Clinical Case Report

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Extraskeletal myxoid chondrosarcoma of the leg in a child

A case report

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Abstract

Rationale: Extraskeletal myxoid chondrosarcoma is a slow-growing soft tissue tumor of adults with a propensity for local recurrence and eventual metastasis. Only 17 pediatric and adolescent cases have been reported.

Patient concerns: Here we present an 11-year-old boy with a 3-year history of a slowly growing painless left leg mass. Magnetic resonance imaging of the lesion revealed a subfascial well-circumscribed lesion with intramuscular extension in the medial gastrocnemius muscle of the left leg.

Diagnoses: He underwent wide local excision of the mass and the histomorphological and immunohistochemical findings were consistent with extraskeletal myxoid chondrosarcoma.

Interventions: Possible radiotherapy was the further management plan.

Outcomes: He was in good condition with no evidence of recurrence at 6 months postsurgery.

Lessons: Although pediatric cases of extraskeletal myxoid chondrosarcoma were reported to be aggressive, the tumor in this case demonstrated indolent behavior. Furthermore, the tumor in this case showed primitive round cell foci which adds to a previous study that especially reported this morphology in pediatric cases.

Abbreviations: EMA = epithelial membrane antigen, EMC = extraskeletal myxoid chondrosarcoma, MRI = magnetic resonance imaging.

Keywords: child, extraskeletal myxoid chondrosarcoma

1. Introduction

Extraskeletal myxoid chondrosarcoma (EMC) accounts for almost 3% of all soft tissue sarcomas.[1] The occurrence of EMC is mainly in the sixth decade with a male-to-female ratio of 1.5-2:1.[2] It is a slow-growing tumor with a tendency for local recurrence and eventual metastasis.[2] Most of the cases are deep seated and are located in the lower extremities.[2] Because there is no convincing evidence of cartilaginous differentiation, EMC is categorized as a tumor of uncertain differentiation.[3,4] The size of the tumor ranges from 1.1 to 25 cm with a median size of 7 cm.[5] Histologically, multinodular pattern is evident, and the tumor cells are arranged in strands, cords or epithelial-like aggregates in an abundant myxoid matrix. The individual cells have hyperchromatic nuclei and a narrow rim of cytoplasm with occasional cytoplasmic vacuolization and rare mitotic figures.[1] To the best of our knowledge, only 17 pediatric and adolescent cases of EMC have been reported.[2,6–10] Here we report an 11-year-old boy with a soft tissue tumor in his left leg. EMC was the preferred diagnosis.

2. Case report

This 11-year-old male had a 3-year history of a slowly growing painless left leg mass, first noticed after trauma. Physical examination revealed a fixed mass in the left leg with no skin changes or ulceration noted. Magnetic resonance imaging (MRI) of the lesion revealed a subfascial well-circumscribed lesion with intramuscular extension in the medial gastrocnemius muscle of the left leg (Fig. 1). Clinicoradiological findings could not exclude sarcoma.

Excision was performed, and the specimen measured $3 \times 1 \times 0.5$ cm. Gross examination showed a multinodular whitish cut surface. Microscopic examination revealed a moderately cellular tumor with focal infiltration of the surrounding muscle and fatty tissue. On low power, the tumor was characterized by multinodular pattern and extensive sclerosis (Fig. 2). Most of the cells were arranged in a reticular pattern and were embedded in the abundant myxoid stroma. Characteristic small dark nuclei with narrow rim of eosinophilic cytoplasm arranged in strands and cords were observed (Fig. 3). Some foci demonstrating round cells with a more primitive appearance and lesser amounts of myxoid matrix were also observed (Fig. 4). In addition, cytoplasmic vacuolization of the tumor cells was focally present. Mitoses were rarely observed, and necrosis was not identified.

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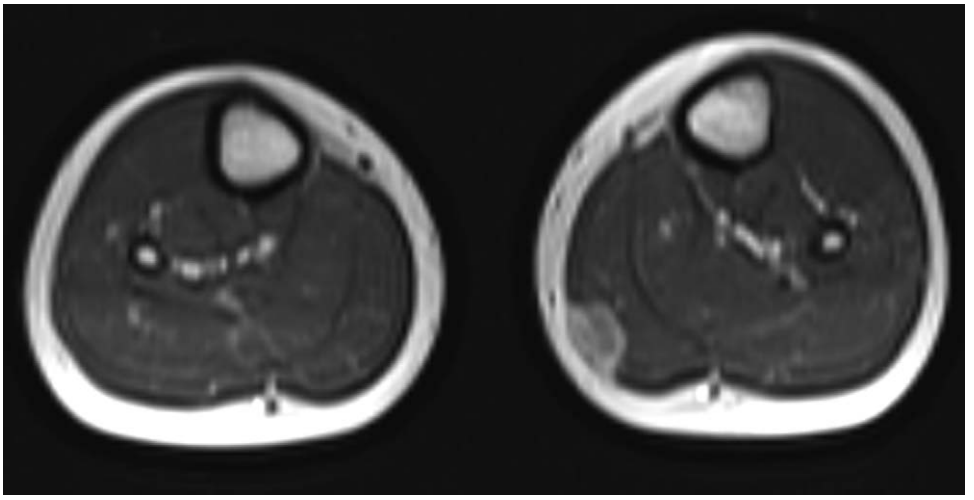

Figure 1. MRI showing a subfascial well-circumscribed lesion with intramuscular extension in the medial gastrocnemius muscle of the left leg. MRI = magnetic resonance imaging.

Immunohistochemical staining for cytokeratin AE1/AE3, epithelial membrane antigen (EMA), desmin, S100, CD99, CD117, INI-1, and Ki67 was performed. The tumor cells were positive for S100. Cytokeratin AE1/AE3 was positive focally in scattered cells. EMA, desmin, CD99, and CD117 were negative. INI-1 protein was retained in the tumor cells, and Ki67 labeling index reached about 10% in some areas. Extraskeletal myxoid chondrosarcoma was the preferred diagnosis. Possible radiotherapy was the further management plan.

Informed written consent was obtained from the patient's family for publication of this case report and accompanying images.

3. Discussion

EMC is a slow-growing soft tissue tumor that occurs mainly in the lower extremities of adults. To the best of our knowledge, only 17 pediatric and adolescent cases of EMC have been reported.[9,10] Moreover, in pediatric population, EMC tends to exhibit more aggressive behavior.[8,10] Histologically, EMC has a characteristic morphology in the form of lobules of cords and strands of chondroblast-like cells in an abundant myxoid stroma.[1] The differential diagnosis of EMC may include the aggressive and rarely reported tumor myoepithelial carcinoma of soft tissue.[11] However, soft tissue myoepithelial carcinoma was excluded due to the following: Cellularity was not pronounced, sclerosis was extensive, necrosis was not identified, mitoses were rare, and the typical areas of EMC with prominent nodularity were present.[1] Unfortunately, we have not investigated NR4A3 rearrangement which is an effective adjunct in diagnosing EMC.[12] We relied on the characteristic histomorphological, immunohistochemical and radiological features of the tumor. Moreover, the only pediatric case of EMC that was investigated for NR4A3 rearrangement showed a negative result.[9] Another entity important for the differential diagnosis was an ossifying fibromyxoid tumor and this was excluded due to the absence of lamellar bone.[13]

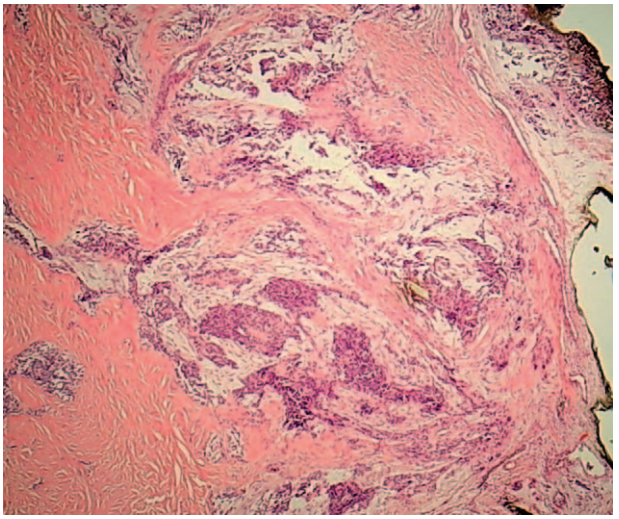

Figure 2. Nodules of tumor cells embedded in the myxoid material and separated by areas of extensive sclerosis (HE staining, ×40).

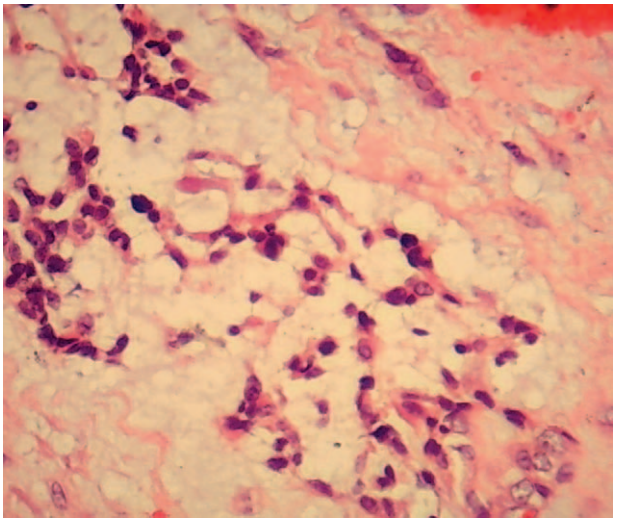

Figure 3. Strands of spindle cells with small hyperchromatic nuclei and a narrow rim of eosinophilic cytoplasm (HE staining, ×400).

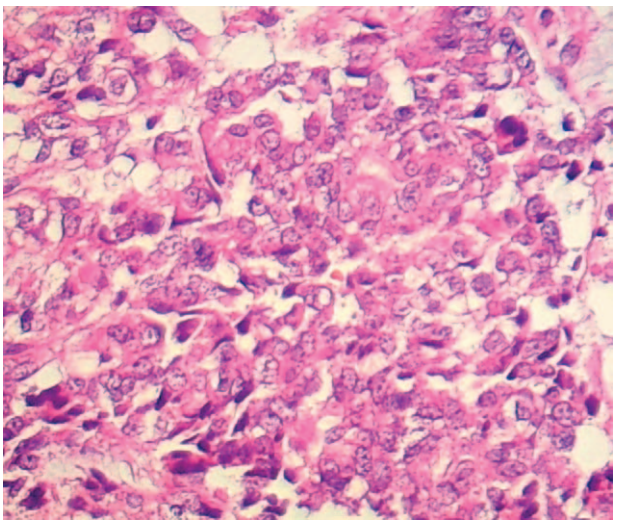

Figure 4. Areas showing round cells with a more primitive appearance and lesser amounts of myxoid matrix (HE staining, ×400).

A feature in our case that may differ from adult EMC was the presence of foci containing round cells with a primitive appearance and small amounts of myxoid matrix. Interestingly, this was comparable to a report by Hachitanda et al.[8]

The lesion was positive for S100, and this was comparable to some reports.[14] Moreover, EMC can be focally positive for pancytokeratin and it tends to be negative for desmin and EMA as in our case.[5]

The small round cell foci previously mentioned were desmin-negative, CD99-negative, and S100-positive. These findings excluded the possibility of the small round blue cell tumors rhabdomyosarcoma, Ewing sarcoma, and neuroblastoma, respectively.[15]

The prognosis for children with EMC is very poor.[10] Hachitanda et al reported death at 7 months postsurgery even with the use of chemo- and radiotherapy.[8] Kauffman and Stout reported death at 6 months postsurgery due to widespread metastatic disease.[6] Enzinger and Shiraki reported recurrence at 2 months, and the patient died after 5 months due to lung, heart, and brain metastasis.[2] Death by 2 months was reported by Jessurun et al and Klijanienko et al.[7,16] Romañach et al reported death after 1 year even with the use of radiotherapy.[17] In a recent report by Ibrahim et al, lung metastasis occurred 5 months postexcision.[10]

The small size reached over a period of 3 years in our case suggests an indolent behavior. In addition, the patient was in complete remission over a 6-month follow-up period. This was only comparable to the case reported by Boyd.[9] Surgical removal with negative margins remains the mainstay of treatment. However, follow-up is mandatory and radiotherapy can be considered to prevent recurrence.[18]

4. Conclusions

Although pediatric cases of EMC were reported to be aggressive, the tumor in the present case demonstrated an indolent behavior. In addition, special morphological features, in this case, were comparable to a previous report of pediatric EMC.

Author contributions

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