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Original Article

Three cases of familial polyposis associated mesenteric desmoid tumors: imaging features and diagnostic value

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Abstract: Desmoid tumors are a rare but life-threatening tumor in patients with familial polyposis after total colectomy. They mostly occur in the mesentery. Due to the lack of understanding of the disease and its imaging findings, they are often misdiagnosed as metastatic tumors and suffered further treatment. But conventional gastrointestinal tumor adjuvant chemotherapy is not effective for desmoid tumors. Surgical resection will stimulate rapid tumor growth and complications, so correct diagnosis has an important clinical significance. This study comparatively analyzed the clinical, pathological, and imaging features of three patients with familial polyposis-related desmoid tumors. The diagnosis and treatment of the lesions on the outcome and the related imaging signs were reported and evaluated. We found that the tumor image characteristics can be divided into two types: type I was common, and it consisted of multiple irregular masses or flakes with blurred margin, which grew around the mesenteric blood vessels and showed extreme hypointensity at the center on the T2-weighted image and slight hyperintensity at the periphery; Type II was rare, and it showed an isolated round-like mass with distinct margin and "black dot and line sign". All three patients progressed after biopsy, and two of them were complicated with intestinal fistula and intratumoral abscess. The intra-tumoral abscess manifested an irregular cavity with a thick wall. Lesions with hyperintensity on the T2-weighted and diffusion-weighted images at periphery progressed rapidly. Therefore, the particular imaging manifestations of familial polyposis-associated desmoid tumors have important value in the diagnosis and the selection of clinical treatment strategies.

Keywords: Desmoid tumors, familial polyposis, X-ray computed tomography, magnetic resonance imaging

Introduction

Desmoid tumor (DT), also known as aggressive fibromatosis, is a rare benign tumor of fibrous link tissue but has the characteristics of invasive growth and easy to reoccur without metastasis [1, 2]. Most DTs are sporadic, often occurring in the abdominal wall or limbs, and often solitary; about 5-15% of DTs are complicated by familial adenomatous polyposis (FAP). DTs combined with FAP often occurred after FAP total colectomy, and they were almost multiple and about 80% occurred in the mesentery [2, 3]. Since it often surrounds mesenteric blood vessels and intestinal wall, resection of the lesion is difficult, and the recurrence rate is high. Moreover, large tumors may complicate with intestinal obstruction, perforation, intratumoral abscesses, and bleeding, which are difficult to treat. They are important causes of death after FAP total colectomy [4]. Therefore, the early detection and correctdiagnosis of the DTs play an important role in the choice of treatment and prognosis. Imaging is the preferred alternative to detecting and monitoring DT.

Summarizing the imaging characteristics of FAP-related DT can provide a reference for the selection of clinical treatment strategies. Currently, a few studies have reported the radiologic manifestations of DT, but most of them were about the lesion at the superficial parts or sporadic DT [5, 6], few were about FAP-related DT [7, 8]. Therefore, we reported 3 cases of mesenteric DT related to FAP about their clinical diagnosis and treatment process and image characteristics.

Case report

Clinical data

The data of three patients with familial polyposis who underwent regular imaging follow-up

Table 1. Basic information, diagnosis and treatment of DTs

	Case 1	Case 2	Case 3
Basic information			
Sex	Female	Male	Male
Age (years)	34	28	31
History	FAP	FAP with RC	FAP with CC
Initial diagnosis and treatment of DT			
Time of tumor detection (after total colectomy)	10 months	24 months	15 months
CT diagnosis	Tumor	Metastasis	Metastasis
Clinical diagnosis	FAP-related DT	Metastasis	Metastasis
Pharmacotherapy	-	Chemotherapy and targeted therapy	Chemotherapy
Follow-up after pharmacotherapy	-	CT (enlarged slowly)	CT and MRI (enlarged rapidly)
DT surgery			
Type	Laparotomy and tumor biopsy	Laparotomy and tumor biopsy	Laparotomy and partial tumor resection
Time of surgery (after tumor detection)	19 days	25 months	9 months
Postoperative therapy	Tamoxifen and celecoxib	-	-
Follow-up			
Time of last follow-up time (after laparotomy)	29 months	52 months	60 months
Tumor evaluation	Progression (tumor enlarged)	Progression (tumor enlarged and new lesions appeared)	Progression (tumor enlarged and new lesions appeared)
Complications			
Type	Tumor-duodenal fistula and intratumoral abscess	Tumor-ileum fistula and intratumoral abscess	-
Time of appearance	1.5 months after biopsy	8 months after biopsy	
Symptoms	Recurrent fever and abdominal pain	Recurrent abdominal pain and occasional fever	-
Treatment	Anti-infection	Anti-infection	-

Note: DT, desmoid tumor; CT, computed tomography; MRI, magnetic resonance imaging; FAP, familial adenomatous polyposis; RC, rectal carcinoma; CC, colon carcinoma.

after total colectomy in our hospital from January 2010 to December 2019 were retrospectively analyzed. They all received surgical-resection or biopsy to confirm the diagnosis of DT. Two patients were male, and one was female, with an average age of 31 years. Two patients were diagnosed of FAP with colorectal cancer. The mesenteric lesions were found during CT follow-up after FAP total colectomy. One patient was diagnosed as tumor, and the other two were diagnosed as metastatic tumors preoperatively by CT or MRI. The CEAf levels of all three patients were in normal range. The tumor in case 3 progressed rapidly in the early postoperative period and multiple new lesions appeared, however the lesions progressed slowly in the later period; the other two patients had tumor progression after the operation with tumor-small bowel fistula and intra-tumoral abscess, and repeated sepsis (**Table 1**), then they were hospitalized. Following up to December 2019, all three patients were surviving. This case report has obtained the consent of all patients.

Surgery and pathology

Cases 1 and 2 underwent laparotomy with tumor biopsy, and case 3 underwent laparotomy with tumor biopsy and partial tumor resection. All three cases had multiple lesions, which palpated hard and involved the mesentery. Most of them had invasive margin and unclear border, and some of them were adherent to intestinal walls. Only one lesion had a clear border which was removed in case 3, about 10 × 15 cm (**Figure 2A-D** shown by white arrows on MR images). Its general section was woven and massive wavy fibers with glassy degeneration and interspersed mucous degeneration and scatter spindle cells distributed uniformly in the HE staining. However, HE staining of the biopsy specimens from three cases showed massive spindle cells and wavy fibers with glassy degeneration and interspersed mucous degeneration. Denser fibers distributed around blood vessels, while the spindle cells and mucous degeneration predominated at distant area from the vessels. Immunohistochemistry results of different degrees of positive nuclear

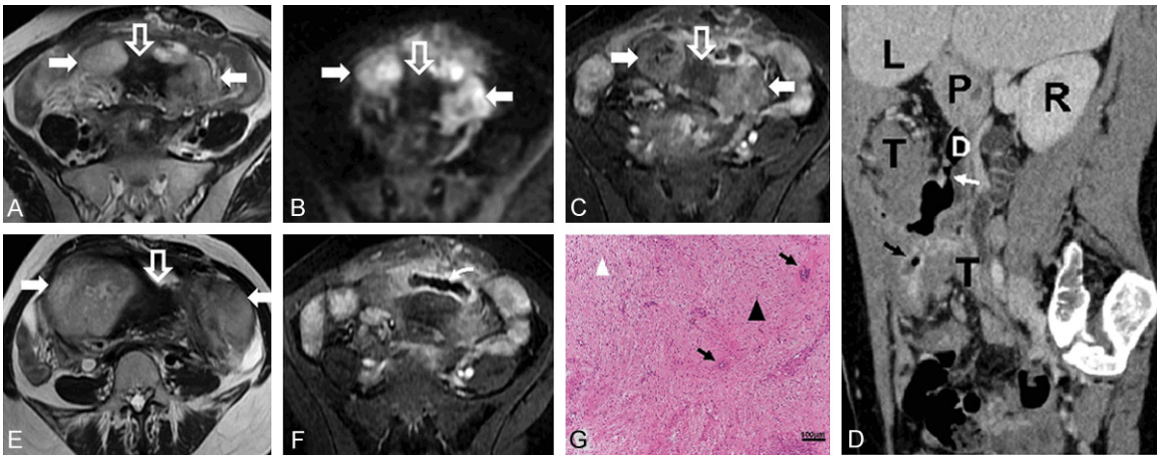

Figure 1. CT, MRI and pathological characteristics of a mesenteric DT with intestinal fistula and intratumoral abscess in case 1. A-D. MRI scans 25 months after the diagnosis of DT. A. Axial T2WI showing a large irregular mass in the mesentery with homogeneous extreme low-signal intensity in the center (empty arrow) and high-signal intensity in the periphery (white arrows; type I). B. Axial diffusion-WI showing low-signal intensity in the center (empty arrow) and heterogeneous high-signal intensity in the periphery (white arrows). C. Axial contrast-enhanced T1WI during the delayed phase showing mild enhancement in the center and heterogeneous moderate enhancement in the periphery. D. Oblique sagittal CT scan showing an irregular pus-filled cavity in the mesenteric DT (T), marked enhancement of the thickened wall (black arrow) and perforation on the horizontal section of the duodenum (white arrow). E, F. MRI scans 32 months after the diagnosis of DT. E. Axial T2WI showing that the size of the central low-signal area remained the same (white arrow), but the surrounding high-signal area was significantly enlarged (empty arrow). F. Axial contrast-enhanced T1WI during the delayed phase showing a pus-filled cavity with wall thickening (curved arrow) within the tumor. G. Pathological HE staining (original magnification × 100) showing a flaky fibrous hyaline degeneration area (black arrowhead) in the central area, and more obvious fibrous hyperplasia around the blood vessel with hyaloid degeneration (black arrow). The spindle tumor cells were more densely distributed in the peripheral area (white arrowhead). L, liver, P, pancreas, R, kidney, D, duodenum; CT, computed tomography; MRI, magnetic resonance imaging; T2WI, T2-weighted image; DT, desmoid tumor.

β-catenin staining in 3 cases supported the diagnosis of DT.

Image findings

There were two types of lesions observed in our cases. Type I lesion manifested as irregular mass or flake with blurred margin, and they were all multiple in all 3 cases, most of them were fused while enlarged. Hence, it was different to describe the size and count them. They grew around the mesenteric vessels and caused compression and occlusion of the internal vein. At MR imaging, the central area of the lesion showed extreme hypointensity on T2 weighted image and mild enhancement on contrast delay phase, while the peripheral area showed hyperintensity on T2 weighted image, restricted diffusion and markedly enhanced on delay phase (**Figures 1** and **2**). On the follow-up MR images in case 1, the lesions enlarged markedly, especially at the peripheral zone (**Figure 1E**).

There was an oval mass with a distinct margin observed in case 3 (**Figure 3A-D**), which was defined as type II. On T2 weighted image, it showed multiple parallel low signal intensity bands accompanied by high signal intensity bands among it, which was defined as “black dot and line sign” by us, and the lesion enhanced heterogeneously (**Figure 3A**, thin white arrow). Besides, a hyperintensity ring was observed around the lesion on T2WI and DWI, and it showed marked enhancement (**Figure 3A**, **3C** and **3D**, thick white arrow). The density of both types of lesions on CT images and the signal intensity on the T1 weighted images were equal or slightly lower than those of muscle. Tumor-small intestine fistula and intra-tumoral abscess occurred in 2 cases during the course of the disease, which manifested as a gas containing cavity with a markedly enhanced thickening wall (**Figure 2**). The contrast enhanced images showed that some intestinal walls were invaded by the tumors and partially penetrated.

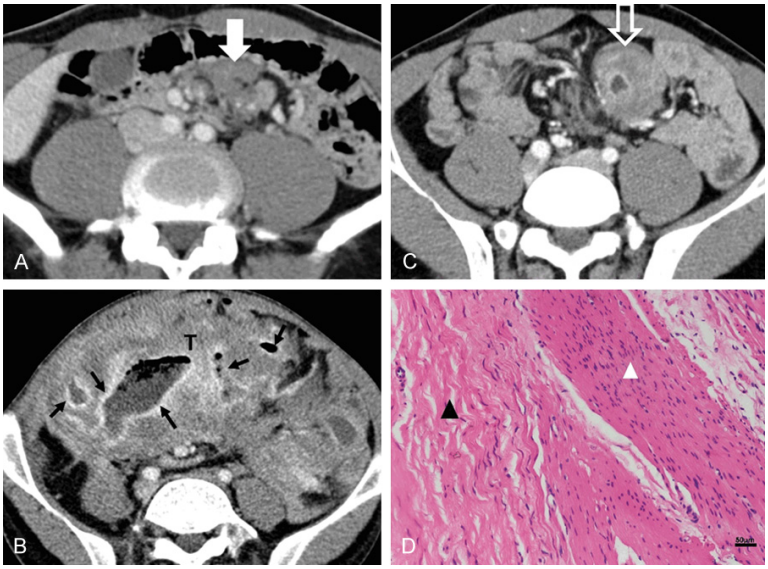

Figure 2. CT, MRI and pathological characteristics of mesenteric DT in case 2. A. Twenty-four months after total colectomy, CT scan revealed multiple mesenteric masses surrounding mesenteric vessels (white arrow). B. Three months after the diagnosis of mesenteric DT by biopsy, CT scan showed that mesenteric lesions were slowly enlarged with intratumoral abscess formation (dark arrows). C. Thirty-three months after the diagnosis of DT, the mesenteric mass was significantly enlarged, accompanied by the formation of abscess in tumor-ileal fistula and intratumor, and the abscess showed significantly enhanced thick-walled cystic space (empty arrow). D. HE staining (200 ×) showed that wavy collagen fibers (black triangle) were alternated with fibrocyte-rich areas (white triangle). CT, computed tomography; MRI, magnetic resonance imaging; T2WI, T2-weighted image; DT, desmoid tumor.

Discussion

The incidence of DT in FAP patients is 800-1,000 times of that in the general population [9]. The morbidity in females and males is about 2:1, and the onset age is mostly 25-35 years. Most of them were diagnosed occasionally in the follow-up within two years after total colectomy (about 65-83%). It was found that about 80% occurred in the mesentery, which could be alone or combined with abdominal wall DT [10]. All three cases of mesenteric DT in this study were initially detected within 1-2 years after FAP total colectomy. It was misdiagnosed as metastasis in the patient of FAP with colorectal carcinoma frequently, because it often presented multiple lesions and enlarged gradually by follow-up, just as two of our cases. But it was not accompanied by increase of CEA level. The occurrence of FAP-related DT is thought to be related to the germline and system mutations of FAP's adenomatous polyposis coli (APC) gene [11], and surgical trauma is a potential initiating factor for DT. Adelaide MC et al. [12] reported that during wound healing after surgical injury, mesenteric mesenchymal cells have mutations in their downstream gene system based on APC germline mutations such as lack of transcription repressor expression, and overexpression of the β-catenin protein, and those factors could cause excessive cell proliferation, and promote the occurrence of DT and postoperative recurrence.

Image manifestations and pathological comparisons

Mesentery DTs are generally pathologically characterized as grayish-white or grayish-yellow soft tissue mass or flat plaque. In our cases, most of the lesions showed infiltrative growth and had an unclear border at laparotomy, which corresponds to the type I lesions. In addition, some small lesions in our study showed an obscure feathery margin (flame sign) in one aspect on cross sectional images as the perverse study reported [13]. Only one lesion had an oval shape and a distinct margin, which coexisted with type I lesions, and Louisa Azizi et al. [7] also reported this phenomenon. Patients with FAP related DTs often have multiple lesions, as we reported in all three cases, while sporadic DTs were almost solitary mass, although they both share the same pathological manifestation [14].

On CT images, soft tissue density of DT can be found in the early stage under the background of the low density of mesenteric fat, so CT scan is an optimal option for detection and surveillance of DT. Although the detection rate of T1WI was found similar to CT imaging [8], CT was applied regularly in the follow-up of patients with FAP rather than MRI. We subsumed the manifestations of mesentery DTs into two types corresponding to the general pathology. Type I was more common and special, which showed in all three cases. They were in isodensity on

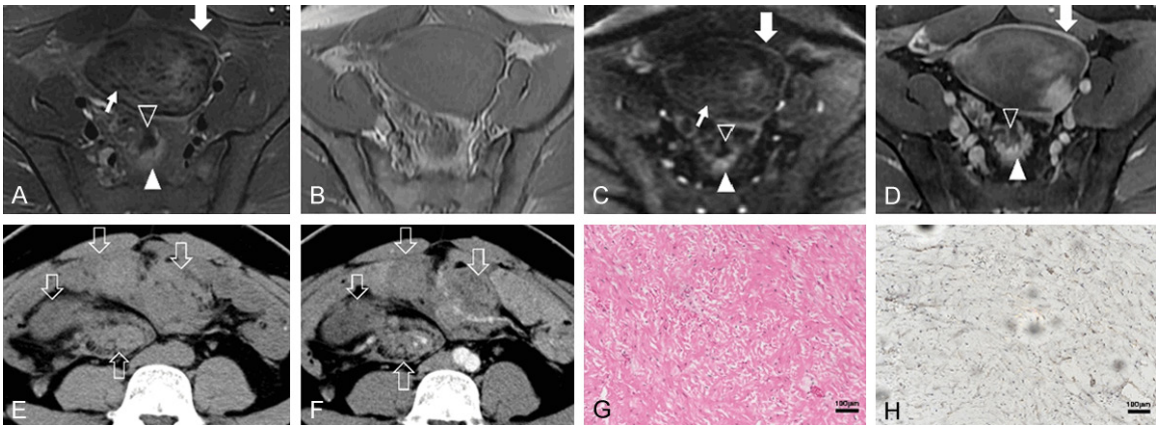

Figure 3. CT, MRI and pathological characteristics of mesenteric DT in case 3. A-D. MRI scans prior to DT biopsy. A. Axial T2WI showing a well-defined round mass with 'black dot and line sign' inside (thin white arrow) and a hyperintensity ring outside (thick white arrow), in the front of the mesentery (type II), which was then resected. An ill-defined flake surrounding a mesenteric vessel located in the back of the mesentery was also observed, which appeared dark in the center (empty arrowhead) and exhibited high-signal intensity in the periphery (white arrowhead; type I). B. On axial T1WI, both lesions showed a similar signal intensity to that of muscle. C. On the diffusion-WI, the well-defined oval mass showed heterogeneous hypointensity inside (thin white arrow) and a hyperintensity ring outside (thick white arrow). The ill-defined lesion appeared dark in the center (empty arrowhead) and exhibited high-signal intensity in the periphery (white arrowhead). D. T1W1-enhanced image during the delayed phase showing heterogeneous enhancement inside (thin white arrow) and marked ring enhancement (thick white arrow) outside the well-defined lesion, as well as a slight enhancement in the center (empty arrowhead) and marked enhancement in the periphery (white arrowhead) of the ill-defined lesion. E. Non-enhanced CT scan showing that the lesions had enlarged and increased 9 months after surgery. F. Contrast-enhanced CT showing the mesenteric vessels encased in the center of the lesion. G. HE staining (100 ×) showing large wavy fiber hyaline degeneration with mucous degeneration. H. Positive nuclear β-catenin by immunohistochemistry (100 ×). CT, computed tomography; MRI, magnetic resonance imaging; T2WI, T2-weighted image; DT, desmoid tumor.

non-contrasted CT images and persistently enhanced on contrasted CT images related to the containing of numerous fibral tissues without central necrosis, and it surrounded the mesenteric vessels with stenosis or occlusion of the encased veins on contrasted enhanced CT images. It may confuse with retractile mesenteritis, but calcification within the latter could distinguish them [15]. The type II lesion was rare and non-special as an oval isodensity mass with distinct margin and heterogeneous mild enhancement on CT images, and it just grew next to the mesenteric vessel. It may be misdiagnosed as Schwannoma or sarcoma on CT images.

However, MRI multi-parameter imaging is superior to CT imaging to differential diagnosis because it can reflect the composition of tumor tissue components to a certain extent. Extreme low signal intensity on T2WI with persistent mild enhancement represented the dense fibers, which was a mark of fiber-origin tumors. For type II lesion, the multiple dark dots and bands on T2WI ("black dot and line sign") indicated the diagnosis of fiber-origin tumors rather than metastasis tumor or Schwannoma. Although some other fiber-origin tumors such as myxofibrosarcoma and giant cell tumor of tendon sheath still cannot be excluded, the diagnosis of DT would also be considered firstly by combination of the history of FAP.

However, for type I lesions, the abundant homogeneous extremely low signal intensity demonstrated around the mesenteric vessels in the center on T2WI with non to mild degree of enhancement was more special. That was almost not observed in metastasis tumors, lymphoma, and sarcomas in mesentery, while mild enhancement of DT in anterior phase was different from the other fiber containing tumors, such as solitary fibrous tumors and carcinoid tumors, but it is difficult to differentiate between the DTs and retractile mesenteritis. The mesenteric vessels in the tumor were compressed severely by the dense fibers. The low signal intensity area showed wider in type I lesions than that in type II lesions. We supposed that was related to the surrounding growth of blood vessels of type I lesions since

the pathological observation of more fibrous hyperplasia around the blood vessel than distant areas in our study. The peripheral zone of both two types of lesions showed hyperintensity on T2WI and DWI and marked enhancement, which represented the containing of rich tumor cells. And the rapid growth of this part after two years of follow-up in case 1 confirmed it. Therefore, previous studies have also suggested that MRI can make certain predictions on the biological behavior of tumors [16, 17], which can guide the selection of non-steroidal anti-inflammatory drugs and estrogen modulators and cytotoxic chemotherapy drugs in non-surgical treatment. Reduced tumor volume, decreased density and enhancement, reduced signal intensity in hyperintensity zone on T2WI or DWI suggested that the treatment was effective [13].

Common complications of mesenteric DT include bleeding, intestinal obstruction, complete parenteral nutrition, ureteral stricture, intestinal perforation, intestinal fistula, intra-tumoral abscess, and so on. Intra-tumoral abscesses usually occurred in large tumors with intestinal involved or after invasive operations. The cause was inferred to intestinal ischemia or bacterial translocation caused by mesenteric blood vessel involvement or surgical operation, or bacterial moved to the tumor along the lymph and vein drainage, or intestinal fistula and ureteral fistula [18]. In our study, two patients had intestinal fistulas and intra-tumoral abscesses after tumor biopsy. The corresponding imaging manifestations of the two patients in our study were similar to those reported in the literature [19]. Abscesses are irregular cavities in the tumor. After the contrast material was administrated, they showed avidly persistent enhancement of the thickening cavity wall composed of inflammation and granulation tissue. They can be distinct from the wall-less central necrosis due to ischemia of the malignant tumors such as fibrosarcoma and malignant fibrous histiocytoma. The treatment and management of intra-tumoral abscesses were difficult and did not reach consensus. A few case-reports described that if the unresectable tumors were complicated with intra-tumoral abscesses but without intestinal fistula, draining the pus and antibiotic lavage combined with drug treatment was effective. However, for patients with intestinal fistula, further surgery was needed to remove the fistula and part of the tumor [19, 20]. Hence, early detection of the complications by CT or MR imaging may change the therapy strategies and improve prognosis.

The importance of image inspection

For the diagnosis of FAP-related DT, the current V2.2019 NCCN guidelines still recommend biopsy [21], and laparoscopic tumor biopsy is often required due to the deep location of mesenteric DT. However, surgical trauma probably promotes the growth of DT. All three of our patients experienced rapid tumor growth in a short period after surgical resection or biopsy. One of them had a duodenal-tumor fistula after the operation, secondary intra-tumoral abscess formation, and recurrent sepsis. The R0 resection rate of FAP-related mesenteric DT was only about 23%. However, R0 resection still did not reduce postoperative recurrence [22], and the postoperative recurrence rate was as high as 44% [23]. Furthermore, radical surgery does not improve the overall survival of the patient, and it can be followed by a series of serious complications, such as intestinal ischemia, short bowel syndrome, severe lower limb lymphedema, and even death [24]. Therefore, in recent years, the diagnosis and treatment of FAP-related DT have tended to non-surgical methods [25]. Both tumor biopsy and radical surgery should be selected with caution. Surgical treatment is only recommended when DT has serious complications, and no radical resection of the tumor is required [24]. Therefore, CT and MRI as a non-invasive way play a crucial role in the diagnosis and evaluation of the FAP-related DT.

In summary, the FAP related DT showed characteristics on CT and MR images, especially on MR images, which could help the radiologists to make a correct diagnosis when they were familiar with those image features. For cases with a clear history of FAP total colectomy, radiologists and clinicians may diagnose and test typical cases only based on the above-mentioned characteristic imaging manifestations of FAP-related DT, thereby avoiding the possible disadvantages of invasive diagnosis and treatment.

Disclosure of conflict of interest

None.

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