Buttock Mass in a 46-year-old Woman

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History and Physical Examination

A 46-year-old woman was referred to the orthopaedic service with a 2-month history of an incidental finding of a right buttock mass. Initially, an abdominal ultrasound performed for an abdominal aortic aneurysm screening identified a liver lesion, so additional studies were ordered eventually revealing an atypical liver hemangioma. An abdominal computed tomography (CT) scan revealed a right buttock mass in the posterior subcutaneous tissue of the buttocks. The mass did not cause pain before the imaging studies, but at the time of referral to the orthopaedic service, the patient complained of pain while seated.

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The pain was intermittent, aching, and dull, which worsened on sitting but did not bother her at night. She denied any history of trauma to the area. The rest of her history was unremarkable.

On physical examination, a soft, nontender mass was palpated. The mass was mobile with respect to the bone and underlying muscle. Soft tissue swelling was present and was most prominent posteriorly. There was no tenderness to palpation of the thigh and no evidence of erythema or warmth over the area. Active and passive range of motion of the hips and knees bilaterally was full and painless. Strength and tone were normal in the bilateral lower extremities. Her lower extremity neurologic examination was normal. Her gait was normal. The remainder of the musculoskeletal examination was normal.

Plain radiographs and MRI scans (Figs. 1–3) of the lesion were performed.

Based on the history, physical examination, and the imaging studies, what is the differential diagnosis?

Imaging Interpretation

Plain radiographs of the lesion showed no soft tissue calcification or bony lesions. MRI of the pelvis showed a well-circumscribed, ovoid, subcutaneous mass, measuring 2.9 cm in its greatest dimension, in the deep subcutaneous fat of the distal buttock, inferior to the ischium and deep to the medial edge of the gluteus maximus (Fig. 1). The lesion had an isointense signal to muscle on the T1-weighted images (Fig. 1) and a hyperintense signal on the fat-suppressed T2-weighted images (Fig. 2). On post-gadolinium images, there was circumferential, irregular rim enhancement with less intense, heterogeneous nodular enhancement centrally (Fig. 3).

Fig. 1 This axial T1-weighted MR image shows a mass in the subcutaneous fat of the right buttock. The mass has a slightly less intense signal than muscle.

Fig. 3 This axial T1-weighted MR image with contrast enhancement and fat saturation shows rim enhancement with some heterogeneous enhancement within the lesion.

Fig. 2 This coronal T2-weighted MR image with fat saturation shows the lesion, which has a bright signal. There is no evidence of surrounding edema within the muscle or subcutaneous fat.

Differential Diagnosis

Peripheral nerve sheath tumor
 Hemangioma
 Chronic abscess
 Myxoma
 Myxoid liposarcoma

Fig. 4 This low-power histology view shows spindle cells set in an abundant myxoid stroma (Stain, hematoxylin and eosin; original magnification, $\times 10$).

Myxoid malignant fibrous histiocytoma
 Low-grade fibromyxoid sarcoma

A CT-guided needle biopsy was performed at an outside institution and was nondiagnostic. Wide excisional biopsy was then performed. On excision, it was confirmed the mass was not connected to bone or the underlying muscle. Histology studies of the excised lesion were performed (Figs. 4–6).

Based on the history, physical findings, imaging studies, and histologic picture, what is the diagnosis and how should this lesion be treated?

Fig. 5 This high-power histology view shows stellate cells with pyknotic nuclei in pools of mucin (Stain, hematoxylin and eosin; original magnification, $\times 40$).

Fig. 6 This medium-power histology view shows cystic spaces containing myxoid material (Stain, hematoxylin and eosin; original magnification, $\times 20$).

Histology Interpretation

Pathologic analysis from the nondiagnostic CT-guided needle biopsy showed fragments of fibroadipose tissue with focal cytologic atypia.

On gross examination, the resected portion was seen to consist of yellow fatty tissue measuring $4 \times 3.8 \times 1.9$ cm. Serial sectioning revealed a circumscribed, tan, mucoid nodule measuring $2.4 \times 1.5 \times 1.4$ cm with a bulging cut surface.

Histologic sections showed a predominantly myxoid neoplasm with benign-appearing spindle and stellate cells (Figs. 4, 5). The cells contained scant cytoplasm and hyperchromatic pyknotic nuclei with no atypia, mitosis, or necrosis. Cystic spaces containing mucoid material were easily discernible throughout the lesion (Fig. 6).

Diagnosis

Extramuscular myxoma.

Discussion and Treatment

Myxomas are tumors of unknown etiology whose myxoid appearance resembles primitive mesenchyme histologically. Extracardiac myxomas were first described in detail in the English literature by Stout in 1948 [42]. Soft tissue myxomas are rare, benign tumors but are the most common type of myxoma encountered outside of cardiac, head, and cutaneous myxomas [42]. To further delineate these tumors, Enzinger [12] described the similar characteristics of myxomas within muscle, which he called intramuscular myxomas. While the etiology is unknown, some have speculated they result from a traumatic incident, with one study [19] showing $\frac{1}{2}$ of the cases with a history of trauma, although it is not a typical finding. Another theory is that they are neoplastic in origin, where the neoplastic cells produce an excessive quantity of polysaccharides [12, 19, 22]. While this case presents a subcutaneous myxoma, many of its features are typical of intramuscular myxomas, such as its presentation, anatomic location, age group, pathology, and radiographic findings.

Intramuscular myxomas, while rare, must be on the differential diagnosis lists of tumors presenting inside muscle. The incidence has been reported as 1.1 per million people [31]. They are found more often in women and are seen commonly in the fourth to sixth decades of life but can occur at any age [26, 31, 33, 40]. They are typically located within the large muscles of the thigh, buttocks, lower leg, and shoulder girdle [12, 19, 26, 31, 33, 40]. While usually asymptomatic, the most common clinical sign is a slow-growing mass that can be painful in less than 20% of patients [9, 12, 19, 26, 33, 40]. Histologically, intramuscular myxomas are composed of a small number of spindle or stellate cells (fibroblasts) in a prominent myxoid matrix (mucus) characterized by hypocellularity, poor vascularization, and no mitotic figures [2, 19, 22, 26, 31, 33, 40]. On gross examination, myxomas are commonly described as gray/white, well circumscribed, lobulated, and gelatinous. On microscopic examination, however, atrophic muscle fibers can often be seen infiltrating the boundaries of the cut section [2, 12, 19, 31, 33, 40]. Alcian blue or colloidal iron stains show the myxoid matrix composed mainly of acid mucopolysaccharides, but only before hyaluronidase treatment [13, 19, 26]. Owing to their relative lack of glycogen, the spindle or stellate cells are negative on periodic acid-Schiff stain [19, 31]. Silver impregnation reveals a classic loose meshwork of reticulin fibers

[18, 19]. Focal, fluid-filled microcystic structures may be seen in some cases [18, 29, 31–33].

Immunohistochemistry shows vimentin as strongly positive in the myxoma stromal cells and desmin, S-100 protein, and endothelial markers as negative [18, 31]. Two studies showed fine-needle aspiration was only diagnostic in a small number of cases, citing the relatively low cellularity, low tumor incidence, and lack of distinct cytologic features as factors [9, 40]. Fine-needle aspiration was nondiagnostic in our case, leading to an excisional biopsy. In our experience, myxoid tumors have been difficult to diagnose using needle biopsy [35].

Radiographic workup of intramuscular myxomas involves MRI. On T1-weighted imaging, they are typically homogeneous and hypointense relative to muscle, whereas on T2-weighted imaging, they tend to be hyperintense [6, 29, 32, 34, 36]. Fat-suppressed T2-weighted images always show increased signal intensity due to the water content of the myxoid matrix [32]. Gadolinium-enhanced images tend to show a heterogeneous increase in signal intensity more accurately showing the solid consistency of this neoplasm [6, 23, 29, 32, 34, 36]. MRI studies of intramuscular myxomas have reported a thin perilesional rim of increased signal intensity appearing like fat on T1-weighted images in 65% to 89% of cases and increased signal intensity in the surrounding muscle on the T2-weighted images suggesting edema in 55% to 94% [6, 29, 32]. Murphey et al. [32] concluded the small amount of tissue resembling fat around the tumors was attributable to surrounding muscle atrophy.

Multiple intramuscular myxomas may be associated with fibrous dysplasia in Albright's syndrome or Mazabraud's syndrome [2, 7, 23, 29, 44, 47]. Mazabraud's syndrome consists of multiple intramuscular myxomas and fibrous dysplasia. Albright's syndrome is characterized by polyostotic fibrous dysplasia, café-au-lait spots, and endocrine hyperfunction, along with intramuscular myxomas. The exact relation of the findings in these syndromes remains unknown. Our patient did not have a history of fibrous dysplasia or cutaneous lesions and no other tumors were found.

The myxoma in our case was present in the deep subcutaneous tissues of the buttock. While the majority of myxomas that are not intramuscular are located in the head, heart, or skin, they can be found in many areas of the body. Myxomas tend to have a distinct histologic pattern described above that varies little with the site of the lesion. Sponsel et al. [41] reported six cases of myxomas before the term intramuscular myxoma was introduced with three in the finger and one each in the foot, in the arm, and under the deltoid, where they were generally superficial, although their exact locations relative to muscle were not mentioned. Three cases of myxomas in the subcutaneous tissue of the

neck have been reported [8, 15]. Since the description of intramuscular myxomas in 1965, at least 16 cases of extramuscular myxomas have been reported in the hand and five in the foot [3, 4, 14, 17, 20–22, 25, 38, 39, 43, 45, 46]. Myxomas in other locations rarely have been described.

Benign lesions on the differential diagnosis list can be easily distinguished histologically, but, because of the myxoid stroma of myxomas, they may be difficult to exclude on imaging [24, 30, 36]. Benign peripheral nerve sheath tumors can have similar imaging characteristics, but intralesional neural tissue and S-100 positivity distinguish them histologically [18]. Hemangiomas typically have more contrast enhancement and occasionally have phleboliths on imaging. On histologic examination they have enlarged vascular channels but lack a myxoid stroma. A chronic abscess has a thick, enhancing capsule with little to no enhancement in the lesion, but a hyperintense signal on fat-saturated T2-weighted imaging is present owing to the high water content. Histologic examination easily determines this diagnosis, showing an encapsulated lesion filled with necrotic material with or without organisms.

Owing to myxoma characteristics, the imaging differential includes several malignant tumors [27, 29, 36] and the histologic diagnosis may be challenging as many tumors show myxomatous change as a prominent feature. The nodular growth pattern, increased cellularity, presence of fat containing cells (lipoblasts), and chicken wire or plexiform vascular pattern separates myxoid liposarcoma from myxoma [19, 33]. Myxoid malignant fibrous histiocytomas have a wide anatomic distribution, so they can be found in similar areas as myxomas [33]. While they are also hypocellular lesions with an abundant myxoid matrix, they always show a greater degree of nuclear pleomorphism than myxomas and contain mitotic figures [18, 33]. Low-grade fibromyxoid sarcomas have alternating myxoid and fibrous areas with whirled cellular arrangements and a predilection for the extremities [18, 33].

Treatment of myxomas is generally marginal surgical excision, although wide margins are recommended by some [10]. No instances of recurrence, metastasis, or malignant degeneration of intramuscular myxomas have been reported in several series of patients with solitary lesions [1, 9, 10, 12, 13, 19, 22, 26, 31, 33, 37, 40, 42]. Three case series that include several types of myxomas have reported six recurrences in solitary soft tissue myxomas outside of the heart, head, and skin: three of 13 cases reported by Attar [5], one of seven cases by Dutz and Stout [11], and two of 55 cases by Ireland et al. [22], with none being intramuscular myxomas. Two of these studies cited inadequate excision as the probable cause of the recurrences [5, 22]. Some isolated recurrences outside of cardiac, head, and cutaneous myxomas have been reported in patients with atypical presentations of myxomas and in

cases associated with Albright's syndrome or Mazabraud's syndrome [11, 14, 16, 23, 28, 29, 43–45]. To our knowledge, spontaneous regression has not been reported.

The lesion in our patient can be considered an intramuscular myxoma because it satisfies all of the histologic components of an intramuscular myxoma except the tumor did not approximate muscle or periosteum and was contained within subcutaneous fat. It also had the characteristic MRI findings presenting in the typical patient age range and anatomic location.

Because the needle biopsy had been nondiagnostic and the imaging provided a broad differential diagnosis of benign and malignant lesions, wide excision was chosen because it treated all possible diagnoses and spared the patient a second operation if the final diagnosis was a malignant tumor or a benign tumor best treated with resection. Wide excision of the mass could be performed without compromising function owing to its lack of involvement of the fascia, skin, or muscle, which was apparent on MRI. The location of the mass within the deep subcutaneous tissue of the buttock permitted wide excision without compromising cosmesis. The patient is now 6 months postexcision without complaints or clinical evidence of recurrence. This case underscores the fact that a myxoma should be considered in the appropriate clinical presentation of a subcutaneous mass.

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