Becker Nevus With an Underlying Desmoid Tumor

A Case Report and Review Including Mayo Clinic’s Experience

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**Background:** Becker nevus is a nevoid melanosis, referred to as Becker nevus syndrome when it is associated with other anomalies. Our objectives were to report the occurrence of a Becker nevus with an underlying desmoid soft-tissue tumor; to review Mayo Clinic’s experience with Becker nevi, concentrating on Becker nevi associated with bone, vascular, neural, and other soft-tissue abnormalities; to inform physicians of the Becker nevus syndrome; and finally to alert clinicians to evaluate a Becker nevus with its associations in mind.

**Observations:** A 46-year-old woman had a Becker nevus with an underlying desmoid-type fibromatosis (desmoid tumor) presenting clinically as a “painful dimple” within the nevus. Review of medical records for 1997 through 2006 at Mayo Clinic, Rochester, Minnesota, yielded 52 patients with Becker nevi, 12 of whom had an associated bone, vascular, neural, congenital, or other soft-tissue abnormality, ranging from liposarcoma to an accessory areola.

**Conclusions:** We add to the literature a unique case of desmoid-type fibromatosis immediately beneath a Becker melanosis, which presented as a painful dimple. We hope to raise awareness that a Becker nevus may be associated with other abnormalities, including an infiltrative soft-tissue tumor. We also emphasize the importance of follow-up, including inspection of not only the surface but also the deep tissues underlying the Becker nevus.

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**BECKER NEVUS IS A SHARPLY marginated and typically unilateral nevoid melanosis commonly arising on the upper extremities or thorax. Although most Becker nevi are isolated findings, they have been associated with various skeletal and other abnormalities included under the designation “Becker nevus syndrome.” Becker nevi do not pursue a malignant course but may become cosmetically problematic. During adolescence, Becker nevi may become more prominent with increased hair growth.**

Desmoid-type fibromatosis (desmoid tumor) is a nonmetastasizing but locally aggressive soft-tissue tumor that occurs both extra-abdominally and intra-abdominally. There is an increased occurrence of desmoid tumors in patients with familial adenomatous polyposis (FAP).

Herein, we describe a 46-year-old patient with a known Becker nevus that became dimpled and painful. Immediately underneath the Becker nevus was a desmoid tumor that was responsible for the cutaneous dimpling. To our knowledge, case reports and reviews in the literature have not identified a similar association. We reviewed Mayo Clinic’s experience with Becker nevi from 1997 through 2006.

**REPORT OF A CASE**

A 46-year-old woman with a known Becker nevus involving the left upper arm and shoulder was referred to the Department of Dermatology at Mayo Clinic, Rochester, Minnesota, in December 2006 when additional associated signs and symptoms of burning pain developed underneath the nevus. The patient reported the existence of her Becker nevus since at least adolescence. The underlying pain was accentuated by pressure, touch, and movement. The patient otherwise felt well. Interestingly, she maintained that the area had “always felt uncomfortable,” but the symptoms amounted to only a mild nuisance. The apparent change in the quality and magnitude of her symptoms during the preceding 2 months prompted the referral. Her medical and surgical history...
Our patient, a middle-aged woman, demonstrated the occurrence of a desmoid tumor beneath a Becker nevus. The patient’s symptoms, as well as the cutaneous dimpling, were secondary to the localized infiltration of the underlying desmoid tumor. In our retrospective case review, a notable percentage of patients (23%) with confirmed Becker nevi had an associated bone, vascular, neural, or other soft-tissue abnormality. These abnormalities, which may be incidental, included malignant neoplasms (lower extremity liposarcoma) as well as congenital anomalies (extra areola; absence of a testicle). One such neoplasm arose close to the patient’s Becker nevus (dermatofibrosarcoma protuberans).

In 1949, Becker\(^1\) described hyperpigmented, sharply marginated, unilateral macules with hypertrichosis located on the shoulders of 2 young men. Today, this is generally referred to as Becker nevus, but has also been termed Becker melanosis, pigmented hairy epidermal nevus, and melanosis neviformis of Becker.\(^2,3\) Becker nevus usually occurs on the thorax or upper extremities, although there have been reports of lower extremity manifestations.\(^4\) In 1995, Happle\(^5\) defined the pigmented hairy epidermal nevus syndrome to include Becker nevus, ipsilateral hypoplasia of the breast, and skeletal defects such as scoliosis and indicated the syndrome may be explained by paradigmatic inheritance. Happle and Koopman\(^6\) in 1997 proposed the new term Becker nevus syndrome for a phenotype characterized by the presence of a particular type of organoid epithelial nevus showing hyperpigmentation, increased hairiness, and hamartomatous augmentation of smooth muscle fibers, and other developmental defects such as ipsilateral hypoplasia of...
the breast and skeletal anomalies including scoliosis, spinabifida occulta, or ipsilateral hypoplasia of a limb. The associated anomalies tend to show a definite regional correspondence, suggesting a common origin from an early postzygotic mutation.

In 1998, Urbani and Betti suggested supernumerary nipple and urogenital anomalies in association with Becker nevus. In response, Happle and Koopman included supernumerary nipples and genital anomalies as part of the Becker nevus syndrome. In 2007, Sugarman, in reviewing the epidermal nevus syndromes, indicated that Becker nevi have been associated with ipsilateral hypoplasia of the breast, hypoplasia of underlying musculature, lipatrophy, and underlying skeletal anomalies, including scoliosis, hemivertebrae, fused or accessory cervical ribs, pectus excavatum or carinatum, and internal tibial torsion.

Happle in 2004 described 5 pigmentary patterns involving mosaic epigenetic and genomic etiologies. According to Happle, the syndrome, which occurs sporadically, has a greater frequency in females, and they may not have hypertrichosis. Haneke described proliferation of the erector pili muscles in the dermis of a Becker nevus that resembled smooth muscle hamartoma, and this feature has caused some to consider Becker nevus and smooth muscle hyperplasia to be the 2 ends—hyperpigmentation and smooth muscle proliferation—of a continuum representing the same disorder. Thus, not every case of Becker nevus syndrome has to manifest every association that has been reported, but the presence of an associated manifestation does raise awareness to consider other anomalies that may indicate mosaic mutations involving cellular lines other than epidermal lines.

Histologically, a Becker nevus demonstrates a hyperplastic epidermis with increased pigmentation of the basal keratinocytes. There may be elongation or “clubbing” of the rete ridges. The dermis may also show increased hair follicles and/or smooth muscle bundles that are unassociated with the surrounding adnexa. Although the epidermis may appear to show a focal increase in melanocytes, there are no nests.

Desmoid-type fibromatoses are synonymous with aggressive fibromatoses and desmoid tumors. These clonal tumors may arise both intra-abdominally and extra-abdominally and may be sporadic or associated with FAP, a hereditary tumor syndrome resulting from germline mutation in the APC tumor suppressor gene. The APC protein is involved in the regulation and degradation of β-
catenin, and mutations in the APC gene allow for defective protein function, resulting in the translocation and accumulation of β-catenin within the cell nucleus, which in turn leads to cell proliferation.15,16 This defect is believed to account for an increased risk of desmoid tumors in patients with FAP.

Sporadic extra-abdominal desmoid-type fibromatoses arise from the connective tissue of muscle and the overlying fascia, although the actual cell of origin is not known.17 They typically present between puberty and 40 years of age, with women affected more than men. They most often involve the shoulder, chest wall, and back. Although these desmoid-type fibromatoses are histologically similar to desmoid tumors occurring in patients with FAP, the mechanism of β-catenin dysregulation is different and involves mutations of the β-catenin gene rather than loss of function of APC.18 The clinicopathologic differential diagnosis of extra-abdominal desmoid-type fibromatoses should include leiomyoma, a low-grade sarcoma, and reactive spindle-cell proliferations (ie, nodular fasciitis).19 In our case, a more focused differential diagnosis would include another bland spindle cell neoplasm, such as a leiomyoma. Both tumors can be positive for both smooth muscle actin and desmin.20 However, the lack of tumor circumscription should alert the pathologist to a diagnosis of desmoid-type fibromatosis. In difficult-to-distinguish cases, desmoid tumors demonstrate nuclear positivity on immunostains for β-catenin,21 which reflects the importance of β-catenin in this tumor's pathogenesis.22

Becker nevi have been reported to have an increased amount of androgen receptors, which may explain its overall male predominance as well as the phenomenon of increased hypertrichosis during pubertal years.23,24 Hypertrichosis is evident in 50% of cases and is less evident or absent in females. Interestingly, occasional desmoid tumors have demonstrated immunoreactivity for androgen receptors; in 1 study, positive immunostaining was limited to the extra-abdominal desmoids rather than the intra-abdominal ones.25 As mentioned, our patient’s desmoid tumor was negative for androgen receptor staining.

We propose the following for clarification of terminology pertaining to the Becker disorders:

1. Becker melanosis: We suggest this designation be restricted to pigmented macular changes. This term would apply to hyperpigmentation in the regions identified by Becker1 and by Happle10 in his discussion of mosaic patterns. This term would include, but not be limited to, prepubertal hyperpigmented macules.

2. Becker nevus: We suggest this term be applied to cases with clinical macular pigmentation and associated contiguous adnexal changes involving follicular components (hypertrichosis), smooth muscle hamartomas, desmoid tumors, dermal and subcutaneous alterations, and epidermal tumors. This category would include the case described in this report.

3. Becker nevus syndrome: We suggest this term be used when, in addition to the above 2 categories (ie, Becker melanosis and Becker nevus), associated abnormalities identified by Happle and Koopman6 and by Sugarman26 are also found. This category would include several cases in our Mayo Clinic review.

CONCLUSIONS

Reassurance and monitoring the patient for both intraincidence and underlying changes are appropriate. Given the propensity for a Becker nevus to be associated with other abnormalities, the clinician should always inquire if the patient has noticed changes in his or her Becker nevus and then examine the nevus to detect additional abnormalities, especially if there is either pain or restricted motion.

Becker nevus is a nevoid melanosis, and it can be associated with other anomalies for which the term Becker nevus syndrome is used. We add to the literature the association of desmoid-type fibromatosis beneath a Becker nevus, which presented as a painful dimple. We hope to raise clinical and pathologic awareness that a Becker nevus may be associated with other abnormalities.

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REFERENCES


**Correction**

Errors in Byline, Text, and Data. In the Evidence-Based Dermatology: Review article titled, “Retrospective Analysis of the Association Between Demodex Infestation and Rosacea,” by Zhao et al, published in the August issue of the *Archives* (2010;146[8]:896-902), several errors occurred. In the byline on page 896 and in the correspondence in the right-hand column on page 901, the first author’s name should have read as follows: Ya E Zhao, MPH. On page 897, right-hand column, “Sensitivity Analysis” subsection of the “Methods” section, the sentence should have read as follows: “Four approaches were applied to identify the sensitivity of the studies: we compared the pooled effect sizes (1) with different statistical models; (2) of Chinese- and English-language articles; (3) before and after excluding studies of small sample size; and (4) of studies using the cellophane tape method, skin pressurization, and skin surface biopsy.” Beginning with the third sentence of the “Sensitivity Analysis” subsection of the “Results” section, the text should have read as follows: “Twenty-eight of 48 articles included had a sample size smaller than 40 (OR, 6.96; 95% CI, 3.69-13.15). After these articles were excluded, the pooled OR of the remaining ones was 8.42 (95% CI, 5.49-12.90) (Figure 4), which was not significantly different from that of 7.57 (5.39-10.62) in the pre-exclusion meta-analysis. The ORs of studies using the cellophane tape method9,13-15,18,20,21,23-26,28,30-32,34-36,39,40,42,49 (OR, 9.29; 95% CI, 5.60-15.40), skin pressurization technique10,22,37,38,43-48,50-56 (5.24; 2.85-9.63), and skin surface biopsy3,6,7,9,14,19,27,29,33,41 (8.87; 3.28-24.01) were coincident.”