CASE REPORT

Neurofibrolipoma of lower extremity: Report of a rare case

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Abstract

A lipoma is one of the commonest types of benign tumor and a neurolipoma is one of its rarer variants. It is also called as Neurofibrolipoma (NFL) or lipomatosis of nerve. It can occur in a person with neurofibromatosis. The most common sites of occurrence are the volar aspects of the hands, wrists, and forearms. These NFLs are observed in young people, usually men. The median nerve is most commonly involved. Lower-extremity cases are extremely rare. We report here a rare case involving the lower limb without any skeletal deformities occurring in a young man.

Keywords: Lipoma, Neurolipoma, Neurofibrolipoma, Neurofibromatosis, Lower Extremity

Introduction

Neurofibrolipoma (NFL) is a known pathological entity. It is identified by various different names such as lipofibroma of nerve or lipomatosis of nerve or fibromatosis of nerve or fibrolipomatous hamartoma of nerve. It is considered as a true hamartoma as it has all three fibrous, neural and fatty components. Its exact etiology still remains obscure though. NFL is considered as benign hamartoma that are seen in close proximity of nerve wrapped on soft tissue sheath mainly involving peripheral nerves. They may be painful or painless. Histopathological features of NFL are characteristic. There is fibrofatty infiltration within the affected nerve and perineural fibrosis which is also very peculiar. Median nerve is most commonly affected followed by ulnar and radial nerve. Lower extremity involvement seldom occurs. Here, such a lower extremity NFL case is being reported.

Case Report

A 30 year old male presented with a swelling on the left leg for the last 2 years. There was no history of any fever. It was not associated with any pain or tenderness. The swelling was insidious in onset and gradually increased in size. There was no history of trauma or restricted mobility. The patient complained of skin paresthesia and tingling sensation along the areas of the swelling. Past history, family history, or personal history revealed nothing significant. On general examination, patient had mild pallor. There were multiple neurofibromatosis type lesions all over the body as shown in Figure 1. On local examination, the swelling was solitary, measuring 4 cm × 3.5 cm × 2 cm situated at the postero-lateral aspect of the left leg. It was non mobile; soft in consistency, temperature was normal and the overlying skin was normal. Rest of the parameters was within normal limits. The systemic examination was unremarkable. On investigating, local sonography of the lower extremity conformed a cystic swelling at the site. Magnetic Resonance Imaging (MRI) was planned but as the patient was not affording at all so we decided to go ahead with the plan of excision biopsy of the lesion.

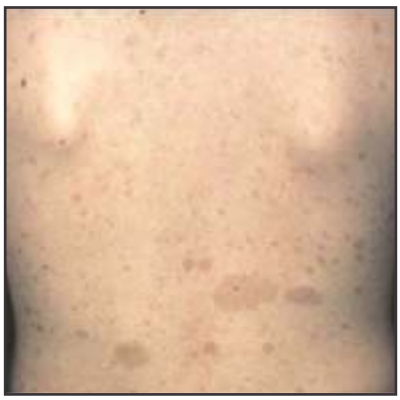

Figure 1: Back lesions on general examination

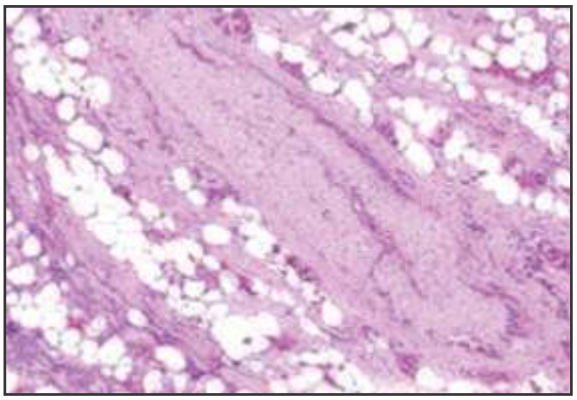

Figure 2: Histopathology image of NFL

Operative details

An incision was made over the swelling on the postero-lateral aspect of leg longitudinally over the swelling extending from the ankle to knee level. As the incision deepened, long swelling extending along the course of sural nerve was found. Clear areas of nodes of Ranvier of the nerve were visible. The proximal end of the nerve was found to enter the muscle. The entire nerve thickening was completely dissected out from the surrounding tissue. Hemostasis secured and washes given. The closure of the subcutaneous layer was done with vicryl 2.0 and closure of the skin was done with the skin stapler.

Postoperative condition

Postoperatively, the patient was in a stable condition. There were no motor complications or paralysis resulting from the procedure. There was no seroma or local hematoma formation either. The skin staples were removed on the 10th postoperative day. Patient had a normal walk without any pain.

Microscopically, it showed admixture of mature elements comprising of adipocytes in lobules with interspersed capillary sized blood vessels along with nerve bundles, thick walled blood vessels, and fibro-collagenous tissue (Figure 2). All these features suggested it as a NFL. We followed up our patient for one year and did not find any local complication or any recurrence however, later we lost the patient to follow up as he migrated to other state.

Discussion

NFL tumors can easily be mistreated initially considering the swelling to be a simple lipoma [1]. But one may get surprised when intra-operatively it is within the sural or the peroneal nerve all along the course of the nerve [2, 4]. Here, the MRI of the lesion would have given us better guidance which we missed due to patient's financial constraints. The person was also suffering from NF. He had extensive NF type lesions all over the body including the face. From the experience gained from this case and from a similar earlier case, any lipoma like swelling arising in a patient with neurofibromatosis lesions should be suspected as a NFL [3].

Current recommendation for neurolipoma is that they can be left alone because inadvertent transaction of nerve when the neurofibrolipoma is arising from a major motor nerve can lead to motor

deficits. Here we took care not to transact the nerve though it was arising from the sural nerve and carefully dissected out a major portion of the neurolipoma leaving some nerve tissue intact. Since sural nerve is a cutaneous sensory nerve no motor deficits arose postoperatively. As the lesion is benign it has good prognosis. NFL affects both upper and lower extremities. The exact pathogenesis of this entity is not known. Some of these tumors are hereditary and some of them are not. Timely investigations are useful [5]. Common differential diagnoses are lipoma, vascular malformation, ganglion cyst and traumatic hematoma. MRI remains the investigation of choice for isolating this pathology as many minute features can be easily delineated such as coaxial cable-like appearance on axial sections, and a spaghetti-like appearance on coronal images [6]. Antecedent trauma and chronic nerve irritation are seen in few cases in which the painless multiple swellings are present since birth all over the body and which usually shows no growth. Here, we compare it with one similar report of sole schwannoma of foot [7]. Few have reported involvement of sciatic nerve which showed marked limitation of patient's movements [8]. Also, sometimes it is observed along with some congenital abnormalities like Klippel-Trenaunay syndrome [9]. Plus as mentioned earlier, median nerve being most commonly involved one may also find such type of hamartoma in patients [10].

The treatment options for neural fibrolipoma can pose a dilemma. They can be treated either conservatively or surgically. Surgical treatment includes debulking, external or internal neurolysis, or radical excision with or without fascicular grafts. The treating surgeon needs to individualize the exact option for his/her patient. Follow up of these cases is crucial. One has to also keep a yearly/half yearly follow up of such patients to note for any delayed complications or local recurrence.

Conclusion

It is always prudent to examine the patient well clinically, and then investigate using best of the available modalities, here being the MRI of local part and then plan accordingly the excision biopsy taking care not to damage the encasing nerve which will lead to parasthesia or if it's a motor nerve then motor deficit. Careful and meticulous dissection will prevent the local complications. Such cases should not be mistreated thinking about it as just lipoma or they may complicate or recur.

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