wall(c), Skin fold freckling (Crowe's sign)(d)

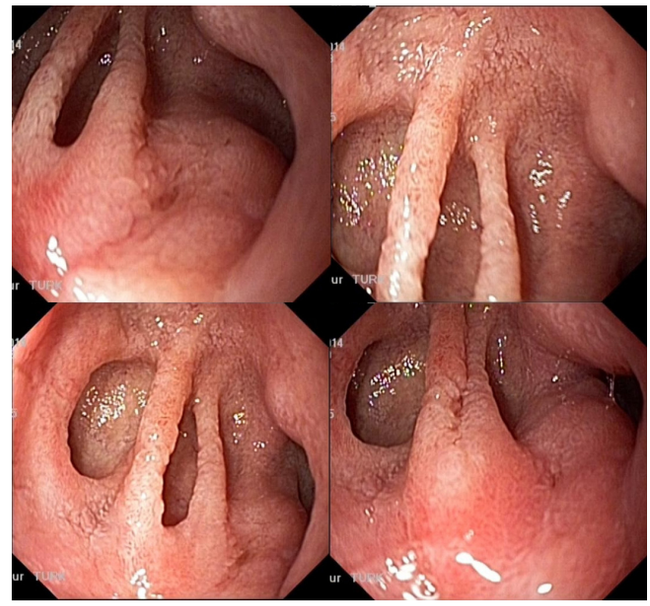

Figure 2. Endoscopic view of multiple duodenal diverticulums.

Discussion

Incidence of all small bowel diverticulosis is reported between 0,06% and1,9%. Incidence of DD is nearly 2%. More common is fifth decade of life and has a male dominancy. DD is more five times more than jejunal diverticulosis. DD can be classified according to genesis, location and structure. DD is classified as congenital an acquired diverticulum according to genesis; extraluminal duedonal diverticulum (EDD) and intraduodenal diverticulum (IDD) according to the structure. EDD are more common than IDD. EDD can be classified into periampullary diverticula (PAD) and juxtapapillary duodenal diverticula (JPDD). Most of the DD is located at the second part of the duodenum with the incidence of 80% to 90%. DD may be in various number; six diverticulum reported in literature. Commonly DD are asymptomatic. Most of the patients were admitted to the hospital with non-specific symptoms and DD diagnosed incidentally. Symptomatic patients are less than 5% of the patients DD were diagnosed. Abdominal pain or discomforts are most common

symptoms. Obstruction of the duodenum ,delayed empting of diverticulum, pressure to the bile duct, perforation of diverticulum, inflammation of diverticulum are reasons of the symptoms [4]. JPDD is suspected of cause to bile duct stones [5]. Inflammatory complications including pancreatitis, cholecystitis and cholangitis may be occurring. DD can be diagnosed during upper gastrointestinal system endoscopy alongside conventional barium radiographies. Computerized tomography and magnetic resonance imaging are advanced imaging methods to diagnose of DD [6]. These studies are also useful to diagnose of complications of DD. Surgical treatment of DD is not recommended unless it is symptomatic or complicated [7]. Neurofibromatosis could affect various organs. Along the digestive system carcinoid tumors and neuroendocrine tumors are more encountered diseases. Gastrointestinal stromal tumors associated with NF1 reported in the literature [8]. Although DD is not associated with the neuroendocrine tumors of digestive system may be associated with NF1 [9]. In this case the location of the DD and coexistence with NF1 is a rare clinical condition. In our knowledge this case will be the first reported DD in the first part of the duodenum in NF1. Although there was not any endoscopic sign of the gastrointestinal neurofibromas in this case; maybe the cause of the acquired DD is neurofibromas that are located around of the duodenum. Absence of the advanced imaging of the abdomen is missing side of the report. In this context endoscopic examinations and abdominal imaging may be useful for the diagnosis of gastrointestinal coexisting disease in NF1. In conclusion neurofibromatosis type 1 could affect multiple systems and it should evaluate carefully.

Competing interests

The authors declare that they have no competing interests.

References

1. Buluş H, Akpınar A, Coşkun A, Aydın A, Yavuz A. Giant duodenum diverticula: a case report. Kolon Rektum Hast Derg 2011;21(1):78-80.
2. Lawler LP, Lillemoe KD, Fishman EK. Multidetector row computed tomography and volume rendering of an adult duodenal intraluminal "wind sock" diverticulum. J Comput Assist Tomogr 2003;37(4):619-21.
3. Boyd KP, Korf BR, Theos A. Neurofibromatosis type 1. J Am Acad Dermatol 2009;61:1-14.
4. Mantas D, Kykalos S, Patsouras D, Kouraklis G. Small intestine diverticula: Is there anything new? World J Gastrointest Surg 2011;3(4):49-53.
5. Chen Q, Li Z, Li S, Ding X, Liu Z, Wu C, Gong J, etal. Diagnosis and treatment of juxta-ampullary duodenal diverticulum. Clin Invest Med 2010;33(5):298-303.
6. Balci NC, Akinci A, Akun E, Klor HU. Juxtapapillary diverticulum: findings on CT and MRI. Clin Imaging 2003;27(1):82-8.
7. Martinez-Cecilia D1, Arjona-Sanchez A, Gomez-Alvarez M, Torres-Tordera E, Luque-Molina A, Valenti-Azcarate V, et al. Conservative management of perforated duodenal diverticulum: A case report and review of the literature. World J Gastroenterol 2008;14:1949-51.
8. Riccardi VM. Neurofibromatosis: past, present and future. N Engl J Med 1991;324(18):1283–5.
9. Salvi PF, Lorenzon L, Caterino S, Antolino L, Antonelli MS, Balducci G. Gastrointestinal stromal tumors associated with neurofibromatosis 1: a single centre experience and systematic review of the literature including 252 cases. Int J Surg Oncol 2013;398570.

How to cite this article:
Turk O, Polat H. Togetherness of the Multiple Diverticulosis of the First Part of the Duodenum with Neurofibromatosis Type I: A Case Report. J Clin Anal Med 2014;5(suppl 2): 197-9.