Basal Cell Carcinoma of the Umbilicus: A Case Report and Literature Review

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GOAL

To gain a thorough understanding of basal cell carcinoma (BCC) of the umbilicus

OBJECTIVES

Upon completion of this activity, dermatologists and general practitioners should be able to:

1. Discuss the unique characteristics of BCC of the umbilicus.
2. Identify patients who may have a higher risk of developing BCC of the umbilicus.
3. Recognize the histology of BCC of the umbilicus.

CME Test on page 138.

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We report the case of a 43-year-old woman with basal cell carcinoma (BCC) of the umbilicus. Although BCC is a common skin tumor, only 2 cases of BCC arising within the umbilicus have been reported previously. Our review of the literature shows that truncal BCCs frequently develop in younger patients, often grow larger, and are associated with an increased risk for developing multiple nonmelanoma skin cancers. Therefore, we advocate a low threshold for performing biopsies on umbilical lesions that are atypical in appearance, course, or response to therapy.

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The umbilicus is an exceedingly atypical site for basal cell carcinoma (BCC). Although tumors on the anterior and posterior areas of the trunk are not infrequently noted, the English-language literature includes only 2 reports of cases of BCC arising specifically in the umbilicus. In this third case report, we describe a woman with umbilical BCC and briefly review other reports in the literature, as well as factors unique to truncal and umbilical BCC.

Case Report

A 43-year-old woman presented to our dermatologic surgery unit with a 20-year history of a slow-growing red scaly plaque at the umbilicus. Previously, a clinical diagnosis of eczematous dermatitis had been made by another physician, and the patient had been treated with topical corticosteroids and topical antibiotics without effect. After the lesion had begun bleeding spontaneously, a biopsy had been performed, and a histopathologic diagnosis of superficial BCC had been made. When the patient reported to our dermatologic surgery unit, the plaque—thin, 2.7×1.5 cm in size, erythematous, and scaling—extended inferiorly from within the umbilicus (Figure 1). Although the patient had fair skin and admitted to having used a tanning bed 3 times several years earlier, she denied any other risk factors for developing BCC (eg, exposure to significant amounts of sunlight, arsenic, radiation, chemical carcinogens). In addition, she denied having any burn scars, ulcers, or chronic trauma to or irritation of the umbilicus. The patient's family had no history of nonmelanoma skin cancer. However, the patient reported that she had simultaneously developed a similar tumor on the left forearm and that her dermatologist had diagnosed the tumor as BCC and had treated it with electrodesiccation and curettage.

Because the umbilical tumor extended deep into the umbilicus and had poorly defined margins, Mohs micrographic surgery was considered appropriate therapy. A tumor-free plane was achieved after 2 stages. Fresh-frozen sections of extirpated tissue showed superficial BCC (Figure 2). The wound was reconstructed with a purse-string closure.

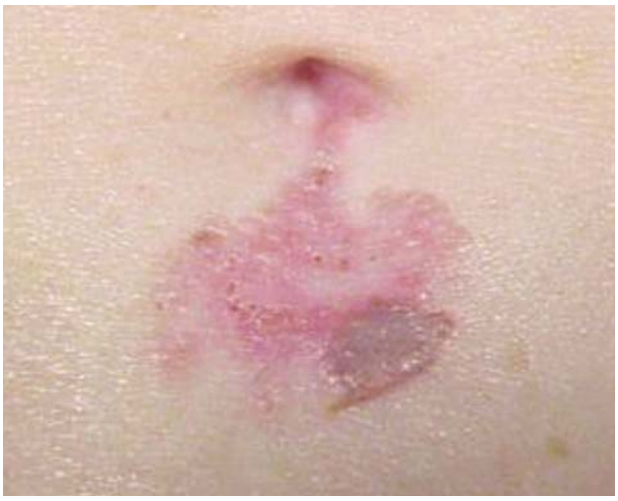

Figure 1. Basal cell carcinoma of the umbilicus presenting as an erythematous scaling plaque.

Comment

The large majority of BCCs arise on sun-exposed skin, most commonly on the head and neck.[1] When BCCs develop on sites that are relatively protected from exposure to UV radiation, often other factors (or combinations of factors) are involved—including exposure to ionizing radiation and arsenic, as well as a history of trauma to or irritation of the site.[2,3] More recently, associations between BCCs and exposure to asbestos, fiberglass dust, dry-cleaning solvents, and luminous paint have been reported.[4] In addition, BCCs may arise in burn scars, vaccination scars, adnexal hamartomas, and areas of chronic inflammation (eg, stasis dermatitis, stasis ulcers, hidradenitis suppurativa).[2,3] Last, BCC can develop in several genodermatoses, including basal cell nevus syndrome, Bazex syndrome, Rombo syndrome, Rasmussen syndrome, xeroderma pigmentosum, and oculocutaneous albinism.[2,3,5]

Unusual sites for BCC have included the breast, axilla, inguinal region, genitalia, periungual region, palm, sole, buttock, and hair-bearing scalp.[2,6-8] Among the multiple English-language reports of unusual sites for BCC and reviews of umbilical lesions, however, only 2 reports of BCCs arising in the umbilicus were found. Steck and Helwig[9] reported both BCCs in their review of umbilical tumors; one BCC was associated with an epithelial inclusion cyst, and the other was heavily pigmented and multicentric. Given that a number of extensive reviews of truncal BCCs did not include details regarding the tumor sites, other cases of umbilical BCCs may exist but may not have been reported as such.

The skin of the umbilicus does not seem particularly unusual. Except for the cicatrix itself, the umbilical adnexae are similar in density to the rest of the anterior trunk.[9] The walls and posterior aspect of the umbilicus are protected from UV exposure even more than the abdomen, which is typically covered by clothing, so fewer sun-induced tumors are expected to develop within the umbilicus than on the anterior trunk.

Some characteristics of truncal BCCs, however, seem unique. The incidence of BCC on the trunk is lower than would be expected based on surface area, and much of the difference is attributed to lack of sun exposure.[10] Interestingly, patients presenting with BCC on the trunk are typically younger than patients presenting with BCC on other sites.[11,12] Furthermore, truncal BCCs seem to develop more often in men than in women.[12,13] Perhaps because the trunk is of relatively minor cosmetic importance and thus is subject to inattention and

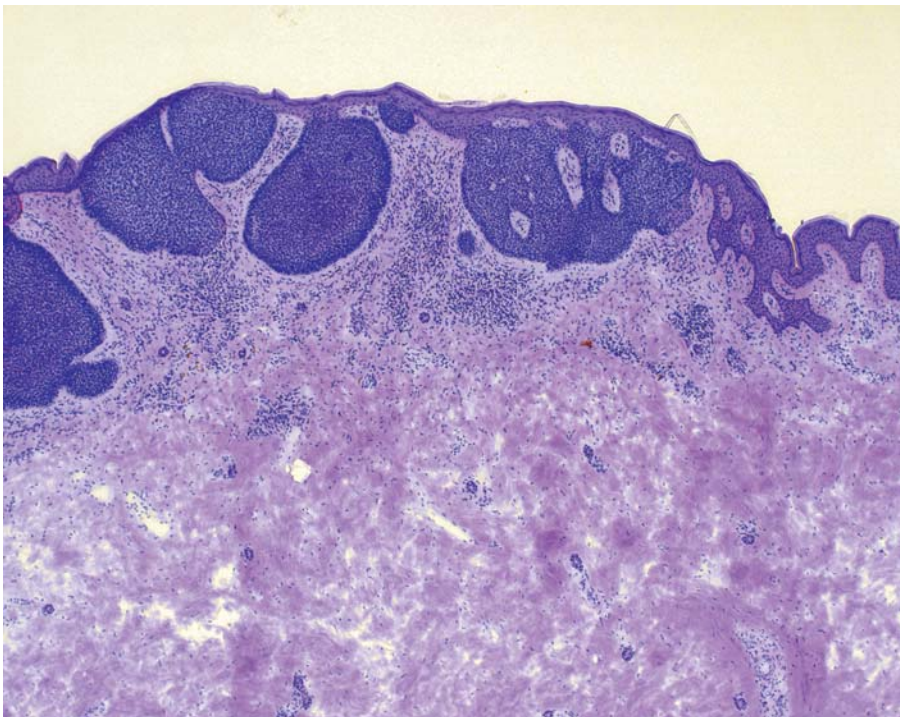

Figure 2. Basal cell carcinoma with typical peripheral palisading and basophilia (H&E, frozen section, original magnification ×80).

neglect, truncal BCCs are typically larger than BCCs on the head and neck.[3] In addition, research suggests that patients with truncal BCCs are at higher risk for developing multiple nonmelanoma skin cancers than are patients with BCCs on other areas of the body.[14,15] This increased risk has been attributed to genetic susceptibility; patients may inherit polymorphisms that result in phenotypes less able to detoxify the products of UV-induced oxidative damage in the skin.[12,14]

Even the histology of truncal BCCs differs from that of tumors on other sites. Superficial BCCs develop significantly more frequently on sun-protected skin[16]; 48.5% of BCCs on the trunk, compared with 22.7% of BCCs on the head and neck, are of the superficial subtype.[13] Indeed, only 14.8% of BCCs are superficial, but 48% of these develop on the trunk.[13]

Although truncal BCCs seem to have some distinctive characteristics, reports of umbilical BCCs are too few to permit the drawing of conclusions regarding the natural history of these tumors. The exceptional anatomy of the umbilicus, however, warrants consideration. All 3 sets of abdominal wall lymphatics drain away from the umbilicus[9] and theoretically may carry tumor to superficial and deep axillary, inguinal, deep femoral, and periaortic lymph nodes.[17] Therefore, the extremely low metastatic potential of BCC may be increased slightly when it develops within the umbilicus, in close proximity to various anatomic structures that may facilitate tumor spread.

The paucity of reported cases of umbilical BCC also prohibits the characterization of its classic clinical features. The presentation of umbilical BCC may mimic relatively benign inflammatory processes such as psoriasis, chronic eczema, allergic contact dermatitis, seborrheic dermatitis, and herpes gestationis.[18] If the lesion is nodular, it may represent a benign tumor such as a seborrheic keratosis, nevus, cyst, pyogenic granuloma, congenital malformation, or endometriosis, to name a few.[18,19] Primary malignant tumors of the umbilicus are rare and include not only BCC but also adenocarcinoma of the urachal remnants, malignant melanoma, and myosarcoma.[9] More common but still rare are cancers metastatic to the umbilicus (so-called Sister Mary Joseph nodules), most often from the gastrointestinal tract.[9] Barrow[19] reviewed 677 cases of umbilical tumors and found that 32% represented endometriosis, 38% were primary umbilical lesions (80% benign, 20% malignant), and 30% were metastatic nodules. Thus, an umbilical lesion has a wide differential diagnosis.

Authors of several articles in the plastic surgery literature have proposed techniques for reconstructing the umbilicus after its surgical or traumatic removal. These techniques include local flaps,[20] bilateral advancement flaps,[21] double V-Y procedures,[22] circumferential rotation flaps,[23] and local

flaps combined with conchal cartilage composite grafts.[24] We believe that, in the case of our patient, using a simple purse-string closure helped us prevent scarring and distortion (outside the cosmetic unit of the umbilicus) that occur with use of the more complex techniques mentioned.

Given our uncertainty about the natural history of umbilical BCCs, we are wary of drawing conclusions concerning their behavior. However, because truncal BCCs frequently develop in younger patients, often grow larger, and are associated with an increased risk for developing multiple nonmelanoma skin cancers, we advocate a thoughtful approach to treating BCCs of the umbilicus and emphasize the need to perform biopsies on umbilical lesions that are atypical in appearance, course, or response to therapy. In addition, more case reports of umbilical BCCs may help us elucidate the pathogenesis, diagnostic and prognostic criteria, and therapeutic options of these tumors.

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