Subcutaneous Myxoma of the Ankle in a Young Child

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ABSTRACT

Soft tissue myxoma is a rare benign disease, and subcutaneous myxoma is extremely rare. It primarily affects middle-aged patients, and its occurrence in children is particularly uncommon. We present the case of a 5-year-old girl with a slowly growing subcutaneous myxoma in the left ankle. We summarize the radiological features of soft tissue myxoma on the basis of our case and the current literature.

CASE REPORT

A 5-year-old girl presented with a palpable subcutaneous mass in the left ankle at our orthopedic outpatient department. The mass had been observed for 6 months. She denied experiencing fever or trauma in this area. Physical examination showed a 2 cm × 2 cm elastic, mobile, and painless tumor in the left ankle without evidence of erythema or warmth over the area. Initially, a ganglion cyst was suspected. Laboratory data were unremarkable. A radiograph of the left ankle showed a soft tissue mass adjacent to the lateral malleolus without bony erosion (Fig. 1). Neither calcification nor a periosteal reaction was observed. Magnetic resonance imaging (MRI) of the left ankle revealed an ill-defined mass lateral to the lateral malleolus (Fig. 2, 3). In T1-weighted images, the mass had low intensity (Fig. 2a), whereas in T2-weighted images, the mass appeared heterogeneous with some areas of distinctly high intensity (Fig. 2b). The lesion was located in the subcutaneous region with a small part extending towards the sinus tarsi, forming an infiltrative growth pattern (Fig. 2c, 3c). Fat-suppressed T1-weighted images with contrast injection showed that the tumor was heterogeneously enhanced, especially within the peripheral region (Fig. 3).

The imaging findings ruled against the likelihood of ganglion cyst. Due to the infiltrative appearance, malignancy could not be excluded. Surgical excision was then performed. During the operation, the tumor appeared to be a 3 cm × 3 cm × 2 cm subcutaneous, lobulated, and gelatinous mass with involvement of the sinus tarsi. Pathological examination showed that the tumor contained few cells and abundant mucoid material. Vascular structure was scanty.
Subcutaneous myxoma

and delicate fibrous septa were present (Fig. 4a, 4b). Thus, the final diagnosis was myxoma.

The girl is doing well after the surgery, and there has been no recurrence for 5 years.

DISCUSSION

Myxoma most commonly occurs in the heart, and extracardiac myxoma is rare. It is a benign soft tissue tumor mostly composed of fibroblasts and myxoid stroma. Myxomas are histologically described as paucicellular, hypovascular, and being composed of cytologically bland cells separated by abundant extracellular myxoid matrix [5].

Soft tissue myxoma usually arises in the intramuscular compartment (82%). Only a small number of cases have lesions in the intermuscular, subcutaneous, or juxtaarticular regions [1]. It primarily affects patients between the ages of 40–70 years, with an average age of 52 years [1-3]. Women are more frequently affected (59%) [1]. The incidence rate is around 0.1-0.13 cases per 100,000 people [2, 6, 7].

The most common location is the thigh (51%), followed by the upper arm (9%), calf or buttock (each 7%). Incidences in the hand, forearm, chest wall, or paraspinal region each account for 4%, while incidences in the groin, knee, toe, or retroperitoneum each account for 2% [1].

Because subcutaneous myxoma is extremely rare, there is no large study discussing the imaging characteristics of this type of mass. However, intramuscular myxomas are well characterized and numerous reports have addressed their imaging characteristics. Radiographs usually appear normal. There is no evidence of calcification, bone involvement, or periosteal reaction [1]. On computed tomography, it appears as a well-defined hypodense mass with a fat rim or a fat cap in some cases. In others, the tumor shows either mild no enhancement [1]. It appears as a well-circumscribed lesion on MRI, typically hypointense on T1-weighted images, and markedly hyperintense on T2-weighted images [1, 4, 8]. Sometimes, a fat rim or a fat cap in the upper and/or lower poles of the lesions is detectable in T1-weighted images, and an edematous change of the peritumoral muscles is observed in T2-weighted images [1, 3, 4, 8]. A pseudocapsule is identified in most lesions [1, 4]. MRI shows 3 different enhancement patterns, including purely peripheral enhancement and peripheral enhancement with patchy or linear internal enhancement [4]. Although the degree of enhancement varies, it is most frequently intense and heterogeneous [8].

The myxoma in this 5-year-old girl is, to our knowledge, the first subcutaneous myxoma reported in a young child. In the literature, only a few young children have been reported to have myxoma, and all of these have been intramuscular masses [9, 10]. Our case is the youngest patient with subcutaneous myxoma.

In addition, the tumor in our case is located in the ankle, which is a rare location for a myxoma. Furthermore, the infiltrative growth pattern of the tumor is exceptionally uncommon. This atypical pattern is likely due to the restricted ambit of the subcutaneous area in the ankle; therefore, the tumor grows deeply into the sinus tarsi.

Nonetheless, in our case, some characteristic findings of the tumor on MRI are still noted, including a hypointense signal on T1-weighted images, a markedly hyperintense signal on T2-weighted images, and a heterogeneous but predominantly peripheral enhancement pattern. Other case reports in the literature have also shown similar results [11, 12]. Consequently, subcutaneous myxomas share several imaging features with intramuscular myxomas, as assessed by MRI even though they occur in 2 anatomically distinct locations.

The differential diagnosis of a soft tissue mass with high water content that mimics a cyst includes the following: benign cystic lesions, neurogenic neoplasms, and myxoid containing soft tissue mass. Cystic masses such as synovial cysts or ganglion cysts may mimic the appearance of myxoma on unenhanced MRI. However, they are easily recognized by

Figure 1. A radiograph of the left ankle in anteroposterior view shows a soft tissue prominence adjacent to the lateral malleolus (arrows). There was neither calcification nor periosteal reaction.
Subcutaneous myxoma

**Figure 2.** MRI of the left ankle.  
**a.** T1-weighted images showed a hypointense infiltrative mass lateral to the lateral malleolus. A major part of the lesion was located in the subcutaneous region (arrowheads) with a slight deep extension to the sinus tarsi (arrows).  
**b.** The mass on T2-weighted images was heterogeneously and markedly hyperintense.  
**c.** Enhanced fat-saturated T1-weighted images showed that the mass was heterogeneously enhanced, especially in the peripheral region.

**Figure 3.** a–c Enhanced fat-saturated T1-weighted images showed that a major part of the lesion was located in the subcutaneous region (arrowheads) with infiltrative extension to the sinus tarsi (arrows). The enhancement was heterogeneous but more significant on the peripheral region.
Subcutaneous myxoma

Figure 4. a. The low-power histology view showed few cells and abundant myxoid stroma. Vascular structure was scanty and delicate fibrous septa were present (H&E staining, magnification × 20). b. The high-power histology view showed spindle-shaped cells containing small uniform nuclei in pools of mucin. (H&E staining, magnification × 200).

the enhancement of a thin rim and delicate septa on CT or MRI obtained after administration of intravenous contrast material. Most neurogenic tumors are intermuscular lesions. Entering and exiting nerves are often seen on imaging, and the target sign may be apparent on T2-weighted MRI. Myxoma should also be differentiated from myxoid-containing soft tissue tumors in the extremities, including myxoid liposarcoma, myxoid peripheral nerve sheath tumors, and myxoid malignant fibrohistiocytoma (MFH). On T1-weighted MRI, myxoid liposarcomas appear as hypointense masses with hyperintense foci that correspond to fat. Myxoid liposarcomas show intense enhancement of the tumor volume in most cases. The above-mentioned features can help to differentiate myxoid liposarcomas from myxomas. Further, identification of a perilesional fat rim or fat cap on T1-weighted images and peritumoral surrounding muscle edema on T2-weighted images are the most reliable radiological features for differentiating intramuscular myxoma from other myxoid soft tissue lesions [8]. Myxoid MFH usually has a far more heterogeneous appearance with areas of hemorrhage and solid nodular regions showing prominent contrast enhancement. The mass in this study can be easily differentiated from a ganglion cyst by the heterogenous enhancement. Myxoid liposarcoma was also not likely due to the lack of hyperintense foci within the mass on T1-weighted images. However, the myxoma in our case showed neither a perilesional fat rim nor peritumoral surrounding muscle edema, making it difficult to differentiate this myxoma from a myxoid MFH.

The prognosis of a myxoma is quite good, and this condition is always curable by surgical excision. Recurrence, even in cases with incomplete resection, is extremely rare [5].

In conclusion, we have shown that the MRI characteristics of a myxoma in a rare subcutaneous location have several visual features in common with those of an intramuscular myxoma. Moreover, subcutaneous myxoma can occur even in a young child. Therefore, when encountering a subcutaneous tumor with a radiological appearance similar to an intramuscular myxoma, the differential diagnosis should include subcutaneous myxoma, even in a young child.

REFERENCES

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