Primary Anorectal Melanoma : A Case Report

Sofiène Sayari, Amir Moussi, Riadh Belhaj Salah, Souad Bakhta Ghérib, Karim Haouet, Abdeljélil Zaouche.

Service de Chirurgie générale A, Hôpital Charles Nicolle, Tunis.

S. Sayari, A. Moussi, R. Belhaj Salah, S. Bakhta Ghérib, K.Haouet, A. Zaouche.

Mélanome anorectal primitif. A propos d'une observation

LA TUNISIE MEDICALE - 2010 ; Vol 88 (n°06) : 430 - 432

S. Sayari, A. Moussi, R. Belhaj Salah, S. Bakhta Ghérib, K.Haouet, A. Zaouche.

Primary Anorectal Melanoma : A Case Report

LA TUNISIE MEDICALE - 2010 ; Vol 88 (n°06) : 430 - 432

RÉSUMÉ

Prérequis : Le mélanome anorectal est une tumeur maligne rare de mauvais pronostic. Les symptômes cliniques ne sont pas spécifiques et le traitement est encore discuté.

But : Le but de cette étude était de rapporter un cas de mélanome anorectal afin de discuter les problèmes de diagnostic et de traitement.

Observation : Un homme âgé de 66 ans se plaignait depuis trois mois avant son hospitalisation de rectorragies intermittentes de faibles abondances et de douleurs anales. Le toucher rectal avait révélé un polype de 2 cm à 3 cm de la marge anale, pédiculé à base d'implantation dure saignant au contact situé à 5 heures en position de la taille. Il a été réalisé une résection endoscopique sous anesthésie générale. L'examen histologique de la pièce de résection avait conclu à un mélanome malin. Les marges de résection étaient saines. Les suites opératoires étaient simples.

Un bilan d'extension tumoral a été réalisé après un mois. Il avait montré un nodule de 15 mm au niveau du poumon gauche. Il a été réalisé une métastasectomie par thoracotomie.

Le patient n'a eu aucun traitement adjuvant. Il est décédé un an après.

Conclusion : A travers ce cas nous avons voulu illustrer les difficultés de diagnostic préopératoire du mélanome malin anorectal à cause de l'absence de symptômes spécifiques. La chirurgie demeure le traitement de base. L'excision locale large associée à la radiothérapie locorégional devrait être préférée si elle est techniquement faisable. L'amputation abdominopérineale n'est indiquée que lorsque la tumeur est volumineuse ou en cas d'envahissement du sphincter anal. La survie à cinq ans est inférieure à 20%. Elle dépend essentiellement du stade évolutif de la maladie.

Mots-clés

Mélanome, anus, rectum

SUMMARY

Background : Anorectal melanoma is a rare but highly lethal malignancy. Clinical symptoms are non-specific and treatment is still debated.

Aim : The aim of this study was to report a case concerning diagnostic and management of Anorectal melanoma.

Case : A 66-year-old man was admitted in our surgical unit with a 3-month history of pain and rectal bleeding. Rectal examination revealed a tender mass arising from the 5 o'clock position of the anal canal that bled on touch. A provisional diagnosis of rectal polyp was made and it was removed by local excision under general anaesthesia. Histopathologic examination reported it as an anorectal malignant melanoma. The postoperative course was uneventful. Extension staging showed a 15 mm nodule on the left lung. The patient underwent a metastasectomy of the left lung. No adjuvant therapy was given. He died one year later.

Conclusion : With this case we want to illustrate that malignant melanoma can be difficult to diagnose, as patients have non-specific symptoms and histology may be misleading. Surgery remains the mainstay of treatment. Wide local excision combined with adjuvant loco-regional radiotherapy should be preferred when technically feasible. Abdominoperineal resection has to be done only in the case of large tumors or when the anal sphincter is involved. Overall 5-year survival is less than 20%. It's correlated to extension of disease regardless of initial surgical therapy.

Key-words

Melanoma, anus, rectum

Primary anorectal melanoma is a rare and aggressive disease accounting for approximately 1% of anorectal malignancies, but it's the most common site of development of melanoma in the alimentary tract (1, 2). It arises from melanocytes present in the transitional zone of the surgical anal canal (3). The clinical features are similar to those encountered in other anorectal lesions which makes difficult to reach the right diagnosis (3). Moreover, a significant proportion of lesions are not pigmented and they can resemble a rectal polyp (3). The nature of the tumor is generally only revealed after histological examination (3). Because of its low incidence, experience in treatment is limited and the optimum surgical procedure is controversial, ranging from radical abdominoperineal resection to wide local excision with or without adjuvant radiotherapy (1, 4). The prognosis of anorectal malignant melanoma is poor with a median survival of 24 months and a 5-year survival of between 10-20% (4).

The aim of this article was to report a case of primary anorectal melanoma and to focus on diagnostic and therapeutic problems of this disease.

CASE REPORT

A 66-year-old man was admitted with a 3-month history of recurrent rectal bleeding and pain. Rectal examination revealed a tender mass at the 5 o'clock position, 3 cm from the anal verge and that bled on touch. The remnant of the physical examination was normal.

Coloscopy revealed two polypoid tumors. The first one was located on the sigmoid colon. Its main line was 15 mm and it had irregular surface surrounded by fragile ulcers. The second polyp was located on the lower rectum. Its main line was also 15 mm and it was covered with a purplish mucous membrane. A provisional diagnosis of colorectal polyps was made.

An endoscopic resection of the first polyp was performed with the diathermic arch. The histological examination of the specimen had shown a tubulo villous adenoma dysplasia of low rank. The axis of the polyp was free from any tumour proliferation. The rectal polyp was removed by local excision under general anaesthesia. A one cm safety margin around the base of the polyp was respected. The histological examination of the resected specimen concluded to malignant melanoma. The margins of resection were healthy. Postoperative recoveries were uneventful. One month after surgery, a tumour staging was realized. Abdominal, pelvic and bone scan were free from any metastases, but thoracic scan revealed a 15 mm nodule on the left lung. The patient underwent a metastasectomy of the left lung. No adjuvant therapy was given. He died one year later.

DISCUSSION

Our reported case was partculary by the diagnostic problem. In fact, histological findings were not evolved before exision. Management is still debated : local excision or abdominoperineal resection ?

Anorectal melanoma is a rare condition accounting for 0.2% to 3% of all melanomas, and 1% of all malignant tumors of the rectum and anus (5). The disease affects all ages, with the highest incidence during the sixth and seventh decade. There is also a slight female preponderance (2, 6). Our patient was a 66 years old man at the time of diagnosis.

Rectal bleeding and pain are the most common presenting symptoms (7). They occur in up to 83% of the cases. They are often considered as symptoms of haemorrhoids or other benign anal lesions. Anorectal mass, tenesmus, and bowel habit changes are other common symptoms. They generally occur when the tumor has a deep penetration and a large size.

The average time from onset of symptoms to diagnosis is four months. Therefore, lesions are discovered at a late stage with metastases among almost 60% of patients in most series (4, 8). Our patient presented himself after three months of pain and rectal bleeding.

The lesion can be pigmented or not. Thirty per cent of the anorectal melanomas are amelanotic. At early stage, they look like a polyp, which makes the diagnosis more difficult even after histological examination. In these cases immunohitochemical studies should always be done. Positive protein S-100, melanoma antigen HMB-45 and Melan-A expressions are strongly suggestive of melanoma (2).

When the lesion is pigmented, it can be taken for a thrombosed haemorrhoid. Indeed, in the Memorial Sloan-Kettering series, 8% had the melanoma discovered on pathological review of hemorrhoidectomy specimens (3).

Surgery remains the mainstay of treatment with two operative options: local excision and abdominoperineal resection (3). Wide local excision is defined as a sphincter saving procedure with 1 to 2 cm side safety margins to the tumor. But there is no consensus at this moment on which surgical approach is preferred (4). This is mainly due to the rarity of this disease and to the difficulty in collecting a consistent number of cases in a homogeneous and rational way. Moreover, the different surgical adopted options have not been directly compared. Thus, optimal surgical treatment for primary tumors is still controversial.

A study had previously suggested that aggressive treatment of the primary anorectal lesion with abdominoperineal resection was associated with improved outcome, with a lower rate of local recurrence possibly due to regional lymphadenectomy (5). However, other studies, have reported that the rate of local recurrence was slightly higher in those who had local excision but the 5-year survival for local excision and abdominoperineal resection did not differ (9, 10).

Two studies showed that sphincter-saving local excision combined with adjuvant loco-regional radiotherapy results in the same loco-regional control with less loss of function compared to abdominoperineal resection (70% vs 74%) (4, 11). Consequently, the current tendency is to consider local excision to be the treatment of choice in case of small tumors, because of lower morbidity and similar overall survival compared to abdominoperineal resection. Only in the case of large tumor or when the anal sphincter is involved, an abdominoperineal

resection has to be done (12-14). The role of adjuvant chemotherapy has not yet been established (15).

The prognosis of anorectal malignant melanoma is poor. Overall 5-year survival is less than 20% (10). This seems to be due to the late detection of the disease at a stage with metastases, and the frequency of recurrences regardless of initial surgical therapy (16).

The most important prognostic factors are the stage of the disease and the tumor thickness. Patients have a relatively good outcome if the tumor thickness is less than 2 mm. The median survival is 34 months for patients with local disease and 10 months for those with metastatic disease.

The presence of tumor necrosis and tumor perineural invasion (PNI) appears to be important histological features associated with poor outcome. In the present series, regardless of surgical approach, all patients who had tumors with either necrosis or PNI recurred with a median recurrence-free survival of 6 months or less (10).

The majority of patients die of distant metastases. At the time of death 58%, 48% and 4% have respectively liver, lung and brain metastases (1).

CONCLUSION

Primary anorectal melanoma is a rare and aggressive disease. Clinical features are similar to those encountered in other anorectal lesions. Surgery remains the mainstay of treatment. A sphincter-saving local excision combined with adjuvant loco-regional radiotherapy should be preferred when technically feasible in case of small tumors.

Abdominoperineal resection must be required only in the case of large tumors or when the anal sphincter is involved. The prognosis of these lesions remains poor. Further study of the molecular mechanisms of anorectal melanoma oncogenesis and tumor progression is needed to develop innovative treatment paradigms that may ultimately impact outcome and improve survival.

Références

1. Belli F, Gallino GF, Lo Vullo S, Mariani L, Poiasina E, Leo E. Melanoma of the anorectal region: the experience of the National Cancer Institute of Milano. Eur J Surg Oncol 2009;35:757-62.
2. Pirenne Y, Bouckaert W, Vangertruyden G. Rectal melanoma. A rare tumour. Acta Chir Belg 2008;108:756-8.
3. Saiprasad BR, Prasad MS, Ravishankar TH, Mathur K. Rectal polyp: can it be a malignant melanoma? Surgery 2006;140:474-5.
4. Van Schaik PM, Ernst MF, Meijer HA, Bosscha K. Melanoma of the rectum: a rare entity. World J Gastroenterol 2008;14:1633-5.
5. Seya T, Tanaka N, Shinji S, Shinji E, Yokoi K, Horiba K, Kanazawa Y, Yamada T, Oaki Y, Tajiri T. Case of rectal malignant melanoma showing immunohistochemical variability in a tumor. J Nippon Med Sch 2007;74:377-81.
6. Ragnarsson-Olding BK, Nilsson PJ, Olding LB, Nilsson BR. Primary ano-rectal malignant melanomas within a population-based national patient series in Sweden during 40 years. Acta Oncol 2009;48:125-31.
7. Kayhan B, Turan N, Ozaslan E, Akdo€an M. A rare entity in the rectum: malignant melanoma. Turk J Gastroenterol 2003;14:273-5.
8. Moozar KL, Wong CS, Couture J. Anorectal malignant melanoma: treatment with surgery or radiation therapy, or both. Can J Surg 2003;46:345-9.
9. Homsi J, Garrett C. Melanoma of the anal canal: a case series. Dis Colon Rectum 2007; 50:1004-10.
10. Yeh JJ, Shia J, Hwu WJ, Busam KJ, Paty PB, Guillem JG, Coit DG, Wong WD, Weiser MR. The role of abdominoperineal resection as surgical therapy for anorectal melanoma. Ann Surg 2006;244:1012-7.
11. Weyandt GH, Eggert AO, Houf M, Raulf F, Bröcker EB, Becker JC. Anorectal melanoma: surgical management guidelines according to tumour thickness. Br J Cancer 2003;89:2019-22.
12. Jai SR, Adraoui J, Khaiz D, Lakhloufi A, Chehab F, Bouzidi A. Mélanome malin anorectal. A propos de trios cas. Tunis Med 2005;83:422-5.
13. Bullard KM, Tuttle TM, Rothenberger DA, Madoff RD, Baxter NN, Finne CO, Spencer MP. Surgical therapy for anorectal melanoma. J Am Coll Surg 2003;196:206-11.
14. David AW, Perakath B. Management of anorectal melanomas: a 10-year review. Trop Gastroenterol 2007;28:76-8.
15. Ishizone S, Koide N, Karasawa F, Akita N, Muranaka F, Uhara H, Miyagawa S. Surgical treatment for anorectal malignant melanoma: report of five cases and review of 79 Japanese cases. Int J Colorectal Dis 2008;23:1257-62.
16. Haddad F, Nadir S, Benkhaldoun L, Alaoui R, Cherkaoui A. Mélanome anorectal primitif. Presse Med 2005;34:85-8.