A reversible severe gait disorder caused by an unusual presentation of a musculoskeletal desmoid tumor: A case report

Tenenbaum Shay, Hershkovich Oded, Shabshin Noga, Chechik Aharon, Liberman Boaz

ABSTRACT

Introduction: Gait disorders are a clinical presentation that can be caused by a variety of etiologies. The classification of the patterns of gait varies according to the type of professionals assessing the patient. While neurologists tend to classify gait abnormalities according to the location of lesion and the anatomic level, orthopedic surgeons and physical and rehabilitation physicians assess gait disorder in a descriptive bio-mechanical manner. Desmoid tumors are benign tumors that can arise at any site in the body and in most cases are confined to the musculature. Desmoid tumors are composed of normal appearing fibroblastic cells in abundant fibrous stroma. They vary from indolent growing asymptomatic tumors to local extremely aggressive tumors which can lead to mortality related to invasion of adjacent vital structures. Case Report: We report an unusual case of a desmoid tumor in the gluteal region presenting as a progressive gait disorder in an otherwise healthy woman. Conclusion: Removal of the tumor after years lead to regaining of the normal gait.

Keywords: Gait disturbances, Gait disorders, MRI, Imaging, Desmoid tumor

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INTRODUCTION

Gait disorders are a clinical presentation that can be caused by a variety of etiologies. The classification of the patterns of gait varies according to the type of professionals assessing the patient. While neurologists tend to classify gait abnormalities according to the location of lesion and the anatomic level, orthopedic surgeons and physical and rehabilitation physicians assess gait disorder in a descriptive bio-mechanical manner [1].

A gait disorder can arise from a central nervous system lesion such as a tumor, stroke, trauma or multiple sclerosis [1]. It can be caused by impairment of the peripheral nervous system such as in Guillain-Barre syndrome, a herniated intervertebral disc disease causing radiculopathy, polyneuropathy or caused by a musculoskeletal pathology such as arthritis, deformation, congenital hip dysplasia, tumors and muscular dystrophy [1].

Desmoid tumors are benign tumors that can arise at any site in the body and in most cases are confined to the musculature [2]. Desmoid tumors are composed of normal appearing fibroblastic cells in an abundant fibrous stroma. They vary from indolent growing asymptomatic tumors to local, extremely aggressive tumors which can lead to mortality related to invasion of adjacent vital structures [2–4].
Recently, modern treatment for tumors has developed based on a multidisciplinary approach. Although wide-margin surgical resection of the tumor is considered as first-line therapy, radiation is used as adjuvant therapy as well as low-dose chemotherapy protocols [4–10]. There are several novel pharmacologic and biologic treatments that are under development, however, long-term follow-up is needed for their implementation [7, 11].

Desmoid tumor is a condition that radiologists should be able to recognize and consider in the differential diagnosis with other soft-tissue tumors. The imaging modalities (X-ray, ultrasound and CT) are of no help in this task due to their intrinsic low spatial and contrast resolution [12, 13]. MRI is considered the gold standard among all second-line modalities and is widely used for the preoperative diagnosis and follow-up. MRI enables not only accurate detection and characterization of the lesion but also evaluation of the relations between the tumor and the neighboring structures, including muscles, vessels, nerves, bone and intra-abdominal organs [14].

We report an unusual case of an intramuscular desmoid tumor in the gluteal region presenting as a progressive gait disorder in an otherwise healthy woman, in whom removal of the tumor after years lead to resolution of the abnormal gait and return to normal function.

The patient gave informed consent and was informed that data concerning the case would be submitted for publication. We also received an approval from our institutional review board.

CASE REPORT

A 39-year-old, otherwise healthy woman, presented to our medical center with left hip pain and a severe gait disorder. Four years prior to her referral she gave birth to a healthy baby boy, by an uneventful normal vaginal delivery. Following the delivery, pain developed gradually in the region of the left hip joint. During the last year she also developed a pelvic tilt and worsening hip pain accompanied by a debilitating gait disorder. Her work-up, in a different country, included physical examination and several imaging studies, including computed tomography (CT) and magnetic resonance imaging (MRI). These were reported negative according to the documents presented by the patient. Three years later and she was referred to our medical center for re-evaluation.

On clinical examination, the patient had a severe waddling gait. She had almost no internal or external rotation of the left hip joint and a 30 degree adduction contracture was noted on left side. She had a functional leg length discrepancy of seven cm and valgus angulation was noted in her left knee. There were no lumbar pain or deformation and no neurological deficits were found. (Figure 1A). Laboratory data, including infectious disease markers (ESR and CRP) were all within normal limits. Electromyography was unremarkable as well. Plain radiographs demonstrated coxa vera and a notable pelvic obliquity that had progressed in comparison with a radiograph preformed two years earlier (Figure 2). Tc-99 Bone scan and lumbar spine MRI were negative.

She was referred for an MRI of the hip which demonstrated a 7x5x5 cm mass in the left gluteus minimus muscle showing low signal intensity on all sequences with a corresponding distortion of the left pelvis (Figure 3). A CT guided biopsy was preformed, yielding connective tissue without evidence of malignancy (Figure 4).

After preoperative planning, a surgical wide marginal resection was conducted. On surgical exploration, a fibrotic, hard-consistency tumor, lying between the gluteus medius and minimis muscles was found, connecting the iliac bone and the greater trochanteric region of the femur. A wide margin surgical resection was achieved, combined with soft tissue release, restoring the lost range of motion (Figure 1B, 2B). Histology of the lesion revealed the diagnosis of a desmoid tumor.

Post treatment clinical follow-up was conducted three weeks and three months after surgery. The patient’s post-operative follow up was uneventful and she had restored to a full range of motion of her hip, experienced no pain and her pelvic gait was normal. She also reported no walking pain or functional limitations.
morbidity. Extra-abdominal desmoid tumors, in the shoulder girdle, chest wall, back and thigh are well recognized [3].

Severe gait disorders and pronounced pelvic obliquity are very rare presenting symptoms of desmoid tumors and to our knowledge have not yet been reported in the literature.

Desmoid tumors usually present as a soft-tissue mass that interrupts the adjacent intermuscular and soft tissue planes. Computed tomography (CT) is of limited use although it can demonstrate asymmetry, presence of a mass, and evidence of bone erosion, if present. On MRI, the tumor may be either hypointense or hyperintense relative to surrounding muscle on both T1- and T2-weighted sequences; also, heterogeneous changes are common. MRI is an excellent means both for outlining the extent of these lesions at presentation and for surveillance following diagnosis and treatment [2].

Until recently wide surgical resection was the main treatment modality, despite high local recurrence rates. Furthermore, even after free margin resection, when the patient was considered disease free, major cosmetic and functional impairment were occasionally noted [2–6]. Operations that preserve function and structure are the primary goal [10]. Based on several studies, the modern treatment is based on a multidisciplinary approach depending on the tumor location, extent and aggressiveness. Wide-margin surgery has been the primary treatment in the past several decades. This has been true despite the high recurrence rate following resection of desmoid tumors. This recurrence rate is among the highest reported for any tumor commonly undergoing surgical resection. In recent years, adjuvant treatment has evolved and now presents a viable alternative to the traditional modalities of wide resection and radiation. In a young patient (<40 years), wide margin surgery and high-dose radiation are unnecessarily morbid in light of the clinical context of a benign tumor [1].

Pharmacologic and biologic treatment approaches are being developed for patients with tumors adjacent to the airway in the neck or upper thorax, radiotherapy is recommended due to the potential of mortality for patients going to wide-margin surgery. Otherwise, since tumor progression rarely causes death, one must consider whether the morbidity of treatment would outweigh the morbidity of disease progression [8].

The patient underwent wide surgical resection of the tumor with no local morbidity, regaining the lost hip-joint range of motion and functional improvement. No adjuvant radiation or chemotherapy was indicated.

Desmoid tumors may have a capacity of self-limitation thus conservative therapy should be considered in symptom-free patients [9]. Follow-up MR imaging of desmoids indicates natural regression of desmoids and more aggressive behavior of recurrences, which may justify a more conservative therapeutic approach. Detection of desmoid tumors on MR images is important for the determination of tumor extent, which allows to decide the surgical method and predict postoperative recurrence [15–18].

DISCUSSION

Desmoid tumors are a benign group of soft tissue tumors with a variable local aggressiveness and
CONCLUSION

We presented a case in which a woman developed severe gait disturbance that was secondary to an intramuscular gluteal desmoid tumor. The multidisciplinary approach including MRI and a CT guided biopsy and the surgical resection restored her normal gait.

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Author Contributions
Tenenbaum Shay – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Hershkovich Oded – Substantial contributions to conception and design, Acquisition of data, Writing the article, Final approval of the version to be published
Shabshin Noga – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Chechik Aharon – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Liberman Boaz – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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