Lipofibromatous hamartoma of sciatic nerve: A case report

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ABSTRACT

Introduction: Lipofibromatous hamartoma of the nerve is the fibro-fatty overgrowth within the nerve. Most commonly they occur in the median nerve, ulnar nerves, and a few other nerves but the involvement of the sciatic nerve is very rare. The fibro-fatty infiltration causes palpable neurogenic mass and clinically presents lump, moderate numbness, tingling sensation, and pain in its territory. Magnetic resonance imaging is the gold standard for diagnosis.

Case presentation: We present a case of a 65 years old female, who presented to OPD with a tingling sensation which progressed to pain in the gluteal region and was associated with a tender swelling. MRI showed a giant space-occupying lesion in the sciatic nerve course. The mass was excised and then sent to the histopathological examination which designated the mass as lipofibromatous hamartoma.

Discussion: Unless debilitating, lipomatosis of the nerve doesn’t require any intervention as it is a benign condition. Lipofibromatous hamartoma is attributed to the accumulation of fatty and fibrous tissue in the epineurium. Diffusion-weighted imaging in association with conventional magnetic resonance imaging has increased diagnostic yield. The lesion was iso-intense to the subcutaneous fat and there were fine fibrillar appearances inside of it. Simple mass excision was performed on our patient without complications.

Conclusion: Lipofibromatous hamartoma of the nerve are rare soft tissue tumors of nerves and sciatic nerve involvement is even rarer. Correct and careful interpretation of the MRI findings can lead to diagnosis with ease and help prevent unnecessary biopsies.

1. Introduction

Lipomatosis of the nerve, which is also known as lipofibromatous hamartoma, is fibro-fatty tumor characterized by palpable neurogenic mass as a result of infiltration and proliferation of mature adipocytes and fibrous tissue within the nerve [1]. It is a rarely occurring nerve hamartoma which is usually seen within first three decades of life [2]. Most commonly, lipofibromatous hamartomas occur in the median nerve and its branches, followed by ulnar and few other sites such as radial, digital, cranial and plantar have also been reported [3,4]. However, cases involving sciatic nerve or superficial peroneal nerve are extremely rare [5]. Lipomatosis of nerve presents as lump, moderate numbness, tingling, and weakness in the territory of the nerve involved. Ultrasound (US) and computed tomography (CT) though useful in diagnosis, magnetic resonance imaging (MRI) with high soft tissue resolution is the gold standard [6,7]. Sarp et al. has reported a case report of giant lipomatosis of sciatic nerve with typical and unique MRI findings, where it was managed with internal neurolysis with microsurgical techniques [8]. We present a similar case report of lipofibromatous hamartoma of the sciatic nerve managed by complete excision. This case report has been reported as per SCARE 2020 criteria [9].

2. Case Presentation

A 65 year-old-female presented to the outpatient department of orthopedics with the complaint of a tingling sensation in the left gluteal region for 6 months. It gradually progressed to pain in the left gluteal region which was gradual in onset, mild, deep aching type, and occasionally sharp and shooting type, which was intermittent in nature, with no radiation. It was aggravated by sitting but had no relieving factors. It was associated with swelling in the left gluteal region which was initially the size of a pea, which later on progressed to be the size of a table tennis
There was no history of trauma, surgery, chronic back pain, and excessive weight-bearing activities prior to the incident. She was a known case of osteoarthritis of both knees. She had a medical history of hypertension and hypothyroidism and was under medication. There was no significant family history. She was a housewife and mother of 3 children. She was a nonsmoker and non-alcoholic post-menopausal female.

On examination, she was obese with a BMI of 31, well oriented to time, place, and person. There was no pallor, icterus, edema or lymphadenopathy. There was a swelling of approximate size 3 × 3 cm with no overlying skin discoloration. It was deep on palpation without elicitation of the margins, with no fluctuation of the mass. Tenderness was elicited on palpation of the mass.

X-ray of the lumbosacral and gluteal region was performed which didn’t show any significant findings. Consequently, High-resolution 3-T MRI imaging was obtained. The T1-weighted axial (Fig. 1) and coronal (Fig. 3) showed a giant space-occupying lesion of size 3.2 × 3.5 cm located in the sciatic nerve course. The tumor was iso-intense to the subcutaneous fat and there were fine fibrillar appearances inside of it. (Short Tau Inversion Recovery) STIR images axial (Fig. 2) and coronal (Fig. 4) show a signal increase at the fibrillar appearance along with suppression of the fat signals within the tumor. The findings suggested the diagnosis of Sciatic Nerve Lipomatosis.

The next step of management for the patient was the excision of the tumor. Following the excision, the specimen was sent for histopathological examination which was suggestive of lipofibromatous hamartoma. The patient is currently asymptomatic and performing her regular activities.

3. Discussion

Lipomatosis of the sciatic nerve is a relatively rare but benign condition that does not require any intervention unless symptoms are debilitating. Nerve lipomatosis was first described in 1952 by Mason whereas sciatic nerve lipomatosis was first described in 1999 by Marom and Helms [7]. Theories have it that there is no known cause or hereditary predisposition for this lesion, but the hypertrophy of mature fat and fibroblasts in the epineurium has been postulated [6]. History of trauma and chronic nerve inflammation is also found to be associated with lipomatosis of the nerve [10,11]. The clinical features are not specific as some remain asymptomatic till late life whereas some become symptomatic early and present with symptoms like tingling, numbness, swelling, and pain [12]. Our patient initially had occasional numbness in the left gluteal region. However, it progressed to pain as the mass gradually increased in size.

Lipomatosis of the nerve is diagnosed with typical magnetic resonance imaging findings of a striking increase in adipose tissue in a fibrous network that involves the peristeum, bone marrow, nerve sheath, muscles, and subcutaneous tissue. In our patient as well, MRI features were suggestive of lipomatosis of the nerve. A conventional magnetic resonance imaging can be of help but is of low specificity, so diffusion-weighted imaging in association with conventional magnetic resonance imaging improves the diagnosis [1]. On T1-weighted images, the aberrant fatty tissue around the nerve fibers and fusiform expansion of the nerve appears spaghetti-like in longitudinal section and coaxial cable-like in cross-section, and they are usually pathognomonic for
lipomatosis of the nerve [7,8]. Despite deviations in the characteristic appearance of lipomatosis of the nerve, the fat interdigitating between the fascicles within the sciatic nerve remains the most important differentiator and confirms the diagnosis [13].

On histological examination, the individual nerve fascicles are segregated from each other by a large number of adipocytes and muscle fibers between them [13]. However, a biopsy of the nerve is contraindicated in lipomatosis of the nerve, as it may cause the motor and sensory deficits. Hence, correct MRI interpretation is crucial for the diagnosis in cases of lipomatosis of the nerve [8]. Also, several atypical features can be present in the MRI as described by Wong et al., due to the variability in the distribution and extent of the abnormal fatty deposition and the odd locations of the lesion [13]. Thus, every radiologist, as well as treating surgeon, must be familiar with the typical and atypical patterns of clinical presentation as well as imaging features for better and timely care of the patients. Complete mass excision is usually performed for cosmetic purposes [1]. Conservative treatments and invasive surgery, including debulking, external or internal neurolysis, or radical excision with or without fascicular grafts, have been described [2,14]. As described by Ying-Ling et al., conservative excision with preservation of the nerve function is recommended when internal neurolysis with meticulous microdissection under high magnification is unavailable or impossible [15]. In our patient, simple excision of the mass was performed and the patient had a good clinical outcome without complications. The specimen was sent for histopathology which was suggestive of the lipofibromatous hamartoma of the sciatic nerve.

4. Conclusion

Lipofibromatous hamartoma of the nerve is a rare soft tissue tumor with the sciatic nerve being a rare site among them. MRI findings are usually diagnostic of the disease and need correct interpretation of the typical as well as atypical features to diagnose it. A biopsy is not recommended to prevent possible nerve damage. Simple excision can also provide a good clinical outcome.

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