



胃肠道间质瘤的误诊因素分析及鉴别

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【摘要】目的:分析胃肠道间质瘤(GIST)的影像误诊因素及鉴别要点。**方法:**搜集术前误诊为GIST患者11例,GIST患者术前未正确诊断2例作为此次研究的对象,对所有患者的影像资料进行回顾分析。13例患者均有手术病理,进行MSCT扫描,观察CT影像特征,分析误诊或未正确诊断GIST的影像学因素及鉴别价值。**结果:**13例误诊患者中,2例为腹膜后神经鞘瘤,2例为低分化腺癌,2例为异位胰腺,1例为类癌,1例为平滑肌瘤,1例为未分化肉瘤,2例为转移性癌,另外2例小肠间质瘤术前未正确诊断。误诊与肿瘤发生部位、大小、血供来源、生长趋势、观察方法等因素有关。**结论:**GIST有一定的影像特点,降低胃肠道间质瘤的误诊,三维重建-肿块准确定位是重中之重,其次应仔细观察肿块细微关键征象,注意肿块血供来源及生长趋势也是鉴别要点。

【关键词】胃肠道;间质瘤;影像诊断;鉴别诊断

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Misdiagnosis factors and differential diagnosis of gastrointestinal stromal tumors

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Abstract: Objective To analyze factors causing misdiagnosis of gastrointestinal stromal tumors (GIST) and provide some key points for the differential diagnosis of GIST. Methods The imaging data of 11 patients who were misdiagnosed as GIST and 2 patients with GIST who were misdiagnosed before operation in Guangzhou Red Cross Hospital from April 2014 to May 2017 were retrospectively analyzed. All 13 cases were confirmed by operation and pathology and underwent MSCT scanning. We observed the features of CT imaging, analyzed the imaging factors affecting the diagnosis of GIST and evaluated their values in differential diagnosis. Results The pathological diagnosis results of 13 patients who were all misdiagnosed included retroperitoneal schwannoma (2/13), poorly differentiated adenocarcinoma (2/13), ectopic pancreas (2/13), gastrointestinal carcinoid (1/13), leiomyoma (1/13), undifferentiated sarcoma (1/13), metastatic carcinoma (2/13) and small intestinal stromal tumor (2/13). Misdiagnosis was related to tumor location, size, source of blood supply, growth trend, observation method and so on. Conclusion GIST has some certain imaging features. To reduce misdiagnosis rate, accurate localization of mass in three-dimensional reconstruction is the priority among priorities; the observation of subtle but key signs about mass are also important; and the source of blood supply and growth trend of mass should also be considered in the differential diagnosis of GIST.

Keywords: gastrointestinal tract; stromal tumors; imaging diagnosis; differential diagnosis

前言

胃肠道间质瘤(Gastrointestinal Stromal Tumors, GIST)是一种起源于cajal细胞的间叶组织源性肿瘤,具有多向分化性及恶性的转化潜能^[1],以前多与纤维

瘤、纤维肉瘤、神经鞘瘤等间叶组织来源的肿瘤混淆。随着免疫组化的发展,逐渐发现GIST是CD117、CD34大部分特异性阳性的肿瘤,从而与其他间叶组织来源的肿瘤区分开来。在MSCT诊断、随访过程中,一些病例往往误诊,为此笔者共收集13例经手术病理证实术前误诊的病例进行回顾性分析误诊因素,以期提高本病的诊断及鉴别水平。

1 材料与方法

1.1 一般资料

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红十字会医院13例手术病理病例,其中11例术前误诊为GIST,术后经病理证实为非GIST;2例小肠远端间质瘤术前误诊为附件粘液腺囊性癌及卵巢癌。所有病例均行3期增强扫描。其中男4例,女9例;最小年龄33岁,最大年龄83岁,平均年龄56.7岁。13例患者主要临床表现腹痛、腹胀、自感腹部包块,2例患者排黑便并1例同时有呕吐咖啡样物,2例腹部B超体检未检出病灶。

1.2 仪器与方法

采用Philips Brilliance 64排128层螺旋CT行3期动态增强扫描。扫描参数:120 kV, 250 mA, 层厚3 mm, 层距1.5 mm, FOV 512 mm×512 mm。均有肘静脉团注非离子型对比剂碘海醇,剂量2 mL/kg,注射速度3.0 mL/s。

1.3 图像分析

由两位副高以上高年资放射科医师分析图像,着重分析肿块生长部位、生长方式、最大径、边界清晰度、强化程度、强化方式、供血血管及有无其他部位转移。根据需要,部分病例加做冠状位、矢状位重建。

2 结果

2.1 误诊因素

术前11例误诊为GIST病人中,2例为腹膜后神经鞘瘤、2例为低分化腺癌、2例为异位胰腺、1例为类癌、1例为平滑肌瘤、1例为未分化肉瘤、1例为转移性鳞癌、1例为网膜囊淋巴结转移性低分化腺癌伴粘液癌。另外2例术后病理证实为小肠远段间质瘤。13例患者共14个肿块。发生部位因素:发生在胃4例、发生在小肠4例、3例在肠系膜、3例发生在腹腔及腹膜后;大小因素:肿块长径大于5 cm的共有9例;生长方式因素:绝大多数肿块呈类圆形或椭圆形生长,仅2例呈不规则或分叶状生长;强化因素,增强扫描可见10例不均匀强化,可见不同程度坏死,4例表现为均匀强化;周围淋巴结因素:肿块绝大部分周围没有肿大淋巴结,仅有2例患者周围可见数个淋巴结,具体影像表现及术后定位及病理可见表1。下面将仔细分析图1~4 4个病例中平时容易忽略的关键征象及三维重建定位的重要性。其中1例术后证实为腹膜后神经鞘瘤(图1),增强扫描动脉期可见中腹部一不均匀轻-中度强化肿块,其内包埋肠系膜动脉分支血管;1例术后证实为转移性鳞癌(图2),增强扫描动脉期可见胃底大弯侧壁一边界清楚、强化不均匀肿块,其内可见坏死,与胃壁关系密切,轴位看似与胃壁分界不清;2例异位胰腺肿块较小均不超过2 cm,1例表现为增强扫描门脉期左上腹空肠内(图3),见

一密度均匀、强化亦明显均匀强化的小圆形结节,表现为明显持续均匀强化;2例术后证实为盆腔小肠巨大间质瘤(图4),表现为边界清晰的以囊性成分为主的囊实性占位,增强扫描囊壁及少许实质明显强化、大部分囊性部分未见强化、囊壁厚薄不均并可见小壁结节;1例低分化腺癌肿块表现为胃肠道壁不规则肿块,增强扫描不均匀轻-中度强化,肿块邻近脂肪间隙模糊,周围可见多发肿大淋巴结。

2.2 误诊病理分类

在13例误诊病人中,2例神经鞘瘤均经术后切除病理证实(图5)。图5a为其中1例大体手术切除实体图,图5b为病理镜下所见:肿瘤组织由成纤维细胞样梭形细胞和泡沫样组织细胞及慢性炎性细胞构成,梭形细胞部分呈编织状,部分细胞核肥大,核分裂像极少见;免疫组织化学:Nestin(+),CD117(-),CD34(-),SMA(-),HHF35(-),CD68(-),CR(-),P53(-),S100(+),Ki67(1%)。2例转移癌均经病理证实,三维重建图和病理图见图6。图6c为转移癌病理镜下病理所见:送检穿刺组织内见异型细胞呈巢团状,片状排列,细胞稍大,胞浆红染,核大深染,可见核仁;部分区域可见角化珠形成,间质纤维组织增生并少量慢性炎症细胞浸润;免疫组织化学:CK5/6(+),CK7(灶状+),CK20(-),Villin(-),CDX-2(+),Vimentin(+),CA199(-),CgA(-),CEA(灶状+),NSE(-),P53(80%+),Ki67(60%+)。2例异位胰腺均经手术病理证实(图7),图7a为其中1例大体手术切除实体图,其中可见胰腺泡、导管及胰岛,图7b为病理镜下可见正常胰腺组织结构,未见任何异型细胞。2例误诊为附件恶性肿瘤术后病理证实为末端小肠间质瘤(图8),图8a为小肠间质瘤术后实体图,图8b为小肠间质瘤镜下病理所见:肿瘤组织由梭形细胞构成,与肠壁平滑肌组织分界不清,无包膜分隔,细胞核长梭形,核分裂象<5个/50HPF,肠粘膜及浆膜未见浸润,局部见小血管血栓形成、间质出血、缺血坏死;免疫组化结果:Dog-1(+),CD117(+),CD34(+),SMA(-),Desmin(-),S-100(-),Ki67(1%),P53(1%+)。1例平滑肌瘤患者行免疫组化分析CD117及CD34均为阴性。其余大部分肿块可见坏死,其内可见不同比例肿瘤细胞分布,异型性明显,部分浸润胃肠壁达浆膜,2例可见淋巴结转移。

3 讨论

3.1 临床与病理

GIST是由Mazur等^[2]在1983年依据肿瘤免疫组化特征提出的,作为消化道最常见的间叶源性肿瘤,组织学上有上皮细胞、梭形细胞及散发的多形性细胞弥漫或束状排列而成,通常表达CD117、CD34阳性的特异



表1 肿块影像表现及术后定位及病理结果

Tab.1 Imaging features, postoperative localization and pathological results of mass

Postoperative site	Size/cm ²	Enhancement	Lymph node	Others	Preoperative diagnosis	Postoperative pathological results
Retroperitoneal	11.0×7.0	Inhomogeneous	No	Mass-embedded mesenteric vessels	Small intestinal stromal tumor	Schwannoma
Retroperitoneal	5.7×4.9	Inhomogeneous	No	Clear regular boundary	Small intestinal stromal tumor	Schwannoma
Stomach	1.0×1.0	Homogeneous	No	Clear regular boundary	Gastric stromal tumor	Ectopic pancreas
Jejunum	2.0×1.4	Homogeneous	No	Clear regular boundary	Stromal tumor or adenoma	Ectopic pancreas
Jejunum	4.2×5.4	Inhomogeneous	Many	Mass communicates with the intestine	Stromal tumor or metastatic tumor	Poorly differentiated adenocarcinoma
Stomach	10.5×9.6	Inhomogeneous	No	Clear regular boundary	Stromal tumor	Poorly differentiated adenocarcinoma
Mesentery	3.3×2.7	Obvious homogeneous	No	Clear regular boundary	Ascending colon stromal tumor	Leiomyoma
Stomach	3.4×4.6	Obvious homogeneous	No	Good mobility	Stromal tumor or adenoma	Carcinoid
Mesentery	Two masses; 4.6×3.7, 8.2×6.7	Inhomogeneous	Many	Masses break through the mucosa	Malignant stromal tumor	Undifferentiated sarcoma
Abdominal cavity	7.3×6.5	Obvious inhomogeneous	No	Unclear boundary	Subserosal stromal tumor	Metastatic squamous cell carcinoma
Stomach	6.6×4.9	Inhomogeneous	No	Unclear boundary, close with lesser curvature stomach	Exogenous stromal tumors	Metastatic adenocarcinoma
Small intestine	13.5×9.3	Obvious inhomogeneous	No	Obvious enhancement of the wall	Mucinous cystadenocarcinoma	Stromal tumor
Small intestine	11.0×8.0	Inhomogeneous	No	Irregular strip reinforcement	Ovarian cancer	Malignant stromal tumor



图1 腹膜后神经鞘瘤动脉期扫描

Fig.1 Arterial phase scan of retroperitoneal schwannoma



图2 胃大弯转移癌动脉期扫描

Fig.2 Arterial phase scan of the metastatic carcinoma of the greater curvature of the stomach

性免疫表型。GIST可以在食管到肛门任何位置发生^[3-5]。目前GIST最终确诊手段依然是免疫组织化学。GIST好发于中老年人，男女发病无明显异常。早期临床表现缺乏典型症状及体征，2 cm以内的间质瘤一般无临床症状，大于3 cm的肿块因其表面附有粘膜，当粘膜发

生溃疡时就会出现胃肠道出血症状^[6]。一般临床症状包括长期反复腹部隐痛、自觉触及腹部包块、排柏油样大便、恶心、呕吐、贫血等，亦有部分是由体检发现入院。GIST可发生在消化道任何部位，其中约50%~60%发生在胃、约20%~30%发生在小肠、约10%发生在结肠、约

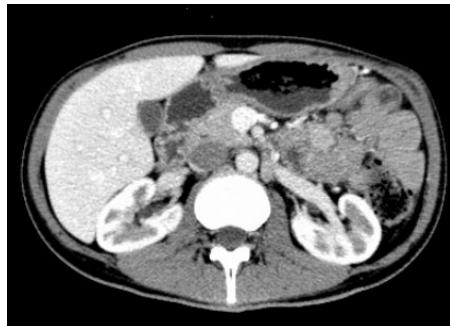
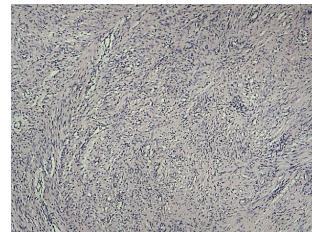


图3 空肠异位胰腺门脉期扫描

Fig.3 Portal phase scan of jejunal ectopic pancreas



a: Surgical specimen of schwannoma



b: Pathological findings of schwannoma

图5 神经鞘瘤术后标本及病理图

Fig.5 Postoperative specimen and pathological findings of schwannoma

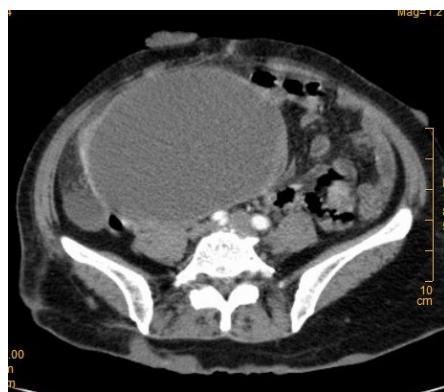


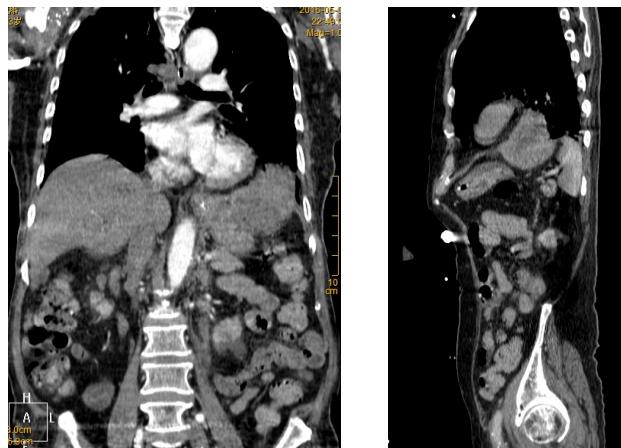
图4 末端小肠间质瘤动脉期扫描图

Fig.4 Arterial phase scan of distal small intestinal stromal tumor

5%发生在食管、肠系膜及腹膜后^[7],均具有潜在恶变的可能。与包括胃癌和结直肠癌在内的胃肠道粘膜肿瘤不同,GIST难以在手术前用内镜检查明确诊断^[8]。而且,由内镜引导的活检可能不足以采集到能做出正确诊断所需的组织量^[9-10]。

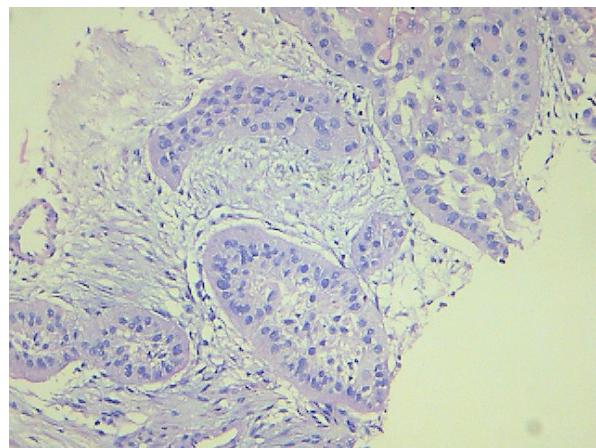
3.2 误诊因素分析

3.2.1 肿瘤定位错误因素 2例下腹部-盆腔的小肠间质瘤由于肿块巨大压迫正常结构,加上盆腔空间相对狭小,导致肿块与周围附件结构较难区分,从而定位诊断方向错误,且更加干扰诊断的是其中1例(图4)肿块大部分为囊性表现,误考虑为右侧附件来源的粘液腺囊



a: Coronal arterial phase scan of metastatic carcinoma

b: Sagittal arterial phase scan of metastatic carcinoma



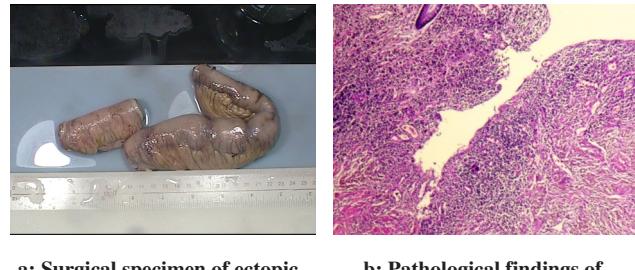
c: Pathological findings of metastatic carcinoma

图6 转移癌三维重建及病理图

Fig.6 Three-dimensional reconstruction and pathological findings of metastatic carcinoma

腺癌。术后反复认真观察影像图片可隐约发现右侧附件,但要相当仔细寻找才能发现右侧附件。1例术前考虑为胃底间质瘤、术后证实为转移性低分化腺癌病例中由于审核医师仅在轴位观察了肿块与胃底关系密切而没有仔细对肿块进行冠、矢状位重建。在后来重建冠、矢状位后(图6a和图6b),我们可以客观地发现肿块

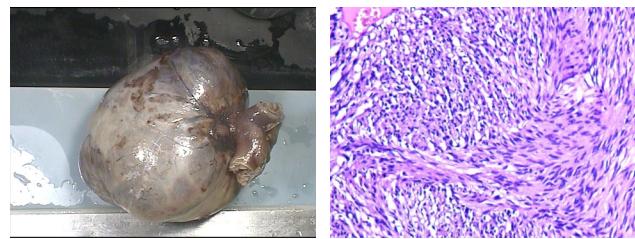
是跨左侧膈肌生长,与胃底联系不紧密。1例术前考虑小肠间质瘤,术后证实腹膜后神经鞘瘤病例中,中腹部见一巨大肿块,术前被肿块与十二指肠水平段关系密切表现干扰,错误地认为是来源于小肠的肿瘤,再加之肿块巨大但周围相对清晰,以致最终误诊为小肠间质瘤。1例平滑肌瘤病例中肿块与升结肠前壁关系密切,



a: Surgical specimen of ectopic pancreas
b: Pathological findings of ectopic pancreas

图7 异位胰腺术后标本及病理图

Fig.7 Postoperative specimen and pathological findings of ectopic pancreas



a: Surgical specimen of small intestinal stromal tumor
b: Pathological findings of small intestinal stromal tumor

图8 小肠间质瘤术后标本及病理图

Fig.8 Postoperative specimens and pathological findings of small intestinal stromal tumor

定位错误,从而导致进一步错误诊断为结肠间质瘤。术后病理为肠系膜来源的平滑肌瘤,再次提示准确定位的至关重要性。

3.2.2 忽略细微关键征象因素 另外1例神经鞘瘤中,肿块位于胃底与脾脏间隙且与胃底外侧壁分界不清,术前误诊为胃底外生性间质瘤。后来发现尽管肿块确实与胃底外侧壁关系密切,但忽略了肿块(图1)包埋肠系膜动脉分支、沿血管神经分布走形、肿块强化程度相对较轻的影像特点,且生长方向是向左肾上腺、胰腺靠近,压迫,有朝着腹膜后血管神经走向趋势。此例病人术前欲做出正确诊断确实较难,再次提醒笔者在以后工作学习中肿块生长是向腹膜后神经血管走形且肿块均匀轻度强化时,注意要把神经鞘瘤的鉴别诊断考虑进去。另外1例网膜囊淋巴结转移性低分化腺癌肿块位于肝左叶、胃小弯侧及胰腺上方间隙,初诊时由于忽略了该肿块对胃小弯侧壁局部侵犯增厚的观察,加上肿块边界清晰,误诊为了胃体小弯侧外生性间质瘤,此病例较难正确诊断。2例低分化腺癌中发现肿块较大呈分叶状、表面不光整且与肠腔相通、周围脂肪间隙模糊、周围可见肿大淋巴结、增强扫描轻度不均匀强化等都与间质瘤诊断相背离,实属对征象分析不仔细所致。

3.2.3 临床少见病例因素 2例异位胰腺的病理结果

确实出乎意料,术前正确诊断确实较难,因为该两例结节都未与正常胰腺密度强化完全一致。但结节都比较小均小于2 cm,且强化是持续均匀明显强化,这些特点可以扩展我们异位胰腺诊断的思维。1例类癌病例中,胃体肿块椭圆形,强化明显,欲术前正确诊断亦较难,但3期增强扫描中发现该肿块形态有变化、相对有活动,该特点是否会有提示意义,这点值得以后进一步病例佐证。1例未分化肉瘤病例中,是1例十二指肠附近多发肿块,平常学习工作中多发肉瘤实在少见,且周围脂肪密度增高伴有渗出、散发淋巴结,这些特点都不支持肉瘤的诊断,故术前误诊为恶性间质瘤。具体误诊原因及各自误诊比重见表2。

3.3 鉴别要点

发生在胃肠道腔内外或跨壁生长,边界清晰的肿块,无论其密度是否均匀、强化程度如何,都应该要考虑GIST。需要鉴别的主要有下列疾病:①胃肠道癌,经典的表现为局部不规则肿块起源于粘膜层,粘膜明显僵硬、中断或破坏征象,周围胃肠壁增厚、僵硬,与周围组织分界不清,周围常常见到淋巴结,但笔者发现绝大多数GIST增强扫描静脉期强化程度高于动脉期强化程度也有助于与胃癌的鉴别,此与张静等^[11]报道相符。②粘膜下肿瘤,当影像考虑肿瘤是来源于粘膜下时,应与胃肠道淋巴瘤、平滑肌瘤、平滑肌肉瘤、神经纤维瘤鉴别^[12]。Choi等^[13]指出平滑肌瘤好发生于胃贲门、均匀强化、肿块内少见坏死这些征象更支持平滑肌瘤的影像诊断。由于GIST与胃肠道神经鞘瘤的影像表现较相似,欲在手术之前准确区分胃肠道间质瘤和神经鞘瘤仍然具有不小的挑战性^[14]。Yang等^[15]指出肿块侵犯胃食管连接处以及长短径之比也高度提示平滑肌瘤的影像诊断。Choi等^[16]认为外生性或混合性生长及较慢倍增时间模式是与胃神经鞘瘤诊断相关的最常见CT表现特征。Choi等^[13]证实GIST与胃肠道神经鞘瘤之间的增强模式,肿瘤大小以及坏死和肿大淋巴结的存在显著不同。He等^[17]提出,GIST显示了恶性肿瘤的特征,包括血供丰富,生长方式更快,体积更大,坏死和囊肿变化频繁,可用于区分胃肠神经鞘瘤。胃肠神经鞘瘤作为良性肿瘤,更可能是均匀强化,并且在神经鞘瘤中囊性变,坏死或淋巴结的存在相对罕见。王静等^[18]表示胃肠道淋巴瘤肠壁增厚、边界清晰,内部积气积液显示“真肠腔征”,往往提示淋巴瘤。神经鞘瘤肿块沿腹腔或腹膜后血管神经走形生长趋势,强化较均匀且程度较轻。Choi等^[16]指出,肿块外生型或混合型生长方式、均匀强化、瘤周淋巴结的出现及肿块倍增时间较慢这些特点都更支持神经鞘瘤的诊断,另有指出肿块坏死是胃肠道间质特异性较高的影像征象而瘤周淋巴结



表2 胃肠道间质瘤MSCT误诊原因及比重

Tab.2 Causes of misdiagnosis and proportion of gastrointestinal stromal tumor misdiagnosed with MSCT

Causes of misdiagnosis	Preoperative conclusions	Postoperative pathological results	Number of cases	Proportion/%
Incorrect localization of tumors	2 cases of adnexal tumors	2 cases of small intestinal stromal tumors	5	38.45
	Small intestinal stromal tumor	Retroperitoneal schwannoma		
	Gastric stromal tumors	Metastatic adenocarcinoma		
	Ascending colon stromal tumor	Mesenteric leiomyoma		
Ignoring key signs	Exogenous stromal tumors	Metastatic adenocarcinoma	4	30.77
	Exogenous stromal tumors	Retroperitoneal schwannoma		
	2 cases of gastric exogenous stromal tumor	2 cases of poorly differentiated adenocarcinoma		
Rare clinical cases	2 cases of gastrointestinal stromal tumor	2 cases of ectopic pancreas	4	30.77
	Gastric stromal tumor	Carcinoid		
	Small intestinal stromal tumor	Mesenteric undifferentiated sarcoma		

出现则是神经鞘瘤特异性较高的影像表现。

简而言之,GIST有一定的影像特点,尤其是胃、小肠的典型间质瘤更具有代表性。笔者认为欲降低GIST的误诊,三维重建-肿块准确定位是重中之重,其次应仔细观察肿块细微关键征象,最后注意肿块血供来源及生长趋势也是鉴别要点。

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