Extraskeletal Myxoid Chondrosarcoma Presenting as a Plantar Fibroma: Case Report and Review of the Literature

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Abstract. *Background: Extraskeletal myxoid chondrosarcoma is a rare tumor with an indolent course, high propensity for local recurrence, metastases, and propensity for the proximal extremities of middle-aged males. Case Report: We present the case of a 44-year-old man with an extraskeletal myxoid chondrosarcoma in the plantar fascia of the medial arch initially thought to be a plantar fibroma. Magnetic resonance imaging of the lesion demonstrated a lobulated subcutaneous mass plantar to the tarsal bones and inseparable from the fascia. Microscopic examination revealed a lobulated lesion composed of cords and nests of round to spindled malignant cells in a blue-gray myxoid matrix surrounded by fibrous septae. The malignant cells displayed variable positivity for S-100. Conclusion: Plantar extraskeletal myxoid chondrosarcoma is a rare occurrence. It should always be considered in the differential diagnosis of masses arising in the plantar fascia of the foot.*

Extraskeletal myxoid chondrosarcoma (EMC) is a rare tumor with an indolent course and high propensity for local recurrence and metastasis. This tumor most commonly presents in the proximal extremities of middle-aged males, and is commonly asymptomatic. We present the case of a 44-year-old man with an EMC in the plantar fascia of the medial arch initially thought to be a plantar fibroma.

Case Report

Our patient, a 44-year-old African American male, presented with a subcutaneous mass in the medial arch of the plantar foot which had been growing in size since 2009. Magnetic resonance imaging (MRI) of the lesion demonstrated a lobulated subcutaneous mass plantar to the tarsal bones which was inseparable from the plantar fascia (Figure 1). The mass showed iso-signaling on T1-weighted images and an increased signal on T2-weighted images.

On surgical removal, the mass was tan-pink to yellow in color with a multicystic cut surface containing yellow mucoid material. Microscopic examination revealed a lobulated lesion composed of cords and nests of round to spindled malignant cells in a blue-gray myxoid matrix surrounded by fibrous septae (Figures 2 and 3). The surgical margin was positive in several areas. The malignant cells displayed variable positivity for S-100 (Figure 4).

Discussion

EMC is a rare tumor which accounts for approximately 2.5% of all soft tissue tumors (1, 2). It was first described by Enzinger and Shiraki in 1972, and was originally considered to be a low-grade lesion based on its histological appearance (1, 3). Case reports and series published later with longer follow-up intervals, however, demonstrated this entity to have a propensity for local recurrence and metastasis beyond 5 years (4-6). Saleh *et al*. followed patients for a minimum of 10 years, revealing EMC to have an indolent but resilient course with metastases developing later in all but one of their patients (4). Overall survival in a separate case series collected by Drilon *et al*. showed overall survival at 5 years was 82%, but at 10 and 15 years, it decreased to 65% and 58%, respectively (6). Greater than 70% of local recurrences and metastases developed beyond 5 years' follow-up, requiring close follow-up of patients for an extended period past initial resection (5).

As histological grade was determined to have no prognostic significance, several case series investigated other prognostic indicators. The most common poor prognostic factors described are larger tumors (the actual dimensions varying between reports), limb girdle involvement, and metastasis at presentation (2, 5, 7). The most common site of metastases is lung in over half of all patients (2). Metastases in deep soft tissue, lymph node, bone and brain, however, have also been reported (2).

Middle-aged men are most commonly affected (4, 6, 8, 9). EMC is largely asymptomatic with most patients describing

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Table I. *Literature review of primary foot and ankle extraskeletal myxoid chondrosarcoma.*

Author (Ref)	Number	Age (years)/Gender	Location	EWSR1 FISH
Enzinger and Shiraki (3)	5	65F, 42M, 56M, 37M, 58M	Foot (N=1), ankle (N=4)	Not performed
Englert *et al*. (16)	1	-	Foot	Not performed
Benini *et al*. (17)	1	-	Foot	Not performed
Kindblom and Angervall (18)	1	37M	Ankle	Not performed
Saleh *et al*. (4)	1	41M	Foot	Not performed
Banfic *et al*. (1)	1	46F	Ankle	Not performed
Jakowski and Wakely (20)	4	61F, 82M, 58F, 57F	Foot (N=3), Ankle (N=1)	Positive
Bhamra *et al*. (21)	1	71M	Ankle	Not performed
Xu *et al*. (22)	2	-	Toe	Not performed
Benini *et al*. (23)	3	18F, 38F, 65F	Foot	Positive
Current study	1	44M	Foot	Not performed

M: Male; F: female; EWSR1 FISH: Ewing sarcoma breakpoint region 1 fluorescence *in situ* hybridization.

a slow-growing painless mass. However, pain, tenderness, and restriction of motion correlating to tumor site have also been described (10). The most common sites are the extremities, but other locations including the neck, foot, orbit, and perineum have been described (2, 5, 8, 9, 11).

To our knowledge, from an extensive PubMed search, there are only 16 documented cases, including ours, of primary tumors of the foot as seen in Table I. Imaging of this entity is not specific, but typically shows a well-circumscribed, lobulated mass which may or may not be partially encapsulated. T1-Weighted MRI may be isointense to hypointense, while T2-weighted images are characteristically hyperintense which is concordant with a cartilaginous lesion (8, 10, 11). Reports of increased signaling on T1-weighted images has also been described, and has been correlated with areas of hemorrhage on pathological examination (10). Fluid-fluid levels are not typically present (10).

Grossly, EMC is a circumscribed, variably pseudo-encapsulated lobulated mass with a tan-pink to gray color. On sectioning, the tumor is glistening gray-white to tan-brown with mucoid material, and may have focal hemorrhage or cystic degeneration (8, 10, 11). On microscopic examination, EMC is multi-lobulated with malignant polygonal to spindled cells forming nests and cords in a myxoid matrix. Tan-pink bland fibrous septae enclose the lobules (1, 8, 10). Areas of rhabdoid differentiation have also been described (8). Belying its cartilaginous nature, EMC is commonly strongly positive for vimentin and variably positive for S-100 immunostaining (1, 8).

Recurrent chromosomal abnormalities present in EMC are t(9;22)(q22;q11-12) and t(9;17)(q22;q11), producing fusion genes Ewing sarcoma breakpoint region 1 (*EWS*)/nuclear receptor subfamily 4, group A, member 3 (*CHN*) and TAF15 RNA polymerase (*RBP56*)/*CHN*, respectively, with t(9;22) being most common (2, 6). Additionally, gene profile studies have been published identifying an array of diagnostic markers out of which the most common are: Dickkopf WNT signaling pathway inhibitor 1 (*DKK1*), neuromedin B (*NMB*), delta-notch-like EGF repeat-containing trasmembrane (*DNER*), chloride channel 3 (*CLCN3*), and differentially expressed in FDCP 6 homolog (DEF6) (6). The significance and utility of these markers is still being determined (6).

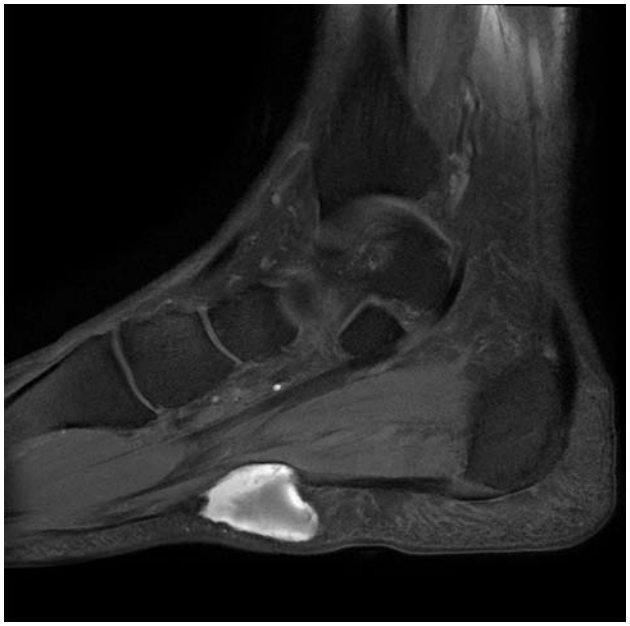

Figure 1. *Magnetic resonance image of lesion.*

Wide local excision is the mainstay of treatment (6, 11, 12). Radiation has been used in cases with positive microscopic margins with some success and in unresectable lesions with variable results. EMC is relatively

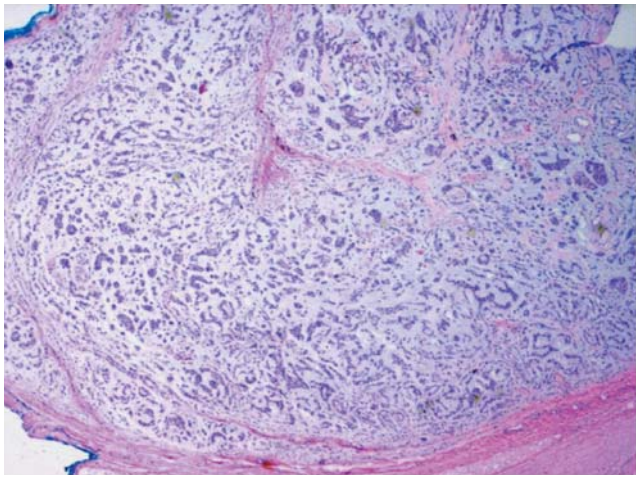

Figure 2. *H&E-stained section of mass, ×4 magnification.*

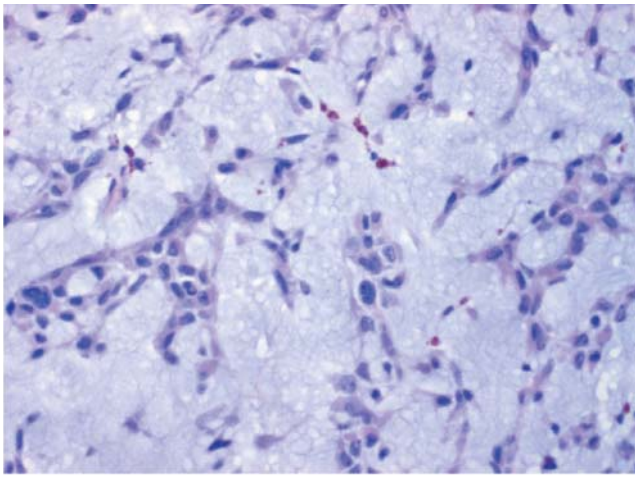

Figure 3. *H&E-stained section of mass, ×20 magnification.*

nonresponsive to chemotherapy. Although two cases have been reported of complete remission status post high-dose combination chemotherapy, it is a largely an ineffective treatment (2, 13).

Our patient, although slightly younger than the average patient with EMC, had similar clinicopathological features to previously reported cases. The site of our patient's tumor, however, was relatively unique in that it arises in the plantar fascia which led to an initial clinical diagnosis of plantar fibroma. Like our patient's tumor, plantar fibroma is typically a well-circumscribed nodule arising within the plantar fascia (14). Plantar fibroma is also a common lesion in middle-aged males, and is characteristically a slow-growing painless nodule (14, 15). On imaging, plantar fibroma most commonly demonstrates hypointense signaling on both T1- and T2-weighted imaging, however, increased signaling on T2-weighted images, as seen in our patient, has also been described (14, 15). Differing signal intensities in fibroma have been attributed to increased cellularity and myxoid changes (15). Although excision is a common treatment of plantar fibroma, wide margins are not necessary and may cause unnecessary morbidity (15). Since patients with EMC have a better prognosis with negative margins status post excision, it is important to consider EMC as a possibility, especially for lesions which have increased signaling on T2-weighted MRI.

EMC is a relatively rare soft tissue sarcoma that has an indolent course with a high propensity for late metastases. Although EMC has been previously described in the foot and ankle in a handful of cases, the presentation of our patient demonstrates the importance of keeping EMC in the differential diagnosis of masses arising in the plantar fascia of the foot.

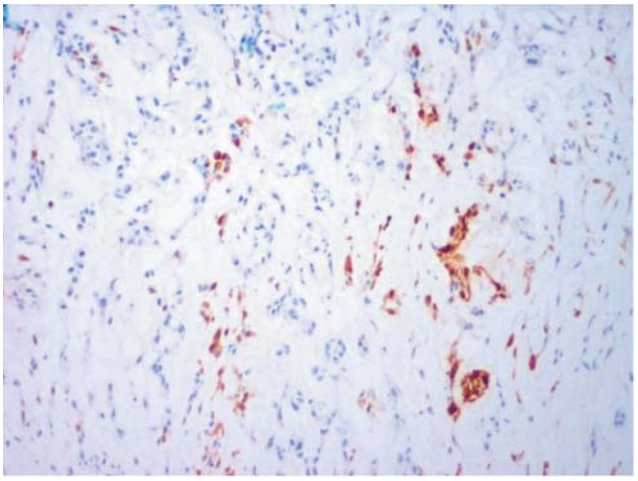

Figure 4. *S100-stained section, ×10 magnification.*

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