Extra-abdominal Periosteal Desmoid Tumor of the Third Toe

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Abstract

Extra-abdominal periosteal desmoid tumors are uncommon nonmetastatic tumors of the extremities with a propensity for local recurrence. Lesions in the distal extremities are rare; a majority of extra-abdominal lesions occur in more proximal portions of the upper and lower extremities. This article reports a patient with an extra-abdominal periosteal desmoid tumor in the toe.

A 37-year-old woman had a mass in her left third distal phalanx that was originally noted 3 years prior to presenting to the authors’ institution. She reported the mass expanded during pregnancy. The toe was red and elongated and had expanded to approximately the same size as her great toe. The plantar aspect of the toe was thick and callused, and the toenail was slightly elevated. Marginal excision with retention of the nail was performed without complication. The mass was determined to be an extra-abdominal periosteal desmoid tumor and was successfully removed without recurrence. To date, the patient remains asymptomatic, with no pain and complete sensation in her third toe.

Although extra-abdominal periosteal desmoid tumors have been identified in the extremities, to the authors’ knowledge none have been reported as far distal as the toe. Identifying this lesion in the distal extremity will allow a hasty diagnosis and treatment in future cases of similar presentation. Knowledge of the existence of this type of tumor in the distal extremity may also assist in narrowing differential diagnoses.

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Desmoid tumors are rare, locally infiltrative, nonmalignant tumors that can be found either within the abdomen or in the extremities, at which point they are referred to as extra-abdominal desmoid tumors. Lesions in the distal extremities present with pain and limited motion and are rare; the majority of extra-abdominal lesions occur in more proximal portions of the upper and lower extremities and often arise at areas of repetitive stress, such as in the origin or insertion of tendons into bone.\(^1\) The most common sites of involvement of an extra-abdominal desmoid tumor are the shoulder and the thigh, at 21\% and 13\%, respectively.\(^1\) Extra-abdominal desmoid tumors have a female predominance and a peak incidence during ages 25 to 35 but have been described to occur from the onset of puberty to 60 years of age.\(^2\) Risk factors are believed to include prior surgical scar, injury to the area, elevated estrogen levels, and a possible somatic mutation in the adenomatous polyposis coli gene.\(^1,2\) Hormonal changes may contribute to the growth patterns of desmoid tumors.\(^3,5\)

This article describes a case of an extra-abdominal desmoid tumor occurring in the third toe of a healthy adult woman with no medical or familial predisposing history. To the authors’ knowledge, this is the first report of this type of tumor found as far distal as the toe. This case report will qualify the existence of the lesion in the distal extremity, allowing appropriate, timely diagnosis of the lesion in future cases. Informed consent and agreement was obtained from the patient that this case would be submitted for publication.

**Case Report**

A 37-year-old woman presented with a solid plantar mass on the distal phalanx of the left third toe, which had progressively increased in size for the past 3 years. The mass was first noticed as a discomfort while wearing shoes. The patient was evaluated by her primary care physician 3 years prior to presentation at the authors’ institution, at which time the mass was subjectively small, well-circumscribed, nontraumatic, nonpainful, and nonerythematous; did not affect her skin appearance or sensation; and was thought to be a clinically benign process due to a foreign body reaction. No radiographic workup was obtained, and the patient was conservatively followed. Two years after this initial consultation, the patient became pregnant, and the mass enlarged during the course of the pregnancy. She was then evaluated by a podiatrist, who suggested the mass be biopsied, but due to the pregnancy, the patient postponed biopsy until after her delivery and was subsequently seen by a foot and ankle orthopedic surgeon (M.O.) at the authors’ institution.

The patient had an unremarkable medical history. On physical examination, she experienced only mild pain and discomfort due to footwear. The third toe was grossly deformed; it appeared red, elongated, and had expanded to approximately the same size as her great toe (Figure 1). The plantar aspect of the toe was thick and callused, and the toenail was slightly elevated. Magnetic resonance imaging (MRI) showed a tuft enlargement of the third toe, originating from the plantar margin and extending distally (Figure 2). The soft tissue was eroded, but no apparent severe destruction of bone was found. It was determined that an epidermoid inclusion cyst best fit the characteristics of the MRI; however, a slow-growing tumor of subdermal elements was another possibility.

Showing no malignant qualities clinically or radiographically, a marginal excision with retention of the nail was performed without complication. The operation was performed under epidural anesthesia and ankle block. A longitudinal incision was made on the plantar aspect of the third toe directly over the mass. An excisional biopsy was performed, removing the tissue from the distal interphalan-geal joint to the flexor tendon. The mass had a rubbery consistency and was white, focally yellow, and tan soft tissue (Figure 3). The mass was peeled off of the bone, which appeared damaged and was debrided. Once all of the affected soft tissue and bone was excised, the wound was thoroughly irrigated and closed with 3-0 Vicryl (Ethicon, Inc, New Brunswick, New Jersey) and 4-0 Nylon (Ethicon, Inc). On further pathological examination, it was seen that the lesion was mitotically active; however, atypical mitotic figures were not present. The morphological features were consistent with a fibromatosis-like proliferation involving subcutaneous and periosteal tissue, and it was determined that the mass was a locally aggressive extra-abdominal periosteal desmoid tumor.

The patient had an uncomplicated postoperative course and, 6 years post-
operatively, has remained asymptomatic. Currently, the patient experiences no pain as a result of the operation and has complete sensation in her toe. The affected toe is shorter than her other toes, the skin on the plantar aspect of the toe is thick, and she has a mild hook nail deformity (Figure 4). No recurrence of the mass has occurred.

**DISCUSSION**

Although periosteal desmoid tumors have been reported in the literature, to the authors’ knowledge none have been reported as far distal as the toes. Clinical, radiographic, and histological presentations confirm the diagnosis, and a review of the literature shows that hormonal changes affect the growth rate of these tumors. An adequate resection is indicated for definitive treatment of localized symptoms and prevention of recurrence.

Reitamo et al. found a correlation between rate of desmoid tumor growth and estrogen levels in women, providing a possible explanation behind the finding that men of different ages exhibit the same growth rate of this tumor, whereas women of different ages exhibit different rates of growth. This study also noted that desmoid tumors were more common following a patient’s second pregnancy, with accelerated growth during the third trimester. The current patient presented for treatment following the completion of her pregnancy after no improvement was noted in the clinical presentation of the progressively growing mass. Additional studies by Dahn et al. and Strode on the relationship between hormones and desmoid tumors reported that hormones influence tumor growth with spontaneous regression of the desmoids at menopause.

Historically, extra-abdominal desmoid tumors have been known as extra-abdominal fibromatosis, musculoaponeurotic fibromatosis, and aggressive fibromatosis. The presence of peripheral muscular fibers, with a rare extension into the center of the lesion, is a common histological finding. Low cellularity with bland spindle fibers and dense stroma is also noted on histology. Immunohistochemical markers are positive for actin and beta-catenin and negative for desmin, CD34, S-100, and keratin. Radiographically, the lesion occurs at the origin or insertion of muscles and is characterized by focal cortical disruption and sclerosis. Areas of mineralization may exist within the lesion shown on MRI, with an overcall of the reactive area of bone due to the periosteal reaction without soft tissue involvement. These tumors are difficult to detect on radiographs, as seen in a review of 11 preoperative plain radiographs by Markhede et al. in which only 2 patients had detectable signs of pathology and 1 had bony destruction. The current authors’ historical review confirmed the presence of a fibromatosis-like proliferation of the periosteum with local aggressiveness and no signs of distant metastasis, with positive histochemical markers for extra-abdominal desmoid tumor. Markhede et al. reported that recurrence was controlled by adequacy of excision because 3 of 21 tumors recurred after adequate excision and 10 of 14 recurred after inadequate operative excision. Tumors occurring on the upper arm and on the thoracic wall recurred most often.

Differential diagnoses for extra-abdominal periosteal desmoid tumors may be subclassified based on radiographic findings and pathology findings. Similar radiographic findings may be found in periosteal chondroma, fibrous cortical defect, Ewing’s sarcoma, and osteosarcoma. Pathological similarities are seen in osteosarcoma, fibrosarcoma, and fibrous cortical defect. Periosteal desmoid tumors may present as radiolucent defects in bone with an area of sclerosis at the borders; occasionally, cortical irregularity from erosion of the lesion into the cortical bone may exist. These findings are more likely to be seen with fibrous cortical defect based on radiographic findings; however, extra-abdominal periosteal desmoid tumors rarely have bony changes noticeable on plain radiographs, as seen in the workup of the current patient. On occasion, spicules may be noted on radiographs, leading to malignant bony processes such as osteosarcoma and Ewing’s sarcoma over benign conditions.

This case report qualifies the existence of an extra-abdominal periosteal desmoid tumor as far distal as the toe. Operative treatment with marginal excision was successful in removal of the tumor without recurrence.

**REFERENCES**


