An inside story of a thrombosed pile

Yashwant Kumar,[1] Alka Bhatia[2]

[1]Clinical and Pathology Laboratory, Chhuttani Medical Centre; [2]Department of Experimental Medicine and Biotechnology, Post Graduate Institute of Medical Education and Research, Chandigarh, India

Abstract

We report a case of extremely rare variant of ano-rectum malignant tumor. The tumor is often misdiagnosed as either hemorrhoids or rectal polyp, which are benign diseases. On histology also this variant may be confused with other more commonly occurring spindle cell lesions in this area; its recognition is therefore important as it normally has a poor prognosis.

Introduction

In ano-rectum different types of neoplastic as well non-neoplastic lesions can occur. Among the latter, hemorrhoids, anal fissure or polyps are common.[1] This is a report of an anal growth, which was clinically diagnosed as thrombosed pile but histopathology and immunohistochemistry revealed its real nature.

Case Report

A 60-year-old male presented to surgical outpatients department complaints of bleeding from rectum and pain during defecation. He reported a history of weight loss and anorexia, and at examination he was pale and cachectic. On rectal examination a growth about 3×2×1 cm was found in the rectum, 4.3 cm from the anal verge. The clinical diagnosis was of a thrombosed pile, therefore it was excised and sent for histopathological examination.

The tissue was in pieces, firm and brownish-black colored. The examination at the microscopy revealed a tumor replacing the entire wall thickness with ulceration of overlying epithelium. The tumor cells were disposed in fascicles and sheets (Figure 1A) and were spindle-shaped. Few of them showed prominent inclusion like nucleoli and atypical mitotic figures, while many contained blackish pigment (Figure 1B). Schmorl's stain confirmed the pigment to be melanin (Figure 1C). The tumour cells were strongly positive for S-100 and HMB-45 (Figure 1D). Based on these findings a diagnosis of sarcomatous variant of malignant melanoma was made.

Thereafter, an abdomino-perineal resection with removal of bilateral inguinal, pelvic and mesorectal lymph nodes was carried out. On histological examination no residual tumor was identified at the primary site, however one of the inguinal lymph nodes was found to harbor the metastatic tumor deposits. The patient received radiation and chemotherapy and was then discharged and advised for a regular follow-up. The patient however came back after 13 months and on computerized tomography (CT) scan of abdomen showed multiple secondaries in liver. The patient was discharged on his own request and thereafter no follow up could be obtained.

Correspondence: Alka Bhatia, The Pines, Near Ashiana Regency, Chhota Shimla (HP), 171002 India.
E-mail: alkabhatia@ymail.com

Key words: malignant melanoma, sarcomatous, pile.

Conflict of interest: the authors report no conflicts of interest.

Received for publication: 14 March 2011.
Accepted for publication: 19 March 2011.

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Clinics and Practice 2011; 1:e16
doi:10.4081/cp.2011.e16

Discussion

Malignant melanoma is a rare neoplasm in the ano-rectum accounting for only 1-3% of all tumors.[2] Most of the malignant melanomas are of classical type and the sarcomatous variant, although it is well known in skin has been rarely reported at this site.[3] By reporting this case we intended to alert clinicians and pathologist about its occurrence and importance of its recognition. A dermal sarcomatoid melanoma has even worse prognosis than classical malignant melanoma with a 5-year survival rate accounting to only 15%, despite an aggressive, multimodality approach.[4]

It has been reported that 38% of patients have already metastatic disease at the time of diagnosis.[1] However, its prognosis in ano-rectum is not yet clear due to its rarity at this site.

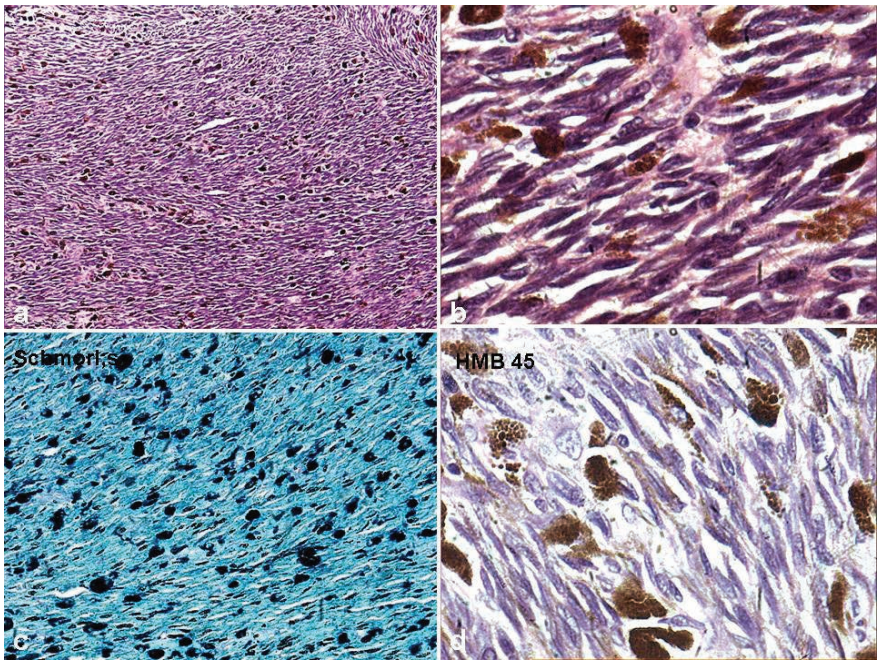

Figure 1. Microphotograph of the tumor in the ano-rectum showing (A) the tumor cells arranged in long parallel running fascicles (B) the high-power view of individual cells with prominent spindling and abundant blackish pigment in many of them (C) Schmorl's stain demonstrating the pigment to be melanin stained as blue green granules and (D) Strong positivity for HMB-45 in the tumor tissue.

The present case also showed a dismal prognosis, as patient developed disseminated malignancy despite aggressive therapy. Further data on the various prognostic factors is needed, which is possible only if more cases are reported.

Its diagnosis also may be difficult even on histology and it may be confused with other more common spindle cell tumors i.e. leiomyoma, haemangioma, haemangiopericytoma, neurilemmoma, neurofibroma, granular cell tumor, spindle cell lipoma, gastrointestinal stromal tumor, malignant fibrous histiocytoma, leiomyosarcoma, rhabdomyosarcoma, fibrosarcoma and spindle cell carcinoma.[5] The key diagnostic feature is the melanin production, which may be absent in approximately 20% of the cases.[4] In such difficult cases only immunostaining can help in achieving the correct diagnosis.

A sarcomatous melanoma in ano-rectum is seldom reported in the literature and the present case highlights several key points. Most importantly, when faced with a spindle cell lesion of anal canal, especially in an elderly, the diagnosis of sarcomatous melanoma must be included in the differential. As occurred in our case, the clinical impression is often misleading, therefore it is important that the pathologist maintain a high index of suspicion. The histologic appearance of sarcomatous melanoma mimics other spindle cell tumors, therefore immunohistochemical stains play an important role in diagnosis. Since present therapeutic strategies may not necessarily alter the prognosis, an early recognition of this unusual variant of malignant melanoma is a key to prolong the survival.

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