

One Example of when a Hemorrhoid is not a Hemorrhoid.....Anorectal Melanoma

Very rarely what appears to be a thrombosed external or internal hemorrhoid can turn out to be an anorectal melanoma. Less than one percent of all cancers involving the anorectum are melanomas. Less than 2% of all melanomas are found in the anorectum, yet it is the third most common site after the skin and eyes. Roughly 75% of all of these melanomas are dark in color, but up to 1/3 may actually share the color of the surrounding anoderm or mucosa. Melanoma has a very poor prognosis as far as cancers go, except for when it is very superficial. Unfortunately, the disease is usually quite advanced when it is finally identified in the anorectum. As a result, the 5-year disease free survival is typically 0-22% depending on the study that you read.

Since this disease is so rare, very little data is available to help guide management. There are no randomized, comparative trials and what does exist are mostly retrospective reviews or institutional case series with small numbers of patients and limited duration of follow up.

The management for this devastating oncologic diagnosis primarily involves surgery. Much debate has developed regarding the optimal surgical treatment. The two choices include abdominoperineal resection (APR) involving an inherent permanent colostomy and wide local excision (WLE) with sphincter preservation. The rationale between the two recommendations is as follows:

Those who support APR point to data from institutions such as Memorial Sloan Kettering Cancer Center in New York where a 64 year experience with this diagnosis yielded only 10 long term survivors out of a total of 85 patients. Nine out of ten of these patients underwent an APR and only one had a WLE.

Physicians at the University of Virginia found that 69 percent of the patients who underwent APR, had mesenteric lymph node metastases in the mesorectal portion of the resected specimen. WLE would not have removed this tissue and hence, eventual mortality would be essentially guaranteed in those patients. They would also point out that to achieve a good margin for all but the most superficial of lesions would require sacrifice of a substantial portion of the underlying sphincter muscles, which might lead to significant fecal incontinence and reduce quality of life.

Those supporting wide local excision would argue that as many as 70% of patients with anorectal melanoma who recur, will do so at a distant site regardless of whether they undergo APR or WLE, as has been shown in a number of studies. Plus, although local recurrence in most studies is almost twice as likely in the WLE group, a statistically significant long term or disease free survival has never been documented for APR. Finally, for a patient population in which there is such a low survival rate, seemingly regardless of treatment algorithm, does it make sense to subject the patient to the morbidity (close to 40% in some series) of an APR? They would suggest that it does not. The biggest problem with all of this is the data itself. How was it decided in each case series and each institutional review which patients were selected for APR and which underwent WLE? Upon review of these papers, this information is often unclear or not present at all. The potential selection bias, as a result, greatly reduces the potential to draw a strong conclusion as to which is the superior or more appropriate treatment.

Unfortunately, little evidence suggests that adjuvant therapy is of much help for this disease process. Adjuvant treatments used have included chemotherapy (dacarbazine, vincristine, nimustine, BCG, levamisole, and interferon). None of these have shown a statistically significant survival benefit in cutaneous melanoma data. Response rates are at best 20%.

Sentinel lymph node mapping, which has become standard of care in cutaneous melanoma due to its survival benefit, has been used in anorectal melanoma, but the numbers are too small to reliably extrapolate the same conclusion. Studies that have looked at empiric lymph node dissection of the groin have shown that survival did not improve and morbidity was substantial. Therefore, empiric groin dissection is not recommended unless a palpable node is present or if a PET scan suggests there may be an involved node. Interferon alpha has been used for cutaneous melanoma and some benefit may exist, but the side effects are substantial. One case study did actually show a 2.5 year unexpected survival and resolution of a pulmonary metastasis while on the Interferon.

Radiation therapy has been shown to improve locoregional control in cutaneous melanoma in the head, neck, axilla, and vagina. Again, data for the anorectum is lacking due to limited numbers. One study involved 23 patients undergoing WLE with radiation therapy, using 30 Gy over 2.5 weeks, showed a 5 year local control rate of 74%, which compares with the local control for APR in most studies. However, this data is potentially tainted by the fact that all patients were not treated similarly. Specifically, nine of those patients underwent concurrent chemotherapy and some did have an inguinal lymphadenectomy.

In determining a recommendation for treatment at Colon and Rectal Care, Inc., we view this as a patient specific process. For example, if there is any evidence of metastatic disease at presentation, any role for an APR except in the case of palliation for a very large lesion simply would not exist, particularly given the high recurrence rate. However, a very small lesion,

incidentally identified in a young patient with very little co-morbidity, would at least call for a discussion regarding a possible APR. Although the available literature does not support this (possibly due to the lack of available data), in theory, one would think that there should be some patient population with very early regional lymph node metastasis to the mesorectum and no other distant disease that might be treated with APR, but missed with WLE. Certainly, the biologic behavior of this disease probably limits this likelihood or else a significant difference in disease free survival would have already been shown between the two treatment modalities. Nonetheless, the patient deserves to make the choice, understanding that any benefit to an APR would be theoretical only given what information we do have.

As far as the role for adjuvant treatments, we feel that a medical oncologic evaluation is appropriate, especially given the possibility of future therapies maybe showing some improved efficacy. The role for sentinel lymph node mapping needs to be determined on a case by case basis, especially since there will likely never be a study with an adequate number of cases to establish its efficacy.

Since 5-year survival is 0-22% in this disease, certainly quality of life is of paramount concern and should be emphasized. *Dis Colon Rectum* 1995;38(2):146-151. *Dis Colon Rectum* 1997;40(6):661-668. *Clinics in Colon and Rectal Surgery* 2006;19(2): 78-87. *Melanoma Res* 2002;12:395-98. *Dis Colon Rectum* 2002;45(10):1412-17. *Dis Colon Rectum* 1982;25:693-703. *Dis Colon Rectum* 1999;42(9):1203-8. *Dis Colon Rectum* 2007;50(7):1004-10. *J Clin Oncol* 2002;89:4555-58. *Am Surg* 2006; 72(10):917-20. *Discomfort Colon Rectum* 2007;50(7):1004-10. Summarized by Joseph C. Muller, MD.

This newsletter is produced by Doctors Rama M. Jager, Shekar Narayanan, Joseph C. Muller and Arun Gowdamarajan, specialists in Colon and Rectal Surgery. Our practice includes an onsite anorectal floor lab and ambulatory surgery center. Comments or requests to subscribe can be e-mailed to info@colonandrectalcare.com.

