Carcinoid of the Rectum

CASE REPORT AND SURGICAL MANAGEMENT

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SUMMARY

A case of rectal carcinoid is presented. The malignant potential of these tumours is emphasised. An approach to treatment of these lesions is outlined. Liver scintiscanning and peritoneoscopy are recommended as useful diagnostic aids in the assessment of these patients.

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Carcinoid tumours of the rectum present particular problems in management because microscopically they have the cytological features of a benign tumour, yet a significant number are malignant in their behaviour. Drastic differences in surgical management are required for the non-invasive and invasive stages of the neoplastic process, so that accurate evaluation of the tumour is mandatory. This case report demonstrates the malignant potential of these tumours and suggests the use of liver scintiscanning and peritoneoscopy as important facets of the diagnostic profile.

Otto Lubarsch[1] published the first description of a carcinoid tumour and established its origin from the crypts of Lieberkühn. Oberndorfer[2] coined the term carcinoid (carcinoma-like) because in his opinion the tumour was benign in behaviour, though having the appearance of a carcinoma. Saltykow[3] recorded the first case of a rectal carcinoid, found incidentally at autopsy. Masson[4] demonstrated that both the Kulchitsky cells found at the base of the crypts of Lieberkühn and carcinoid cells contained cytoplasmic argentaffin granules, thus confirming the origin of this tumour. The argentaffin reaction is not obtained in rectal mucosa or rectal carcinoids. Erspamer[5] isolated serotonin in the Kulchitsky cells and Thorson *et al.*[6] elucidated the clinical and biochemical aspects of the carcinoid syndrome. MacDonald[7] regarded all extra-appendiceal carcinoids as malignant and described a classification of invasiveness.

CASE REPORT

A Black male aged 60 years was admitted to hospital in January 1973 complaining of discomfort on defaecation and the occasional passage of a blood-streaked stool. Physical examination was non-contributory, except for a fingerbreadth, smooth hepatomegaly. Sigmoidoscopic examination revealed a pale yellow submucous nodule on the lateral wall of the rectum 6 cm from the anal verge, with maximum diameter 1,7 cm and minimal surface ulceration.

Biopsy of the base of the lesion showed a carcinoid tumour involving the muscle layers of the rectum. Radiological examinations, including barium enema, chest radiograph and abdominal lymphangiography, were all negative. A full blood count, blood urea and electrolyte estimations and liver function tests were also non-contributory. Urinary 5-HIAA estimation showed no elevation. The histopathological features prompted a liver scintiscan, which demonstrated numerous cold areas occupying the right lobe. The presence of liver metastases was confirmed by peritoneoscopy and biopsy of the liver under direct vision (Fig. 1). Because of the presence of liver metastases, the rectal lesion was removed by wide local excision and a rectal carcinoid, invading the muscle layers and with a maximum diameter of 1,7 cm, was confirmed (Figs 2 and 3).

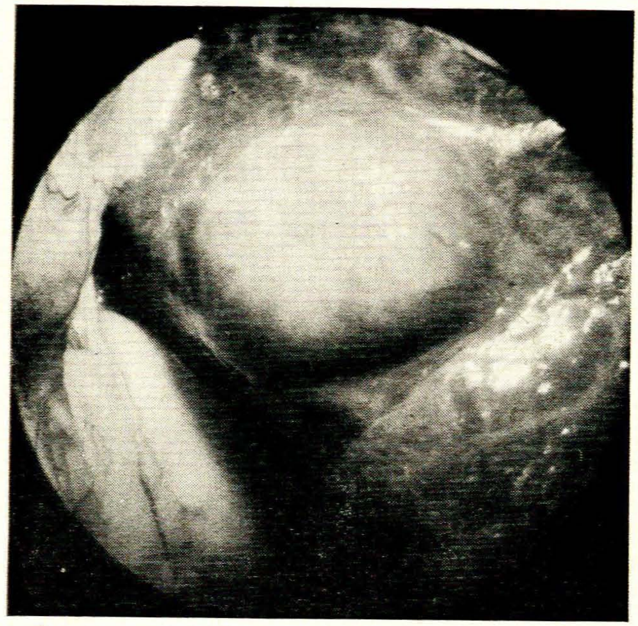

Fig. 1. Peritoneoscopic view of metastasis in the right liver lobe. The gall bladder is visible immediately beneath it.

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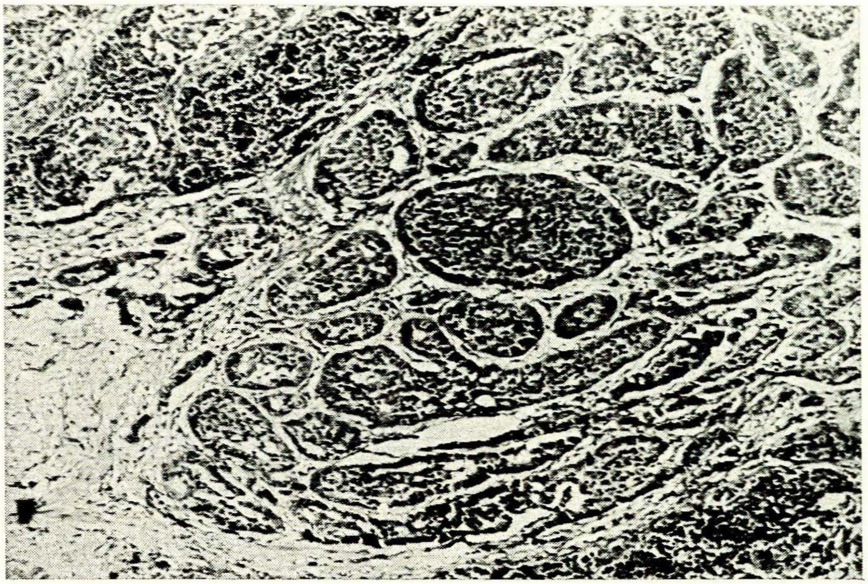

Fig. 2. Low-power histological section showing 'alveolar arrangement' of small round cells typical of carcinoid.

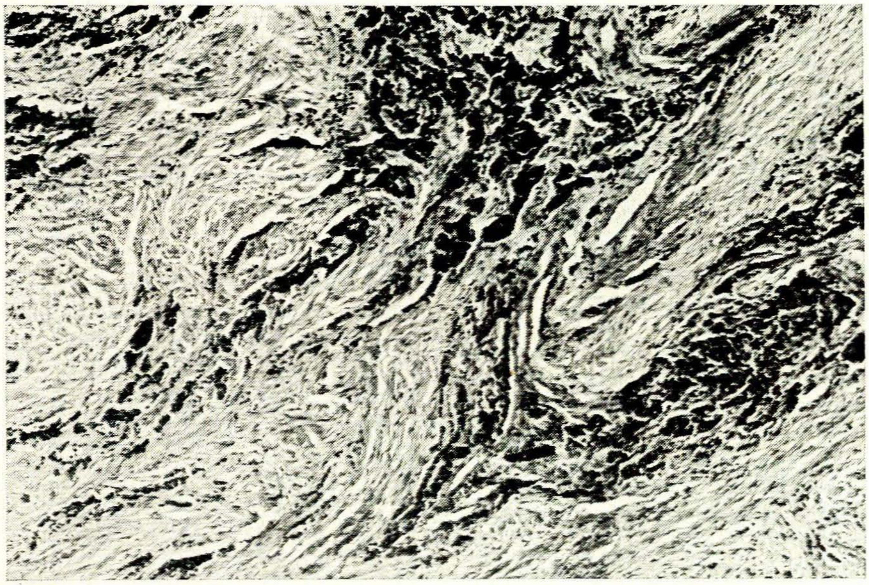

Fig. 3. High-power histological section showing invasion of muscularis by carcinoid cells.

DISCUSSION

This case demonstrated many of the classic features of this disease. Carcinoid tumours of the rectum are either benign or extremely malignant, although microscopically there may be no differentiating histological characteristics.

Pathology

Carcinoid tumours constitute less than 1% of gastro-intestinal neoplasms. They occur most frequently in patients in the fifth and sixth decades, and there are no significant differences in sex or racial incidence.[8] The lesions are usually solitary, the incidence of multiple tumours being 2 - 4%. The lesion appears as a yellow submucosal nodule with intact mucosa overlying it. Occasionally the tumour may be ulcerated, bleeding, polypoid, or it may even present as an annular constricting lesion. Rosettes, ribbons or masses of uniform small, round or polygonal cells with round nuclei and acidophilic cytoplasmic granules are found on microscopic examination. These cells, which characteristically do not give the argentaffin reaction, are found in a fibrous stroma which varies from scant to desmoplastic. Stout[9] believed that carcinoids that do not take a silver stain are tumours derived from Kulchitsky cells in the pre-enterochrome stage.

Malignant Rectal Carcinoid

Carcinoid tumours of the rectum characteristically appear benign on microscopic examination, according to the usual histological criteria. Tumour size and histological evidence of invasion into and beyond the muscle layers of the rectum are helpful in distinguishing between benign and malignant tumours. Caldarola *et al.*,[8] in a series of 139 cases, found that the chance of a carcinoid tumour of less than 1 cm in diameter showing deep invasion or metastasis was minimal. Tumours greater than 2 cm in diameter were always associated with metastases. In a collective review of published cases, Bates[10] reported that only 1,7% of 234 rectal carcinoids that measured less than 1 cm in diameter were malignant, and 10% of 77 tumours which measured between 1 and 2 cm in diameter were malignant. In contrast, 82% of 45 rectal carcinoids larger than 2 cm in diameter proved to be malignant. He also emphasised that muscle invasion is not invariable evidence of malignancy.

Orloff,[11] reporting a personal series of 38 patients, found that tumour invasion of muscle was invariably associated with lymph node or distant metastases. The principal sites for metastases are the regional nodes and the liver.

The rectal carcinoid removed from our patient was 1,7 cm in diameter and clearly showed muscle invasion. The use of liver scintiscanning and peritoneoscopy unequivocally demonstrated the presence of liver metastases. These investigations prevented unnecessary laparotomy and eliminated any consideration of radical surgery.

Clinical Features

The majority of patients with rectal carcinoids are asymptomatic.[12] The most common symptoms are rectal bleeding, constipation and tenesmus. Obstructive symptoms may develop when the lesion is large, and constitutional manifestations of malignancy occur with metastases. To date, one instance of the malignant carcinoid syndrome has been reported in association with a rectal carcinoid.[13] Elevated urinary 5-HIAA levels have been reported in 2 further cases without convincing manifestations of the syndrome.[14,15] Despite the presence of hepatic metastases in our patient, the urinary 5-HIAA levels were normal.

Treatment

Because of their location, carcinoid tumours of the rectum present particular problems in treatment. If they are benign all that is required is a minor local excision, but if they are malignant, radical extirpation necessitates an abdominoperineal resection.

For freely movable lesions less than 2 cm in diameter a wide, full-thickness local excision should be carried out, so that muscular invasion can be assessed. If no muscular invasion is present, local removal constitutes adequate treatment. If there is muscular invasion on microscopic examination and no liver metastases are present, the more radical procedure is indicated.

Abdominoperineal resection is indicated for: (*a*) lesions 2 cm or more in diameter; (*b*) lesions showing muscular invasion; (*c*) lesions recurring after local excision; and (*d*) fixed or annular lesions.

Large reported series[8,10,11] substantiate the validity of these therapeutic criteria.

Prognosis

Benign rectal carcinoids have a high cure rate after complete local excision. Rectal carcinoids with malignant characteristics have a worse prognosis than benign rectal carcinoids and metastasising carcinoids from other locations. Freund[16] reported an average survival time of 26,3 months in 34 patients after radical operation for malignant rectal carcinoid. Orloff[11] reported a 5-year survival rate of 40% in 15 patients with lesions measuring 2 cm or more in diameter, in contrast with the 100% 5-year survival rate of 23 patients with tumours measuring less than 2 cm.

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