

LOS ANGELES

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Unusual Malignant Tumors of the Anus

The Good News? These are Rare Lesions.

PERIANAL TUMORS:

Paget's Disease Perianal Paget's disease is thought to be a neoplasm arising from the apocrine glands of the perianal skin. Unlike its mammary counterpart, perianal Paget's disease may begin as a benign condition in the apocrine glands, and later transform into an invasive adenocarcinoma, spreading into the epidermis. Another theory holds that the disease begins as an intraepithelial adenocarcinoma with a lengthy preinvasive phase, which subsequently spreads to the deeper dermis.

Anal Paget's disease is exceedingly rare, with less than 200 reported cases. It affects people in the seventh decade of life and affects men and women equally.

Pruritus is a common presenting symptom. Other presenting symptoms include an erythematous perianal skin rash with eczema, oozing or scaling, weight loss, inguinal adenopathy or constipation. The diagnosis is often delayed because of the tumor's similarity to many other benign anal skin conditions and because of physician unfamiliarity with the disease. The presenting lesion may be circumferential and extend proximally to the dentate line. The average duration of symptoms prior to diagnosis is 3 years. Anal Paget's disease may present in up to 40% of cases with an associated invasive adenocarcinoma.

A thorough anorectal examination must be performed looking for an associated carcinoma requiring more extensive treatment. A full colon evaluation and a search for visceral carcinomatosis should be conducted. Visceral spread occurs in up to 50% of cases. Common sites of spread include the GI tract,

prostate, neck and nasopharynx.

A biopsy of the affected area will confirm the diagnosis.

TREATMENT OPTONS: For patients who do not have an invasive cancer, a wide local excision is the treatment of choice. With a non-invasive tumor, survival may approach 100%.

Patients initially presenting with invasive disease may be candidates for an abdominoperineal resection (APR), with an added inguinal lymphadenectomy if documented nodal disease is present.

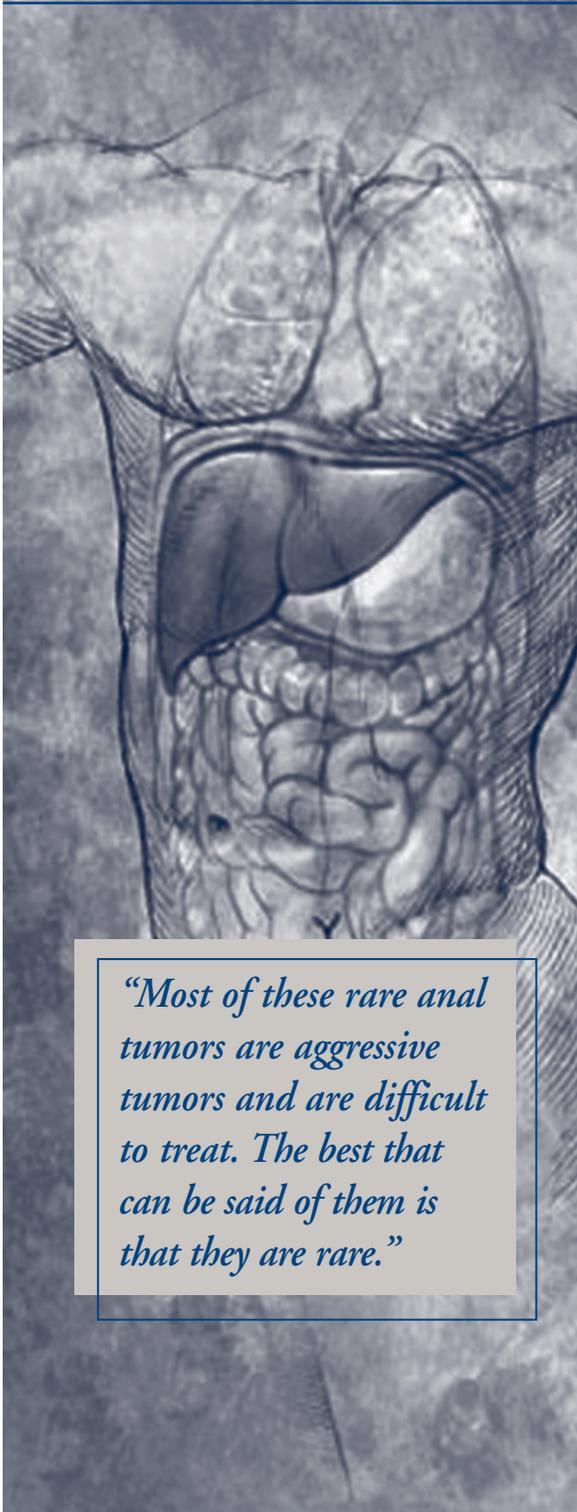
Recurrence rates are reported ranging from 37% to 100% and most recurrences can be treated with repeat wide excision.

Basal Cell Carcinoma Even rarer is the perianal basal cell carcinoma. Occurring in the seventh and eight decades of life, more commonly in men than women, the lesion looks like other cutaneous basal cell cancers with raised, rolled edges and a central ulceration. They rarely metastasize. Up to one-third have a history of a basal cell cancer at another bodily location. Lesions are usually smaller than 2 cm. They may also present as papules, plaques, nodules or ulcers.

TREATMENT OPTONS: They are superficial and mobile making wide excision straightforward. As they are of a low invasive potential, treatment is gratifying. Five year survival with adequate excision or re-excision for recurrences is 100%.

Verrucous Carcinoma Associated with

continued on back...



"Most of these rare anal tumors are aggressive tumors and are difficult to treat. The best that can be said of them is that they are rare."

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HPV-6 and 11, verrucous carcinoma or Buschke-Lowenstein tumor is a large, soft, fleshy cauliflower-like cancer that is painful and slow growing. It affects men almost three times more often than women in the fifth decade of life. Symptoms include pain, abscesses, pruritus, bleeding, a malodorous smell and altered bowel habits. Located on the perianal skin or in the distal anal canal or rectum, these lesions are relentless in their growth. Although benign when viewed under the microscope, they must be considered malignant in their behavior, with the potential for local erosion or invasion into the ischioanal fossa and perirectal tissue. CT scanning will define the extent of invasion. Microscopically, verrucous carcinoma looks like condyloma acuminata. It does not metastasize.

TREATMENT OPTIONS: Wide local excision is the treatment of choice, with APR performed in unusual cases of late-stage disease with sphincter invasion. Radical excision may be necessary in order to achieve a cure. Chemotherapy has been shown to be of little benefit in treating these lesions.

ANAL CANAL TUMORS:

Adenocarcinoma Anal adenocarcinoma accounts for 10% of all anal cancers. The three types of anal adenocarcinomas are those arising in the anorectum, those arising in anal glands, and those arising in an anorectal fistula.

Because there are so few reported cases of anal adenocarcinoma, uniform treatment recommendations based on solid data are difficult to obtain. It is clear that treatment results are relatively poor and this disease seems to be much more aggressive than its anal squamous cell counterpart when matched for local recurrence rates and survival rates.

TREATMENT OPTIONS: Treatment regimens have included wide local excision (WLE), abdominoperineal resection, radiation, chemoradiation, and combined modality therapy, usually pre- or post-operative chemoradiation and an abdominoperineal resection. The five year survival and disease-free survival rates have been found to be best in the chemoradiation group, with five and ten year overall survival rates of 58% and 39% respectively.

Melanoma Anal melanoma is an ominous tumor. Fortunately, it is also a rare tumor. Fewer than 500 cases have been reported and it constitutes between 0.5% and 5% of all anal malignancies. Melanoma of the anal canal is the third most common site of occurrence after skin and ocular melanoma. Those with anal melanoma are often white females with an average age of 63 years old. The disease is rarely reported in the African-American or Asian populations. Women are twice as likely as men to have this lesion.

The lesion may arise at any location in the anal canal. Bleeding is the usual patient complaint, followed by pain. Altered bowel habits and tenesmus are also reported. Weight loss is common. The most common presenting sign is an anal mass. Seventy five percent of patients have a mass greater than 1 cm in size, and the average diameter is 4 cm. Many patients have associated involved inguinal lymph nodes.

Anal melanoma is often confused with a thrombosed hemorrhoid. Larger lesions may be polypoid and ulcerated distinguishing the lesion from a thrombotic hemorrhoid. In one study, 71% of patients had "gross and/or histologic pigmentation," while other studies have found these lesions to be amelanotic in 41% of cases.

Between 38% and 62% of patients presented initially with metastatic

disease to lymph nodes, and not uncommonly to distant sites such as liver and lung.

In a review of the Mayo Clinic experience, no single factor predicted survival. Factors evaluated were "gender, size of the lesion, presence of melanin, depth of penetration, positive perirectal lymph nodes and wide local excision versus APR". Interestingly, the Memorial Sloan-Kettering series found that the only long-term survivors were women.

TREATMENT OPTIONS: Although there are conflicting issues in trying to choose a treatment modality and predict survival rates using various parameters, one point is clear; five year survival rates are dismal. Survival rates range between 0% and 29%. One study showed that in patients whose tumors are thicker than 10 mm, cure is not possible.

Using wide local excision or abdominoperineal resection, overall five year survival rates have been found to be 17% and 22% respectively. Most authors note that patients with documented regional metastatic disease or distant metastases should be spared an APR and permanent colostomy, as long-term survival rates are almost non-existent. However, in those with bulky tumors, or in patients in whom the surgeon is unable to obtain 1 to 2 cm tumor-free margins with a wide local excision, or in patients with tumor involvement of the anal sphincters, or in those patients who would be rendered incontinent after a wide local excision, an APR may be the treatment of choice.

Numerous immunologic and adjuvant chemotherapeutic treatments have been tried with little benefit in patients with anal melanoma. Radiation therapy is of unproven benefit as well. Local control, while possible with surgical treatment, is often useless, as distant metastatic disease is a major cause of death.

Neuroendocrine Carcinomas Also called a small cell carcinoma or a large cell neuroendocrine tumor, or a Merkel cell carcinoma, this tumor is so named because it is derived from cells of both the endocrine system and the nervous system. Neuroendocrine cells are found in many organ systems and tissues, with the gastrointestinal tract harboring the largest volume of these cells. These tumors are rare, comprising less than 1% of all lower digestive tract cancers. Most neuroendocrine tumors are found in the rectum and cecum.

Colorectal neuroendocrine tumors are classified as either low-grade carcinoids or high-grade neuroendocrine carcinomas. The neuroendocrine lesions can be further subdivided into small cell carcinomas or large cell neuroendocrine carcinomas.

Diagnosis involves a high index of suspicion in patients with any type of anorectal complaint. An obvious lesion removed surgically will then be submitted for the appropriate microscopic and immunohistochemical evaluation.

TREATMENT OPTIONS: As these tumors are rare, no single series can reliably evaluate the different modes of available treatment and survival statistics. Patients have been treated with excision, radical extirpative procedures with or without adjuvant therapy, radiation alone or chemotherapy alone.

Although not validated, it would make sense to treat patients with disease limited to the anal canal with extirpative surgery and/or chemoradiation, akin to the treatment for anal adenocarcinoma.

Most of these rare anal tumors are aggressive tumors and are difficult to treat. The best that can be said of them is that they are rare.