

Clear cell adenocarcinoma of the colon is a unique morphologic variant of intestinal carcinoma: A rare case report

Neha Bakshi, Shivani Sharma, Vijay Kaushal, Kavita Mardi

Department of Pathology, Indira Gandhi Medical College, Shimla, Himachal Pradesh, India

ABSTRACT

Primary Clear cell adenocarcinoma of the colon is extremely rare, with only a handful of cases reported in literature. We herein report a case of such a tumor in a 42-year-old male who presented with rectal bleeding and pain abdomen. A circumferential growth occluding the lumen at the hepatic flexure was visualized on colonoscopy. The right hemicolectomy specimen revealed an ulceroinfiltrative growth which showed histologic features of clear cell adenocarcinoma.

Key words: Adenocarcinoma, clear cell, colon

INTRODUCTION

Primary Clear cell adenocarcinomas are very rare in the colon. Most of the reported cases show predilection for elderly men and are preferentially located in the left colon. Almost all form part of a larger conventional adenoma. Herein we describe an intriguing variant of Clear cell Adenocarcinoma, occurring in a middle aged man and unusual in terms of both location (the lesion was located in the right colon) and morphology (was not accompanied by a conventional adenoma).

CASE REPORT

A 42-year-old male presented with rectal bleeding and pain abdomen of four months duration. Clinical examination revealed a firm lump in the right hypochondrium. CT Abdomen showed circumferential

wall thickening in the proximal transverse colon and adjacent hepatic flexure. A circumferential growth occluding the lumen at the hepatic flexure was visualized on colonoscopy. Preoperative serum CEA level was 3.34 u/ml. An extended right hemicolectomy was done. The surgical specimen consisted of an 18 cm long segment of colon revealing an ulceroinfiltrative tumour measuring 4 x 4 cm located 5 cm from one end, involving the mucosa and infiltrating through the full thickness of the colonic wall. Two lymph nodes were dissected out from the specimen.

Histology of the tumour revealed an ulcerated mucosal surface covered over by exudative material. The underlying tumour was composed of tumour cells arranged predominantly in a glandular pattern, having moderately pleomorphic vesicular nuclei with prominent nucleoli, abundant clear cytoplasm and well defined cell borders. Mitotic activity was low (1-2 mitoses/10 high power fields). Areas of necrosis were present. The tumour infiltrated through the full thickness of the colonic wall into the surrounding fat. On immunohistochemical staining, the tumour was positive for CK20 and negative for CK7. There was no residual classic adenoma at the periphery of the tumour. Both the resected lymph nodes were free from metastatic tumour deposits.

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Address for correspondence: Dr. Neha Bakshi, Flat no 2, Nikunj Bhawan, Anji, Vikasnagar, Shimla, Himachal Pradesh, India.
E-mail: bakshi.nb@gmail.com

DISCUSSION

In 1964, Hellstrom and Fischer described the first case of clear cell adenocarcinoma of the sigmoid colon, composed of neoplastic clear cells which resembled the physaliferous (clear) cells of chordoma tumors.^[1] Clear cell adenocarcinomas generally develop in in organs of mullerian duct origin such as the kidneys, ovaries, extraovarian endometriosis, uterine cervix, upper vagina, or lower genital tract.^[2]

Most reported cases of clear cell adenocarcinoma in the colon have been identified as a metastatic focus from other organs, including renal clear cell adenocarcinoma or those arising from an ovarian malignancy.^[3] Primary clear cell adenocarcinoma of the colon is an extremely rare occurrence.^[2,4,5] In a study involving 3,486 cases of colon cancer, only 0.086% had clear cell changes.^[6]

The histologic similarity may make it difficult to differentiate between primary colonic clear cell adenocarcinoma and metastatic clear cell adenocarcinoma.^[4] Preoperative investigations to rule out a primary malignancy at other sites

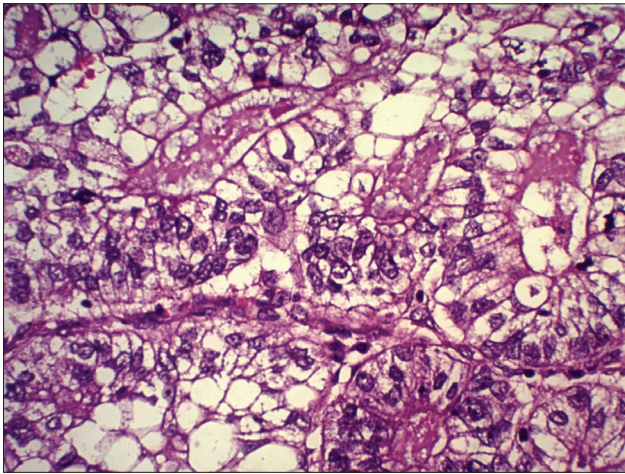


Figure 1: Tumour cells arranged predominantly in a glandular pattern, having pleomorphic vesicular nuclei, prominent nucleoli, abundant clear cytoplasm and well defined cell borders. (H and E, x400)

are important in such cases. In our patient both kidney and prostate tumor were ruled out preoperatively.

The mechanism causing clear cytoplasm remains unknown. Most reported cases have been attributed to accumulation of glycogen granules (PAS positive and mucicarmine negative).^[5] In contrast, some workers have failed to detect glycogen in the clear cells and suspected elution of glycogen granules during fixation or staining. These differences indicate the heterogeneous nature of the clear cells, and until now the pathogenesis and diverse potency of this malignancy remains unclear.^[5]

Almost all Clear Cell Carcinomas form part of a larger conventional adenoma, thus supporting the hypothesis of a linear carcinogenetic sequence from conventional adenoma, to clear cell-type adenomas and Clear Cell Carcinomas.^[7] Further, the different morphology of Clear Cell Carcinoma does not seem to reflect a distinct biological entity, but an unusual morphological variant with similar molecular profile of conventional colorectal carcinoma.^[7]

Most affected patients are elderly males and the tumor tends to be located on the left side.^[2,7] However, our patient was a middle-aged man, the lesion was located in the right colon and was not accompanied by a conventional adenoma.

Immunohistochemical study may prove indispensable in establishing the primary colonic origin of the lesion. 75-95% of primary colonic adenocarcinomas are CK20 positive and CK7 negative, whereas 80-100% of Müllerian origin adenocarcinoma are CK20 negative and CK7 positive.^[8] CDX2 is expressed in the majority of primary colonic adenocarcinomas and is negative in the Müllerian origin neoplasms.^[8]

Since most reported patients treated with polypectomy were not subjected to follow up, the prognosis of clear cell carcinoma remains unclear. Further reports are therefore necessary for elucidation of this rare entity.

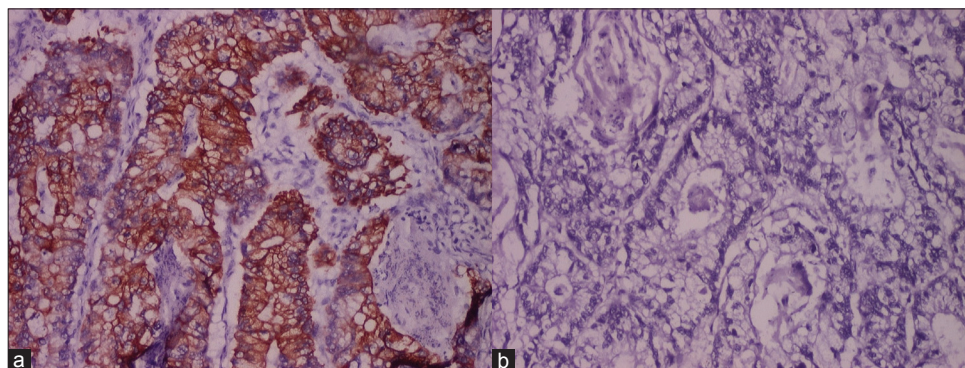


Figure 2: (a) Tumour cells revealing strong positivity for CK 20, (b) Tumour cells are negative for CK 7

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