

## LARGE RECTAL VILLOUS ADENOMA PRESENTING WITH OBSTRUCTION AND McKITTRICK - WHEELLOCK SYNDROME

### MEKANİK TIKANMAYA YOLAÇAN REKTAL VİLLÖZ ADENOM: McKRITICK - WHEELLOCK SENDROMU

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**ÖZET:** Büyük villöz adenomlar belirgin olarak rektumda yerleşirler ve sıklıkla rektum karsinomu ile birlikte dirler. Çoğu zaman yumuşak ve parçalanabilir olmalarına karşın, nadiren tam barsak tıkanmasına da yol açabilirler. Nadir olarak şiddetli sıvı ve elektrolit kaybına yol açan aşırı diyare ile karakterli ilginç bir sendrom oluşturabilirler. Cerrahi öncesi yoğun replasman tedavisi gereklidir. Artmış malignite gelişim sıklığı nedeniyle, malign dokunun bulunup bulunmadığını tam bir eksizyon ile saptamalıdır. Malignite barındıran büyük villöz adenomlu bu olgu sonununun amacı , rektumun bir adenomatöz polipi olan bu özel formunu tanıtmak ve uygun tedavi seçeneklerini tartışmaktadır.

**Anahtar Kelimeler:** Villous adenoma, rektum, obstrüksiyon, mckittrick - wheellock sendromu

#### INTRODUCTION

Villous adenomas constitute only about 4% to 10% of adenomatous polyps in the colon and rectum<sup>1-4</sup>. While tubular adenomas are more evenly distributed throughout the large intestine, VAs occur most frequently in the rectosigmoid. There is universal agreement that carcinoma is often found in VAs, the reported frequency varies from 30% to 70 %<sup>1-4</sup>. Biopsy is absolutely unreliable as a means of rulling out the presence of carcinoma<sup>1-5</sup>. Their distinct morphologic characteristics, particularly in very large and bulky tumors involving the entire circumference of the bowel may rarely be responsible for the development of clinical manifestations of rectosigmoid obstruction and McKittrick-Wheelock Syndrome.

#### REPORT OF A CASE

A 73- year-old female patient with a sixth months past history of symptoms of satiety, fatigue, fainting , distention, abdominal pain, excessive discharge of mucus, and intermittent diarrhea and rectal constipation was admitted to the Emergency Unit complaining of dehydration, weakness, nausea, incontinence and weight loss for seven days. She was known to have primary hypertension. She presented with severe hypotension, oliguria, and circulatory collapse: her blood pressure,

80/60 mm Hg; the pulse rate, 105 beats per minute; and respiration rate, 20/min. The temperature was 36.2oC. Her initial laboratory values were as follows: WBC:16.400 cu. mm.; platelets:269.000 cu. mm.; BUN:12 mg/dl; Creatinin:4.7 mg/dl; Na:130 mEq/L; K:2.6 mEq/L; Ca:8.2 mg/dl; Hb:14.2 g/dl; pH:7.3 and HCO<sub>3</sub>:10.7. The main physical findings were abdominal pain, distention, and profuse diarrhea with mucus, except rectal bleeding. Prompt resuscitative therapy was provided in an emergency setting besides the diagnostic work-up. Urgent proctosigmoidoscopy revealed a sessile polypoid lesion situated at the tenth centimeter of the rectum, showing the typical appearance of VA that was not suitable for endoscopic removal. After the period of fluid and ion replacement therapy, she was operated for resective surgery: it was found to be a very large, approximately 13 cms. in its largest diameter, rectal VA obstructing the bowel (Fig. 1) and the Hartmann's procedure was performed with clear resected surgical margins. Adenocarcinoma was found in VA penetrating through the lamina propria and muscularis mucosa, without any tumor deposits in eight lymph nodes that removed. Her postoperative course was uneventful.

#### DISCUSSION

The VA is distinct from the tubular adenoma in its occurrence, morphology and, behavior. Compared with tubular adenomas, VAs are uncommon<sup>1,3</sup>. They develop more often in men than in women, in a ratio of about 3.2, and their peak occurrence rate is in the sixth and seventh decades of life<sup>1,3</sup>. In contrast to tubular adenomas, VAs occur predominantly in the rectum<sup>1-4</sup>. For example, of

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219 VAs found in 215 patients, Quan and Castro reported 144 in the rectum and 54 in the sigmoid<sup>1</sup>. They are typically sessile and only occasionally pedunculated, and arise from the mucosa over a broad and spreading base, sometimes involving the entire circumference of the bowel, varying in size from 0.5 to 12.0 cms<sup>6-7</sup>. The surface of the lesion has a cauliflower-like appearance and is soft, friable and velvety (Fig. 1)<sup>1,6,7</sup>. So it may easily be missed by digital examination, even when located in the lower rectum<sup>1,3</sup>. Areas of firmness should raise suspicion of malignancy.



**Fig. 1: Large VA having a shaggy surface with obvious papillary fronds**

VAs are distinguished morphologically by their larger size than other types of polyps<sup>5</sup>. On gross examination, VAs tend to have a shaggy surface with obvious papillary fronds<sup>6</sup> (Fig. 1). Microscopically, they consist of finger-like processes extending outward from the surface. These contain a core of lamina propria and are covered by the neoplastic epithelial cells growing toward the bowel lumen<sup>7</sup>. The mucus content of the adenomatous cells is usually decreased and occasional adenomas have abundant mucus production<sup>7</sup>. The reported frequency of malignancy varies from 30% to 79% of VAs, the likelihood of malignancy increasing with the size of the lesion. Synchronous and metachronous carcinomas are also common: Quan and Castro reported that 49% of their patients had more than one adenoma, 16% had associated bowel malignancies and, remarkably, 12% had extracolonic cancer<sup>1</sup>. Thomson et al. (1977) found one or more additional tumors in 30 of 121 (25%) patients with sessile VA of the rectum. Jahadi and Baldwin (1975), Adair and Everett (1983), and Christiansen et al. (1979) found that 12% of their patients had an associated lesion<sup>3</sup>. Although the gross appearance of VAs is sufficiently characteristic to have a provisional diagnosis, biopsy specimens may be misleading<sup>1-8</sup>. In Quan and Castro's series of 219 VAs, over half the initial biopsies called "benign" were turned out to be tumors later found, when examined whole, to have malignant changes<sup>1</sup>. It is important therefore, to totally excise and carefully section the whole<sup>1-8</sup>. Even though

some tumors present incidentally, larger lesions often cause severe symptoms like rectal bleeding, bowel habit changes, distention, abdominal discomfort, excessive discharge of flatulence and/or mucus, prolapsing and, interestingly expelling the fragments of tumor tissue, and diarrhea, which can lead to fluid and electrolyte depletion. Very occasionally, patients will present with the full-blown syndrome—namely McKittrick and Wheelock Syndrome—and complain of dehydration, confusion, weakness, weight loss, nausea, depression, and incontinence with severe hypotension and circulatory collapse due to profuse mucous diarrhea<sup>1-4</sup>. Hypovolemia, prerenal azotemia, severe hypokalemia, hyponatremia, and hypochloremia are often found. Unless it is recognized and prompt resuscitation is provided preoperatively, death from remote organ failure may ensue<sup>1-4</sup>. Babior (1996) emphasized the presence of large stool volumes, sometimes up to 4 liters in 24 hours, and the high incidence of hypokalemia<sup>1</sup>. The definitive therapy is complete surgical resection of the tumor. Because 20 to 25 percent of sessile VA contain foci of invasive cancer, and random biopsies are unreliable in detecting malignancy, surgical management of large rectal villous tumors may be particularly difficult, thus explaining the variety of surgical approaches (the transanal, the transsphincteric or the transabdominal route) that have been described in the management of these lesions. Complete regression of VAs of the colon using the nonsteroidal anti-inflammatory drug, piroxicam, was also documented<sup>9</sup>. Smaller villous lesions can be removed by a combination of snaring and fulguration (SF) or transanal excision (TE) described by Parks and Stuart<sup>8</sup>. However, TE results are more reliable tumor eradication than SF<sup>2,8</sup>. The transsphincteric approach of Bovan, and later popularized by Mason, provides good access to the rectum; but most surgeons are reluctant to divide the anal sphincter<sup>2,3,8</sup>. For very extensive or high-situated rectal villous tumors that can not be dealt with in these ways, some form of anterior resection or coloanal (CA) excision, or even abdominoperineal resection (APR) will need to be performed<sup>2-4,8</sup>. For extremely large tumors that extend to dentate line, CA anastomosis is appropriate whilst the APR should be reserved for those patients with a diagnosis of invasive carcinoma in whom lesser procedure would not constitute adequate treatment<sup>2-4,8</sup>. In very occasional emergent cases presenting with delayed obstruction and the clinical features of syndrome, Hartmann's procedure may be an acceptable alternative to the other surgical modes of therapy. More recently, transanal endoscopic microsurgery described by Beuss et al, a form of minimal access surgery, has been used to treat a variety of rectal lesions including villous tumors<sup>2,8</sup>. The authors have no experience with this technique and fail to see a significant role for its use because of its expense, the need for specialized training, and the infrequency with which other transanal techniques are insufficient.

## KAYNAKLAR

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