Recurrence of Extranodal Natural Killer/T-cell Lymphoma Presenting as Tarsal Tunnel Syndrome

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Take-Home Points

- A thorough review of systems, physical examination, and personal review of a patient’s advanced imaging is critical to avoid missed diagnosis or delays in diagnosis.
- Any mass lesion encountered in clinical practice, no matter how benign appearing, should be presumed malignant until proven otherwise.
- Fluorine-18 fluorodeoxyglucose positron emission tomography CT (18-FDG PET-CT) should include whole-body scans when evaluating patients for recurrence of malignancy.

Nasal-type, extranodal natural killer/T-cell lymphoma (ENKTL) is a rare form of non-Hodgkin lymphoma (NHL). Malignancies account for only 10% of NHL in Asian and South American populations. However, in Caucasians, it represents <1% of all cases. In addition, at 3:1 male to female ratio, the disease most commonly affects male patients who are 50 to 59 years old. The etiology of this malignancy is strongly related to prior infection with Epstein-Barr virus (EBV) as EBV-encoded early small ribonucleic acid on in situ hybridization of lymphoma cells is positive in 95% of cases.

Typical sites of involvement include the nasal cavity, nasopharynx, and sinuses, causing patients to present with nasal obstruction, chronic sinusitis, or epistaxis. Additionally, ENKTL can occur primarily in the skin, gastrointestinal tract, spleen, and testis, whereas the bone marrow may be involved in 10% of cases. Although rare, unusual sites, including muscle, adrenals, and ovaries, have been published.
Staging is best performed using the T-staging system, which accounts for the extent of local tumor involvement. Higher stages, such as T3 /T4, equate to locally advanced disease and imply a worse prognosis.\textsuperscript{9,10} Computed tomography (CT) and magnetic resonance imaging (MRI) help define local soft tissues and bony involvement. Furthermore, CT of the chest, abdomen, and pelvis as well as bone marrow biopsy are performed as part of the staging process. Lastly, fluorine-18 fluorodeoxyglucose positron emission tomography CT (18-FDG PET-CT) is often used to detect extranodal spread, define the extent of involvement, differentiate between lymphoma and inflammatory masses, and monitor for recurrence.\textsuperscript{11}

Treatment for local ENKTL involves concurrent chemoradiotherapy followed by 3 cycles of etoposide, ifosfamide, cisplatin, and dexamethasone, which results in a complete response rate of 80%, and is the most favorable when comparing treatment modalities.\textsuperscript{12} Unfortunately, recurrence rates reach as high as 50%, whereas the 5-year survival rate is 59%.\textsuperscript{13,14} For recurrent or disseminated disease, high-dose chemotherapy and hematopoietic stem cell transplantation remain as alternative treatments for patients who have undergone 2 complete remissions and can be curative in some instances.\textsuperscript{13,15}

In summary, ENKTL is a rare form of NHL which classically presents in the nasal cavity; however, this type of lymphoma may present in a variety of extranodal sites.\textsuperscript{7,8} Despite the numerous published reports on ENKTL, no study has reported either primary or recurrent ENKTL in the feet or hands. To our knowledge, this is one of the first published cases of a patient who developed a rare and recurring ENKTL in the foot and ankle. The patient provided written informed consent for print and electronic publication of this case report.

**Case**

A 59-year-old Caucasian woman was referred to the orthopedic foot and ankle clinic by her primary care physician for right medial ankle pain, skin ulceration, and numbness over the plantar aspect of her right foot. Upon questioning, the patient noted that the pain and numbness were present for almost 6 months. She denied trauma to the concerned area. Previously, the patient was observed and treated elsewhere for plantar fasciitis and was prescribed a brace before being immobilized in a controlled ankle motion (CAM) boot for 6 weeks. At follow-up with her outside provider, the patient had developed skin breakdown over the medial aspect of the right ankle, and this condition was presumed to be caused by the boot. After local wound care failed to improve her skin ulceration, she returned to her primary care physician, who ordered an MRI of the area and referred her to our specialty clinic.

Upon review, the patient’s past medical history included a diagnosis of nasal-type ENKTL. Her malignancy was treated with chemoradiotherapy 2 years prior to her consultation with the foot and ankle clinic.

The patient was noted by her medical oncologist and interventional radiologist to be in complete stage 4 remission since being treated. She underwent routine MRI and CT scans of the head and neck at 6-month intervals and FDG PET-CT scans at 3-month intervals, as per institutional protocol. The examinations showed no evidence of malignancy or metabolically active disease. The last imaging study occurred 2 months prior to admission to the foot and ankle clinic.

The patient consulted her medical oncologist 1 month prior to presenting to our clinic and was noted to exhibit an “excellent response to chemoradiotherapy” and “continues to remain disease free at 2 years.” She was instructed to continue routine follow-up. However, the office notes mentioned no ankle pain and non-healing wounds.

During physical examination, the patient presented an antalgic gait on the right side. Inspection demonstrated an increased circumference of the right ankle compared with the left, with a soft, palpable mass over the medial
aspect of her right ankle. A 3 cm × 2 cm, grade 2 abrasion of the skin was observed over the medial mass just posterior to her medial malleolus. Range of motion was within normal limits. The patient exhibited a palpable posterior tibial artery pulse and full strength upon muscle testing of the lower extremities. She featured a positive Tinel’s sign and discomfort over the mass itself, with the pain radiating down to the plantar aspect of her foot and diffuse numbness over the plantar aspect of the foot.

Review of her plain radiographs demonstrated no bony abnormalities, fractures, nor visible deformity (Figures 1A, 1B). MRI of the foot and ankle was reviewed and demonstrated a large soft-tissue mass over the medial aspect of the ankle extending posteriorly to engulf the medial flexor tendons and medial neurovascular bundle (Figures 2A-2C). Interestingly, the radiologist’s report only mentioned a “large region of devitalized tissue underlying the known medial ankle ulcer which extended to the tibiotalar and subtalar joints and bone marrow changes in the talus most compatible with osteomyelitis.” FDG PET-CT images from the patient’s past 2 years were reviewed, and the radiologist was contacted to confirm the negative findings. Unfortunately, FDG PET-CT performs no routine imaging below the elbow or knee when evaluating for this particular malignancy (Figure 3).

At presentation, our differential diagnosis included recurrence of the malignancy, secondary malignancy, infection, and inflammatory disease. After a lengthy discussion with the patient and consultation with our institution’s musculoskeletal oncologist, the decision was made to perform a right-ankle mass biopsy and marginal excision with wound irrigation and débridement and tarsal tunnel release.

The patient was placed in the supine position with standard prepping and draping. The medial eschar was excised in an elliptical fashion, and a curvilinear, longitudinal approach was performed within the compartment to access the mass along the posteromedial aspect of the ankle. Although no evidence of infection was observed, the tissue was thickened with areas of necrosis down to the flexor retinaculum. Once the flexor retinaculum was opened, a fibrous, plaque-like mass was observed, and it was encased with flexor tendons and neurovascular structures of the tarsal tunnel. After mass excision, a complete tarsal tunnel release was performed until the neurovascular bundle was free. Irrigation and débridement of the ulcer were performed along with complicated wound closure, and the patient was placed in a well-padded postoperative splint.

Pathology was finalized as a recurrent, EBV-positive, and nasal-type ENKTL. The patient underwent bone marrow biopsy, which yielded negative results. CT of the chest, abdomen, and pelvis were negative for the disease. FDG PET-CT, which included the extremities, was performed and demonstrated increased uptake in the right ankle, consistent with the malignancy (Figure 4). The patient was placed on her second cycle of chemoradiotherapy and again showed complete remission. At over 1-year follow-up, the patient’s wound had completely healed, and FDG PET-CT of the extremity was negative for ENKTL recurrence (Figure 5). At present, the patient’s medical oncologists recommend a stem cell transplant as the patient exhibited 2 positive responses to chemoradiotherapy.

Discussion

ENKTL is an uncommon form of lymphoma and is exceedingly rare in Caucasian females.\textsuperscript{1-3} Although the patient’s primary occurrence was in the nasal cavity, recurrence in the foot and ankle must still be described.\textsuperscript{7,8} To our knowledge, this article is one of the first published cases of a patient who developed a rare-recurrence ENKTL about the foot and ankle. Occurrence in extremities is extremely rare that the staging protocol does not include FDG PET-CT of these areas. The patient’s “negative” scans led many providers to neglect the symptoms in her right ankle until the lesion had ulcerated through the skin. If one would have relied on imaging reports and outside records alone, the diagnosis would have been delayed longer or missed all together. This case illustrates the importance of a thorough medical history and personal review of imaging studies, and how a systematic
approach can produce the correct diagnosis for any unknown lesion. Furthermore, this case may prompt oncologists to consider obtaining whole-body FDG PET-CT when evaluating for recurrence in patients.

**Key Info**

**Figures/Tables**

Figures / Tables:

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![Figure 1](aynardi0518_f1.jpg)

**Figure 1.** Normal appearing (A) anteroposterior and (B) lateral radiographs of the right ankle.

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Figure 2. (A) Axial T1-weighted magnetic resonance imaging (MRI) with fat suppression. (B) Coronal T2-weighted MRI. (C) Sagittal T1-weighted MRI with fat suppression.
Figure 3. Fluorodeoxyglucose positron emission tomography computed tomography of the patient included in the routine screening, which yielded negative results.
Figure 4. Fluorodeoxyglucose positron emission tomography computed tomography of the extremity, demonstrating uptake in the right ankle.
Figure 5. Fluorodeoxyglucose positron emission tomography computed tomography after chemoradiotherapy of the right ankle, demonstrating absence of recurrence at 1-year post-treatment.

References


Multimedia

Product Guide

Product Guide

- STRATAFIX™ Symmetric PDS™ Plus Knotless Tissue Control Device
- STRATAFIX™ Spiral Knotless Tissue Control Device
- BioComposite SwiveLock Anchor
- BioComposite SwiveLock C, with White/Black TigerTape™ Loop

Citation

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