

A Case of Nevus Lipomatosus Superficialis in a 14-year-old Filipino Female*

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Abstract

Introduction: Nevus lipomatosus superficialis is a rare benign hamartomatous skin lesion. It is classified into 2 types: solitary and multiple. Lesions consist of multiple, flesh-colored or yellow papules and/or nodules in a segmental pattern, with a linear, zosteriform, or along the lines of skin folds distribution. Surfaces are smooth, but may also have verrucous or cerebriform appearance. Pathogenesis is unknown, but there is speculation that precursor cells around dermal blood vessels give rise to mature fat cells in a mosaic pattern. Incidence is presently unknown, but this is the first reported case in this institution. Histopathology reveals presence of aggregates of mature adipose tissue among the collagen bundles of the dermis.

Case Summary: DF is a 14-year-old, Filipino, female, who consulted due to multiple, pedunculated, soft papules over the right lower back, and extending to the right flank, with a clustered arrangement. There was no history of trauma or manipulation, nor was there pain, pruritus, or other associated symptoms. Past medical, family, and social history were non-contributory. Initial impression was acrochordon. Excision biopsy revealed mature adipose tissue in the dermis, which is diagnostic for nevus lipomatosus superficialis. Serial excision was done.

CONCLUSION: Excision is the treatment of choice. Other treatment options that can be explored are CO₂ laser, cryotherapy, and intralesional injection of phosphatidylcholine, which yield promising results recommended by other studies. Dermatopathology plays a vital role in the diagnosis of this condition. A high index of suspicion, a good clinical eye, and dermatopathologic analysis are essential tools in clinching the diagnosis.

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INTRODUCTION

Nevus lipomatosus superficialis, also known as nevus lipomatosus superficialis of Hoffman and Zurhelle, is a rare benign hamartomatous skin lesion. It is divided into 2 subtypes: solitary and multiple. The solitary type presents as a single, soft, pedunculated papule located in any part of the body. The multiple type appears within the first 2 decades of life, where lesions consist of multiple, grouped, flesh-colored or yellow papule and/or nodules in a segmental pattern. In this report, we describe the first known case of nevus lipomatosus superficialis in a 14-year Filipino female, which was managed by excision of lesions.

CASE REPORT

This is a case of a 14-year-old Filipino female from Quezon City who sought consult due to appearance of pedunculated papules over the right lower back.

Two years prior to consult, patient noted appearance of skin-colored papules associated with intermittent pruritus starting on the right lower back, which later involved the right flank. Papules were gradually noted to increase in number and size, with a clustered arrangement. There was neither history of trauma, manipulation, pain, loss of sensation, nor other associated symptoms. No intervention or consult done. Lesions persisted, hence consult.

The review of systems, past medical history, and family history were unremarkable.

Physical examination was unremarkable. Cutaneous examination showed multiple, ill-defined, clusters of linearly arranged, round to irregularly-shaped, soft, skin-colored papules and nodules over the right flank.

The initial working impression was acrochordon. Differential diagnoses include linear verrucous epidermal nevus, neurofibromas, and nevus lipomatosus superficialis.

The plan was to do excision of the lesions, and was explained thoroughly to the patient and her mother. Informed consent was obtained prior to the procedure. Excision site was marked with a pen. Skin surface was cleansed using povidone-iodine solution, which has a broad antimicrobial spectrum, including fungi. The sterile area was covered with a drape, followed by local instillation of anesthetic agent (lidocaine-epinephrine). A fusiform incision was done,

and excision of lesions done. Specimen was sent out for histopathology reading. Subepidermal layers were closed with interrupted dermal stitch using vicryl 4-0, and epidermal layers were closed with prolene 4-0 using running stitch. She was prescribed with Cefalexin 500mg/tablet, 1 tablet every 4 hours for 7 days, Mefenamic acid 500mg/tablet, 1 tablet every 8 hours as needed for pain. She was also advised proper daily wound care and to come back after 1 week for removal of sutures.

On third consult, patient came in for follow-up. She was conscious, coherent, ambulatory, and was compliant with prescribed medications and instructions. Cutaneous examination revealed a solitary, well-defined, linearly-shaped, surgical wound over right flank. Wound dehiscence was noted. Histopathologic examination of the excised specimen revealed an exophytic polyphoid structure with papillomatosis and deposition of mature adipose tissue in the dermis. These features are consistent with a diagnosis of nevus lipomatosus superficialis. It is a benign condition, and treatment was done for cosmetic purposes. Removal of sutures was done. Na fusidate 250mg/tablet, 1 tablet 2x a day for 7 days was prescribed, as well as compression dressing. She was advised to come back after 1 week for follow-up.

DISCUSSION

Nevus lipomatosus superficialis, also known as nevus lipomatosus superficialis of Hoffman and Zurhelle, is a rare benign hamartomatous skin lesion¹. It is divided into 2 subtypes: solitary and multiple. The solitary type presents as a single, soft, pedunculated papule that may be located in any part of the body, and is usually seen in individuals in the 3rd-6th decade of life. The multiple type appears within the first 2 decades of life, and consists of multiple, grouped, flesh-colored or yellow papule and/or nodules in a segmental pattern. Distribution of the lesions is usually linear, zosteriform, or along the lines of the skin folds. Surfaces are smooth, but may also have verrucous or cerebriform appearance. Pathogenesis is unknown, but there is speculation that precursor cells around dermal blood vessels give rise to mature fat cells in a mosaic pattern¹. Ectopic fat cells in dermis are derived from perivascular mesenchymal tissue. Histopathology reveals presence of aggregates of mature adipose tissue among the collagen bundles of the dermis². Nevus lipomatosus superficialis is a benign condition, and management is done for cosmetic purposes³. Excision is the treatment of choice¹. Since the patient is female and an adolescent, she is

conscious of the unsightly appearance of the lesions, hence she opted for excision. The plan is to do serial excisions because of the number of lesions. Excision biopsy was done on the first session, which was both diagnostic and therapeutic. Subepidermal layers were closed with interrupted dermal stitch using vicryl 4-0, and epidermal layers were closed with prolene 4-0 using running stitch. Polyglactin 910, also known as vicryl, is a synthetic braided copolymer of glycolide and L-lactide and is absorbable. It has good ease of handling, fair knot security, and low tissue reactivity that makes it a good choice for subcutaneous closures¹. Polypropylene (prolene) is a non-absorbable suture that has high plasticity and stretches with tissue swelling, which reduces the likelihood of postoperative track marks¹.

Patient came back for consult three weeks after the scheduled follow-up. Upon physical examination, there was a solitary, well-defined, irregularly-shaped surgical wound over right flank. Wound dehiscence was noted. There were no symptoms of bleeding, pain or fever noted. Wound dehiscence may be caused by location of the excision site in the body, infection, pressure on sutures, injury to the affected area, weak tissue or muscle at the wound area⁴. In the patient's case, this may be due to the location of the excision site on a highly mobile area of the body, which is the right flank.

Nevus lipomatosus superficialis can also present in a variety of ways. Brasanac et al (2005) reported a case of a 47-year-old female from England that had a history of skin-colored papules and nodules in the sacral region since birth, associated with pain. Resection of lesion was done. Histology showed infiltration of fat cells in dermis with multiple folliculosebaceous cystic hamartomas and dermoid cysts⁵. Kwon et al (2002) reported a case of a 72-year-old male from South Korea with chief complaint of left nasal obstruction of 10 years. On physical examination, there was a round, fleshy, skin-colored mass connected to skin of left nasal vestibule. Excision was done. Histopathology revealed presence of mature ectopic fat tissues embedded in dermis. There was no recurrence of the lesion after 10 months of follow-up⁶.

Other treatment options that can be offered to patients who are unable to undergo surgery are CO₂ laser, cryotherapy, and intralesional injection of phosphatidylcholine. In a study by Jung et al (2007), they treated affected patients with CO₂ laser with staged excisions and covered with local

advancement flaps. Patients were reported to be satisfied with the results, and recurrence was not observed⁷. Cryotherapy was used by Al-Mutairi et al (2005) in a case of a 43-year-old male diagnosed with nevus lipomatosus superficialis. Liquid nitrogen cryotherapy was administered by spray method at 2-week intervals, and after 3 sessions, there was considerable size reduction of lesions⁸. Another treatment option, which is not available in the Philippines, is the use of intralesional phosphatidylcholine. Phosphatidylcholine is a class of phospholipids that incorporate choline. It is a major component of biological membranes, and can be obtained from a variety of sources, such as egg yolk and soybeans. Intralesional injection with phosphatidylcholine has been initially used in the treatment of atheroma plaques in cardiac diseases. Hexsel and Serra (2003) reported their experience on the use of 250mg/mL phosphatidylcholine injections in the treatment of localized fat deposits among patients with a minimum interval of 1 week and mean interval of 15 days between injections. Results showed that phosphatidylcholine was effective in reduction of fatty areas⁹. Similar studies done by Kim et al (2011), Rittes (2009) and De Paula et al (2009) also showed promising results of with intralesional injection of phosphatidylcholine^{9,10,11}. These treatment options can be fully explored provide alternative treatment options for patients who are hesitant to undergo excision, and for those with large areas of affectation.



Figure 1. A 14-year-old Filipino female presented with multiple, ill-defined, clusters of linearly arranged, round to irregularly-shaped, soft, skin-colored papules and nodules over the right flank



Figure 2: Excision, which is both diagnostic and therapeutic, was done



Figure 3. Surgical site closed with prolene 4-0

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