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Case Report

Primary neuroectodermal tumor: a case report and review of the literature

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Abstract: The primary neuroectodermal tumor (PNET) arising in the colon is extremely rare. We reported a case of 26-year-old male colonic PNET patient whose initial symptoms were just diarrhea and paroxysmal pain. This PNET patient's journey from onset to death only lasted one and a half months, losing opportunities of surgery, radiotherapy and chemotherapy. We wish to draw the attention of our colleagues to the occurrence of PNET of colorectal origin. At the same time, long-term observation or medical follow-ups may be extremely important for a patient whose cause is not fully understood.

Keywords: Primary neuroectodermal tumor (PNET), colon, colorectum

Background

Primitive neuroectodermal tumors (PNET) are a class of rare neurogenic small round cell tumors with highly malignant potential. According to the literature, it can occur in the brain [1], spine [2], kidney [3] and so on, but rarely in the colon [4] and other colorectal tracts. The incidence of PNET of colorectal origin is extremely low. Up to now, only one case has been reported in the colon [4] and no more than 10 cases in the colorectal tracts. Owing to its rapid development, lack of effective treatment options and poor prognosis of patients, early detection is particularly critical, so is the experience of admissions doctor. In short, in this study, we reported a case of PNET patient occurring in the colon and reviewed PNET patients occurring in the colorectum for the reference to clinical workers.

Case presentation

A 26-year-old man came to our hospital for a month of diarrhea and paroxysmal pain, but no fever, chills, emesis or other positive syndromes in January 12, 2017. He said he had a contaminated diet and was diagnosed with gastroenteritis after an abdominal computed tomography (CT) scan in a French hospital because of the same symptoms in December 14, 2016 (no written material). After being detained a day in the hospital for observation, he got some oral drugs and left without symptom relief. Physical examination on admission revealed slight abdominal swelling and tenderness in the right upper quadrant of the abdomen. The results of laboratory examinations were: WBC 11.89×10^9/L, neutrophilic granulocyte 79.7%, RBC 3.77×10^{12}/L, Hb 108 g/L, PLT 506×10^9/L, CRP 51 mg/L, ALT 189 U/L, AST 108 U/L, PT 16.8 s, D-Dimer 15.29 mg/L. We rechecked CT scan which showed multiple malignant intra-abdominal masses and liver metastases (**Figure 1**). The result of liver puncture showed positive immunoreactivity for CD99 (Supplementary Figure 1) and Vim, but negative immunoreactivity for CD117, CD34, CD56, CgA, CK, S-100, SMA, Syn, EMA, Desmin and HMB45. However, because of rapid deterioration of his general condition, he was transferred to ICU in January 20, 2017 and followed by multiple organ failure. In January 29, 2017, the patients gave up the treatment and died.

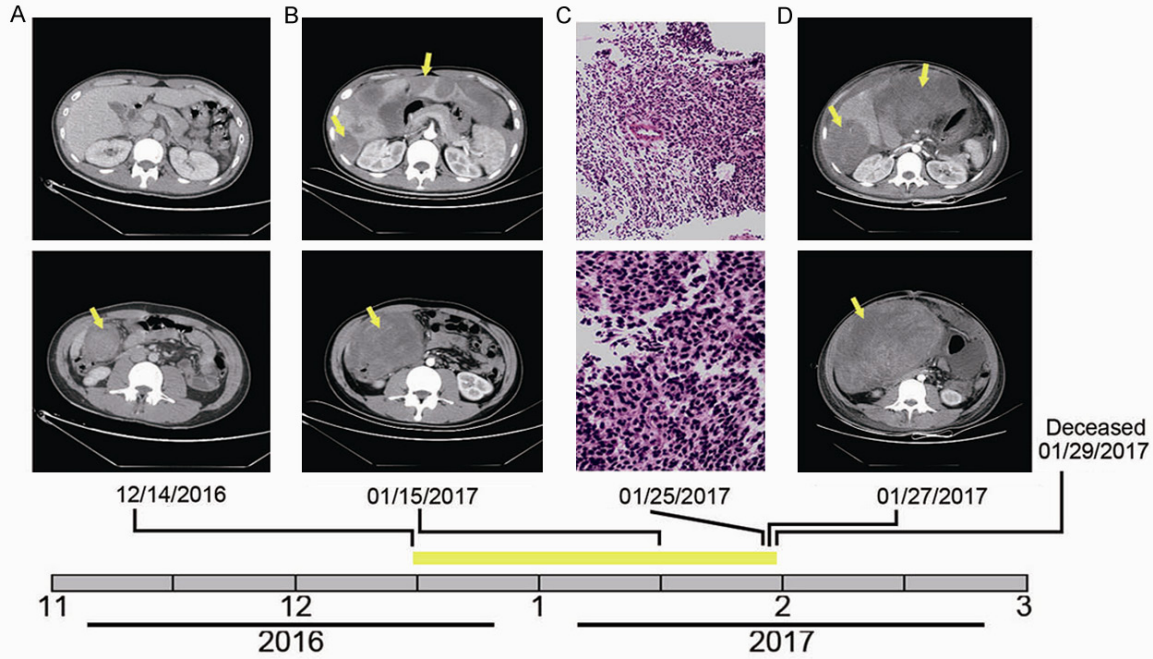

Figure 1. Time line of patient's colonic PNET diagnosis. A: Abdominal CT scan depicting an irregular mass in the colon. B: Enhanced CT revealing increase in heterogeneous mass and occurrence of liver metastases. C: HE staining of the tumor showing tumor nests (40×, magnification), at a higher (magnification of 100×). D: Enhanced CT revealing increase in heterogeneous mass and occurrence of liver metastases. Yellow rectangle near the time line represents this PNET patient's journey from onset to death.

Table 1. Clinical features of 6 colorectal PENT patients

Authors (Year)	Age (yr)/ Gender	Symptoms	Location (Size)	Positive immunomarker	Follow-up
Present case	26/M	Abdominal pain	Colon (9.9*10 cm)	CD99, Vin	1.5 mo Died
Peng *et al* (2014) [9]	36/F	Abdominal pain and Abdominal mass	Ileocecum (15*15*13 cm)	CD99, FLI1, Vim	34 mo Died
Aboumarzouk *et al* (2009) [10]	34/F	Anorectal pain and Rectal bleeding	Rectum (10 cm)	CD99	7 years DFS
Kuwabara *et al* (2006) [4]	59/M	Mass	Colon (10 cm)	CD99, CgA, MIB-1	7 mo Died
Vardy *et al* (2005) [11]	53/M	Rectal bleeding	Rectum (NM)	CD99, Vim	2 years Died
Drut *et al* (2003) [12]	17/M	Intermittent pain and Rectal bleeding	Rectum (4.5*4*4 cm)	CD99, S100	1 year DFS

NM, no mention; DFS, disease-free survival.

A written informed consent for the case report was obtained from the patient. The consent procedure was approved by the Ethics Committee of the First Affiliated Hospital of Wenzhou Medical University.

Discussion and conclusions

PNET was first reported by hart and Earle to describe a class of neuroectodermal tumors in the brain [5]. With the increase of related literatures, it was found that it can occur in the central nervous system and other peripheral organs. Therefore, it is divided into central and peripheral PNET. However, up to now, the study of PNET has been more frequently found in case reports, but less in large sample studies.

Due to lack of characteristic clinical manifestations, suggestive blood markers and imaging features, it is easy for PNET to be misdiagnosed and missed diagnosed [6]. Immunohistochemistry results show that Homer-Wright daisy-group and CD99 (+) are specific for diagnosis in the actual clinical work [7]. Surgery is the preferred treatment, but the results were unsatisfactory according to the current reports. The main reasons are as followed: 1. Patients have liver, lung and other distant metastasis, 2. Extensive resection is difficult for a wide range

of lesion. So the major treatment for PNET patients is the combination of surgery, radiotherapy, and chemotherapy [8].

Only one case of colonic PNET has been reported in English literature that the patient died for tumor recurrence in the retroperitoneal metastasis after 7 months of his first operation [4]. There are still not enough cases to summarize the clinical features of colonic PNET patients. Our report may be useful. Thus we collected data of PENT patients arising in the colorectum but did not contain mesentery (**Table 1**). It includes 2 females and 4 males, with an average age of 37.5 years old (17-59 years). The main manifestations are abdominal pain, rectal bleeding and symptoms caused by rapidly increased tumors. They are not special, so are the results of diagnostic imaging presentation. Especially the patient in this report, the initial imaging presentation and medical history is confusing, which needs to distinguish from intussusception, stromal tumor, enteritis and others, so the clinical experience is very important. Due to fast progression of disease and rejection of autopsy, we did not carry out further examination, but taking the histological results and disease history into account we still considered it is PNET.

In summary, the PNET occurring in the colorectum has the characteristic of nonspecific clinical manifestations, rapid development, poor prognosis and so on. At the same time, early imaging features and suggestive blood markers are not clear, so it is easy to be misdiagnosed and missed diagnosed. We hope that clinical workers can learn some experience from this article. For patients with rapidly increasing abdominal mass and rectal bleeding, we should take colorectal PNET into account. At the same time, long-term observation or medical follow-ups may be extremely important for a patient whose cause is not fully understood.

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Disclosure of conflict of interest

None.

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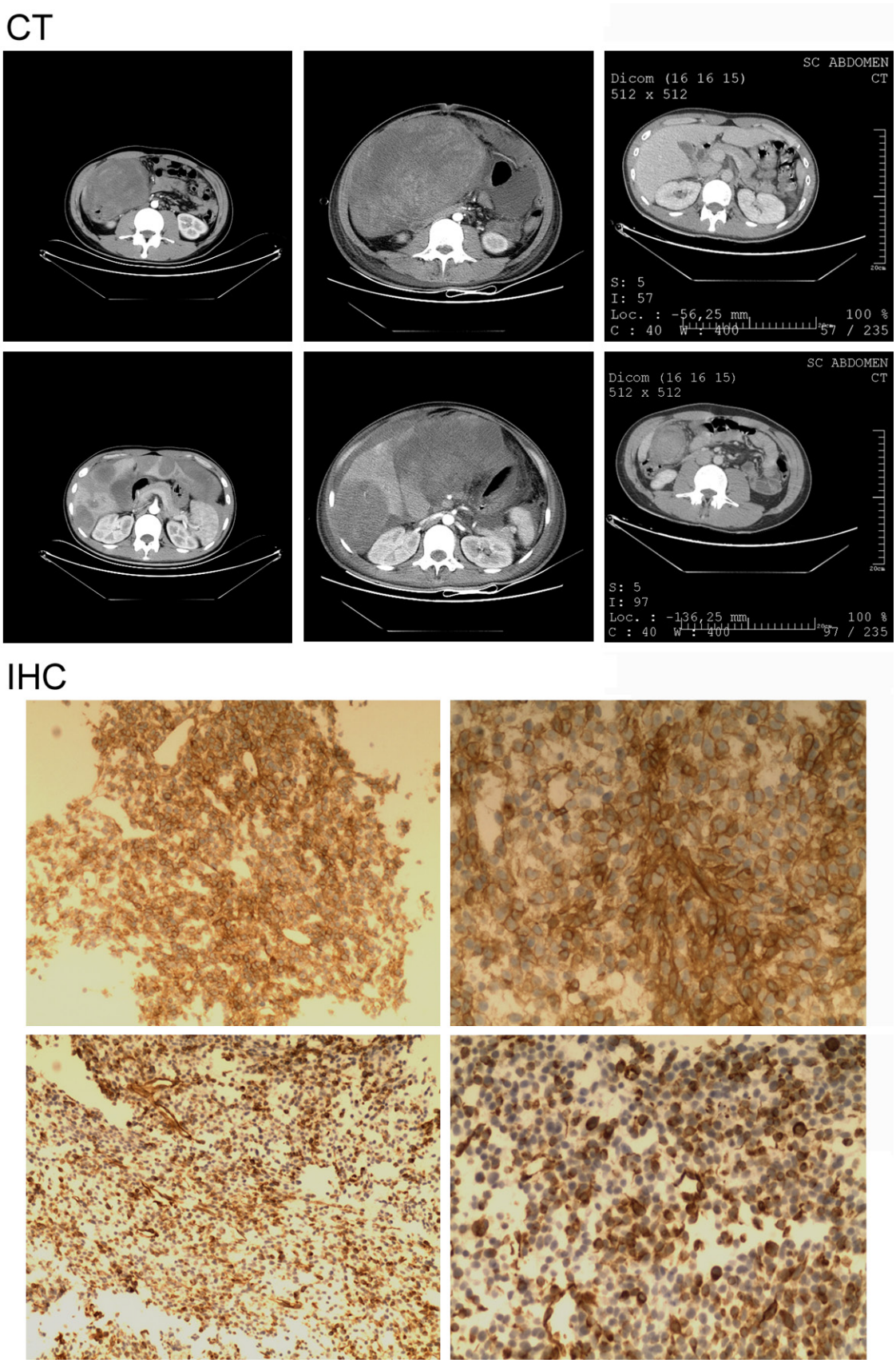

Supplementary Figure 1. CT: Original Images of CT, IHC: Strong CD99 positivity in tumor cells (40×, magnifcation), at a higher (magnifcation of 100×).