Primary Rectal Malignant Melanoma

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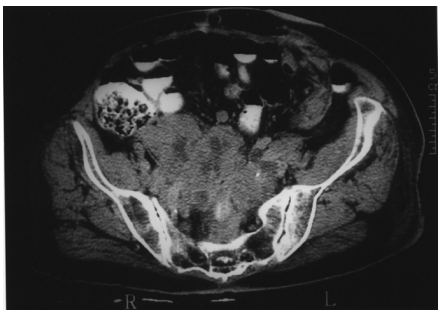
Unlike cutaneous melanoma that is usually diagnosed early, anorectal melanoma is diagnosed late in its course, resulting in a very poor prognosis. We report a case of initial large malignant melanoma of the rectum with a rapid and fatal course.

Case Description

A 78-year-old woman was admitted to our department with recurrent bleeding and 5 kg weight loss during the previous 8 months. Her medical history included non-insulin-dependent diabetes mellitus and chronic obstructive lung disease. Her previous surgical record disclosed hysterectomy due to prolapse, and hip replacement.

On physical examination a non-tender rectal tumor was palpated. Stools were positive for blood. Electrolytes were normal and hemoglobin concentration was 10.1 g/dl. Colonoscopic examination revealed a 2 cm polypoid tumor in the rectum 4 cm proximal to the anus. Transrectal ultrasound confirmed a rectal tumor larger then 5 cm occupying more than 270°. Computed axial tomography revealed a well-defined lobulated soft tissue mass in the rectum, which invaded the perirectal and retrovesical fat planes [Figure]. Biopsies were taken during colonoscopy and directly under general anesthesia. Both revealed malignant melanoma. Under general anesthesia a formal abdominoperineal resection of the rectum was performed, with a total mesorectal excision including the lateral ligament lymph nodes. An ulcerated 6.5 cm tumor was found in the rectum. The tumor penetrated through the entire rectal wall and reached the perirectal fat. On histological examination the tumor was found to be a malignant melanoma. Metastatic tumor was found in 12 of 12 regional lymph nodes. The surgical margins were free of tumor. The patient was discharged from our department 10 days after the operation following a non-eventful course.



An axial CT scan taken at the lower abdomen revealed a huge non-homogeneous mass of enlarged lymph nodes.

Three months later she was re-admitted with deep vein thrombosis of both legs. The CT scan demonstrated bilateral hydronephrosis and massive retroperitoneal lymphadenopathy. A huge lobulated mass was also seen in the lower abdomen. The patient refused any treatment and died a month later.

Comment

The first rectal malignant melanoma was reported by Moore in 1857 [1]. This tumor is very rare, with a reported incidence of 0.4–1.6% of all malignant melanomas in humans. Primary melanoma of the rectum and anal canal represents 1% of tumors in this area [1]. The tumor has been reported mainly in older patients with a median age of 66 years [2], and the common initial symptoms are rectal bleeding (87%) and/or anal pain (33%) [2]. The clinical diagnosis could be missed due to non-specificity of clinical symptoms, or confused with hemorrhoids, polyps or benign skin pigmentation of the peri-anal area. Barium enema has a limited diagnostic value, while CT demonstrates the tumor extension and the distant metastases. Rectal ultrasonography accurately demonstrates the depth of invasion of the tumor. In view of its high accuracy, rectal ultrasound is now considered by many authors as the procedure of choice for tumor staging in patients with primary melanoma of the rectum. The role of magnetic resonance imaging today is very important in unusual tumors appearing in the pelvic region, with regard to extension and penetration to adjacent tissues. The final histopathological diagnosis is difficult especially in the amelanotic form, which represents 25% of cases. In these cases the diagnosis is established by complementary immunohistological methods [3].

The optimal therapeutic approach is still disputed but surgery remains the preferred method of treatment. The surgical procedure varies from local

trans-anal excision to abdominoperineal resection. Radiation is a palliative therapy in an extensive tumor. Chemotherapy may be attempted in cases with metastatic spread. In a few cases published [4], patients who refuse surgical treatment usually die within 2 years following diagnosis [2]. Survival is dependent on tumor staging, ulceration and destruction, involvement of lymph nodes, and histological appearance [5]. The reported 5 years overall survival is 6–15% of patients, depending on the stage of the disease and the medical condition of the patient.

We present a case in which the initial surgical treatment for an advanced tumor did not suffice in preventing the tumor spread and the patient's eventual death. Only a combination of all modes of therapy in an advanced disease could provide a chance for a longer temporary remission in this aggressive tumor, although the benefit of any form of surgical treatment in this context is still disputed in the literature.

References

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