

Primary Malignant Melanoma of Rectum

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INTRODUCTION

The vast majority of malignant melanomas arise in the skin, other less common sites include the eyes, leptomeninges. The anorectum is a rare site for noncutaneous melanomas. In fact, primary malignant melanoma of the anorectum accounts for less than 1% of all malignancies in this site and for 0.4%-1.6% of all primary melanomas. Moreover, whether melanomas can arise primarily in the rectum is a subject of great controversy, although Werdin et al and Nicholson et al reported rectal melanomas with adjacent junctional melanocytic proliferation, which suggested that they had arisen from melanocytes located in the rectum. In this paper, we report a case of primary malignant melanoma of the rectum.

CASE PRESENTATION

A 65-year-old man, previously fit and well, presented to the accident and emergency department following abdominal pain, vomiting and absolute constipation. On admission, he was haemodynamically unstable with feature of peritonitis. His abdomen was tense, tender and bowel sounds were absent. Rectal examination revealed an anteriolateral fleshy mass situated 5 cm from the anal verge which bleed on touch.

When questioned, the patient said he had been bleeding per rectum intermittently for 4 months but without any pain and also gives history of constipation. He used to smoke cigarette as well as hukka and was anti-HCV positive. Abdominal x-ray done showed air fluid level and no gas under diaphragm.

With the diagnosis of intestinal obstruction, patient was resuscitated and taken for exploratory laprotomy. The significant finding was perforation of rectum at the site of tumor mass. Biopsy was taken from the mass and sigmoid colostomy was done.

The gross pathological examination revealed a blackish tumor, measuring, 1x 0.5x1cm. Microscopically, the tumor was composed of malignant cells with bizarre and hyper chromatic nuclei. Many melanin pigments could be seen in the tumor cells. Computed tomography of the thorax, abdomen and pelvis showed no evidence of

metastasis. Dermatological and ophthalmological examinations revealed no evidence of a cutaneous or an ocular primary lesion.

Shortly after his diagnosis, the patient underwent an abdominoperineal resection (APR). He made an uncomplicated recovery and was discharged 12 days later. The malignant melanoma was completely excised with clear margins of at least 2 mm. Six out of six lymph nodes were negative for metastasis. He is currently being followed up will be considered for chemotherapy following repeat imaging.

DISCUSSION

Moore described melanoma of the rectum and anus in 1857. It is a rare malignancy that accounts for less than 1 percent of all colorectal and anal cancers. Only 2 percent of melanomas are located in the anorectum, yet this location is the third most common site of primary melanoma, behind skin and eyes.

Primary rectal malignant melanoma is regarded as such only if it arise from benign melanocytic proliferation in the rectal mucosa, and is located more than 4 cm from the anal verge. It is an uncommon highly malignant tumor associated with an extremely poor prognosis despite of aggressive surgical treatment.

The main complaints among patients with anorectal malignant melanoma are anal bleeding, an anal mass, and anal pain.

This disease presents itself usually starting from the fourth decade, predominantly in women, with an increase of incidence in the fifth or sixth decade of life. Almost 60% of patients have already metastases at initial diagnosis, as it is frequently mistaken for benign conditions as either a hemorrhoid or rectal polyp.

Five-year survival is rarely seen, as the large series have shown it to be less than 5%. The Mayo Clinic reported five-year survival as 22% and cure as 16% in their patient population

If a biopsy shows a specimen suspicious for sarcoma (e.g.leiomyosarcoma), one should be alert. Preferably, S-100 staining should be performed in addition to routine stainings. A positive S-100 stain suggests the tumour most likely to be a melanoma. Subsequently dissemination studies, including chest X-ray and CT-scan of chest/abdomen/pelvis are performed. Curative surgical resection has to be

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performed in the absence of metastases proven by dissemination studies.

There has been a debate in the literature regarding the extent of surgery for anorectal melanoma. Despite numerous attempts, there is no international consensus at this moment on which surgical approach is preferred as neither of surgical treatment confers obvious benefit on survival. Moreover, there is no consensus on follow-up guidelines, either. Surgical treatment of this specific tumour varies from very extended and radical operations (APR) to extremely conservatives procedures such as wide local excision (WLE).

A number of studies claim that an abdominoperineal resection (APR) is the treatment of choice. This is based on the hypothesis that the disease spreads proximally via the submucosa to the mesenteric lymph nodes. Retrospective studies have revealed a statistically significant improvement in local-regional control when patients are managed with APR compared with local excision alone.

Other studies, however, have recommended only a sphincter-saving excision for two reasons. Firstly, treatment is often palliative and wide radical surgery is unnecessarily mutilating. Secondly, tumor stage and biological behaviour of the tumor determines survival instead of the choice of surgical operation.

Some like Cooper et al advocate APR because of more effective local control and since it removes clinically undetectable lymph nodes. While others like Ward et al suggested wide local excision (WLE) with a 2 cm margin along with therapeutic inguinal lymph nodes dissection for positive nodes, as majority of the

patients have metastasis at the time of diagnosis and survival is not significantly better with radical surgery.

CONCLUSION

In summary, primary rectal malignant melanoma is an extremely rare tumor. In both anal canal and primary rectal melanoma, abdominoperineal resection may provide a better local control if not longer survival.

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