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Carcinoid tumor in the rectum

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Incidence and distribution

Carcinoid tumors of the rectum are uncommon tumors [1]. However, recent report shows that the number of reported cases increases rapidly in both Japanese and non-Japanese series [2], probably due to the constant increase number of colonoscopic examination. The rectum is the most common site of gastrointestinal carcinoid tumors in Japan and the third most common site in American-European countries[3-5]. Even in US, recent report shows that carcinoid tumors of the rectum represented the most frequent primary site among gastrointestinal carcinoid tumors [6].

It occurs most commonly in indivisuals of their 5th to 6th decades, and the male-to-female ratios is 1.0 to 1.11 [7, 8]. Carcinoid tumors of the rectum usually arise singly, and only less than ten cases of multiple carcinoid tumors are reported in English literature [1, 9-11]. Most of the tumors are situated between 4 and 13 cm above the dentate line [2, 6, 9].

Symptoms and diagnosis

Sympoms include bleeding, constipation, rectal pain, tenesmus and so on, but approximately 50 per cent of the patients with rectal carcinoids are asymptomatic [6, 9, 13]. Carcinoid syndrome is virtually unknown with rectal carcinoids [9], and level of urine 5-hydroxyindoleacetic acid or serum serotoin are within normal limit [8].

Rectal carcinoid tumors are often detected incidentally by digital examination, proctoscope or sigmoidoscopic examination as the distinctive yellow, submucosal appearance with very firm or hard, discrete, smooth and mobile feel [6, 9]. Barium enema study might be useful to confirm the site of the lesion in the rectum and for planning the treatment method. Endoscopic ultrasonography is a useful tool for preoperatively determine the depth of invasion of carcinoid tumors, but the overall accuracy is only 75 % [14]. To evaluate the metastatic disease if suspected, abdominal echosonography, abdominal CT scan, chest X-ray or chest CT scan should be used.

Biological behaviour

Carcinoids are slow-growing tumors of neuroectodermal origin. It was believed that the tumor was similar to carcinoma because metastases could develop, but the clinical course often tended to be relatively benign. Seventy-two per cent of rectal carcinoids are reported to be a localized disease [3]. Several parameters have been suggested as predictive parameters in the assessment of the malignant nature of rectal carcinoid tumors, including tumor size, depth of invasion, lymphatic permeation, histologic growth pattern, presenting symptoms, morphologic factors, atypical histology and so on [1, 2, 9 12, 13]. Tumor size and depth of histologic invasion have been proposed as the two most important guides to the malignant nature of rectal carcinoid tumors [3, 4, 9, 12, 15-19, 21-28] though some exceptional cases have been reported [29, 30].

Tumors greater than or equal to 2.0 cm have 60 to 89 per cent incidence of metastasis, tumors measuring from 1.0 to 1.9 cm have a 10 to 15 per cent incidence of metastasis and tumors less than 1.0 cm have less than 2 to 5.5 per cent incidence of metastasis [1, 31] (Table 1). Histologic invasion of the muscularis propria is reported to be another indispensable combined factor with the size to predict metastases [1, 4, 12, 16, 18, 26]. Shirouzu et al. reported that rectal carcinoid tumors less than 2 cm in diameter had neither muscle invasion nor lymph node metastasis [19].

Table 1 - Frequency of lymph node metastasis and treatment modality

	Tumor size		
	< or = 1 cm	1-1.9 cm	> or = 2 cm
Lymph node metastasis	< 2 – 5.5 %	10 – 15 %	60 – 89 %
Treatment modality	local therapy (EMR*/local excision)	local excision or colectomy	colectomy (lymph clearance)

*EMR: endoscopic mucosal resection

Selection of treatment methods

Approximately 80 per cent of the lesions are less than 1.0 cm in size, submucosal, and show no metastatic spread, therefore local therapy including endoscopic excision (endoscopic mucosal resection; EMR and other methods) or local resection (transanal resection and so on) is an adequate and curative modality for these small and benign lesions [6, 8, 9, 12, 13, 16, 17, 27-29, 32](Table1). For tumors between 1.0 and 1.9 cm in size without preoperative evidence of lymph node metastasis and invasion into the muscularis propria, proper local excision method is recommended providing enough of a specimen to evaluate the depth of invasion, lymphatic permeation and other factors predicting malignant potential [2, 13, 25]. When the neoplasm is 2.0 cm or greater or muscular invasion or node metastases are present, radical extensive surgery including low anterior resection with total or tumor-specific mesorectal excision, or abdominoperineal resection is performed. This view for bigger lesion has been challenged recently by Sauven et al., who found no survival benefit even with aggressive surgery [9, 23]. They concluded that local excision is adequate, provided the entire tumor can be removed. So far, local therapy seems to be adequate for single tumors less than 2.0 cm with no sign of muscular invasion or lymph node spread preoperatively, provided total excision is performed to offer a specimen with proper histologic evaluation. Multicentric carcinoids might be considered differently and be regarded as malignant even when the size of tumor is less than 1.0 cm though the condition is extremely rare [1, 9]. It is because involvement of lymph nodes has been reported in cases with multiple carcinoid tumors of the rectum [10, 11].

Outcomes of each treatment modality

Endoscopic resection

Endoscopic excision of carcinoid tumors has recently been reported as a new local therapy [14, 28, 30, 32-38]. It is the least invasive treatment among local treatment methods. The maximum diameter of the tumors with endoscopic resection has been less than 1 cm in most reports. Several endoscopic techniques including strip biopsy, endoscopic mucosal resection, an aspiration method with a transparent cap and band-snare technique have been performed for this entity [14, 32, 33-38], however incomplete resection of tumors and/or unclear surgical margin or curability have emerged as difficulties due to limited excision up to the submucosal layer and the burn effect [6, 14, 30, 32, 34-36]. Incidence of surgical margin positive or unclear or doubtful has been 24 to 42 per cent on endoscopic treatment and many of them underwent additional operation [14, 32, 34, 36].

Local excision

Conventional transanal local excision is a useful procedure and has often been applied for this pathology as a least invasive method among several local excision procedures, which allow us full thickness excision for determining the histology and curability [21, 27]. However obtaining sufficient visibility has been sometimes difficult with the popular Parks and Mayo anal retractors, which resulted in excessive operating time and bleeding, and moreover access to proximal tumors was hard to perform with pre-existing methods [31-39, 40].

Transanal endoscopic microsurgery (TEM) has appeared as a new modality to access a high tumor with fine visibility [41-43]; however, the limited excisional layer up to the submucosa could not be avoided if the tumor was located above the peritoneal reflection. Moreover, the equipment was expensive and long training was required. The postoperative morbidity has also to be considered [41, 42].

A novel transanal local excision procedure called minimally invasive transanal surgery (MITAS) has been developed to facilitate full thickness excision even for high tumors,[44, 45] and applied for total excisional biopsy of rectal carcinoid tumors. This local excision technique includes the use of a new anal retractor [44, 46] and stapler with several surgical techniques to allow the procedure near the anus [8, 31, 44, 45]. When MITAS was compared with the conventional method (Table 2) [31], median operative time and bleeding volume were significantly less with MITAS than with the conventional method. Full thickness excision with free surgical margin was confirmed by histology in all cases in MITAS. The patients had no recurrent disease for a median of 41 months (range 2-67) after the operation in MITAS.

Table 2 - Outcome in MITAS and conventional method

	MITAS (n=12)	Conventional method (n=7)	p value
Tumor size	8 (3-15)	10 (3-18)	NS
Distance from anal verge	7 (5-12)	5.5 (4-8)	NS
Location of tumor		NS	
Rectosigmoid	1	0	
Upper rectum	5	1	
Lower rectum	6	6	
Operative time	21.5 (7-68)	35 (30-95)	$p < 0.003$
Bleeding volume	0 (0-20)	40 (0-150)	$p < 0.002$

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