INTRODUCTION

Nevus lipomatosus cutaneous superficialis (NLCS) is a rare, benign hamartomatous lesion characterized histologically by the presence of ectopic adipocytes in the dermis. This condition was first reported in 1921 in a German journal for dermatologic and venereal diseases. Such lesions are often reported classically as Hoffman–Zurhelle lesions—the namesake of its founders. They are described as multiple, pedunculated lesions presenting en masse. The other, rarer clinical variant, is characterized by a singular rounded papule with no stalk. Typically after the second decade of adult life, they are uncommonly seen with a necrotic or ulcerating appearance. In this report, we describe a unique case of a torted and infarcted NLCS found in the intergluteal cleft of an 18-hour-old female neonate.

CASE PRESENTATION

A Gravida 4 Para 2 Caucasian lady was referred to the Maternal Fetal Medicine service at our center—a tertiary children’s hospital—for an abnormal “soft tissue mass” discovered in her third-trimester antenatal ultrasound. Geographically, she lived >4 hours away and was receiving...
her antenatal care at a local district hospital. There was no relevant past or family history of any skin conditions, and the fetus had an otherwise normal growth. Except for one episode of gastroenteritis, the pregnancy was uneventful for both the mother and the child.

The birth was uncomplicated, and the female neonate had APGAR scores of 9 and 10, respectively. The soft tissue mass was immediately noticed and the surgical team reviewed her. It was noticed as a 1.5 cm × 1 cm dusky pedunculated mass resembling a grape. (Figure 1A,B) Given the geographical location of the patient and the nature of the lesion, our center was contacted for opinion on further management of the lesion. Owing to the physical appearance and location of the lesion, we decided to perform an ultrasound scan (USS). The ultrasound of the mass and the adjacent vertebral bodies was performed, which ruled out any sequelae or association with Spina bifida.

The anechoic shadows in USS proved that there were no growth defects in the vertebral bones. Similarly, the filum terminale (blue markers) and cauda equina (orange arrow) were both successfully identified with no obvious defects. The soft tissue mass was confidently thought to not contain any meningeal or neural tissue. (Figure 2A,B).

After reviewing the scans, surgical management was unequivocal decision. It was ligated at the base, transected, and sent for histopathological studies. There was no postoperative bleeding, and no scarring was seen on follow-up.

The histopathological report confirmed the mass to have undergone torsion with infarction of the crown. It had a pedunculated appearance with singular base measuring 15 × 10 × 8 mm and was formally returned with the diagnosis of NLCS. The lesion had skin superficially, with remainder of the specimen containing superficially distributed adipose tissue. The adipose tissue contained areas of significant calcification, suggesting chronicity of the lesion but ruling out any form of malignancy. (Figure 3A,B).

The baby was followed up in 6 weeks and there was no evidence of superficial scarring. She has been developing normally as expected without any need for future surgical follow-up.

3 DISCUSSION

There are many skin conditions which present in neonates. These include vesicles, pustules, bullae, and erosions. The features of NLCS underscore its clinical relevance that it can sometimes be mistaken for some other skin condition. Certain systemic conditions such as Goltz syndrome can often present with skin lesions closely resembling NLCSs. Other lesions, such as syphilitic condylomata, can present similar to ulcerated NLCS. This may be why it was first reported in a journal of venereal diseases.

The Fitzpatrick phototyping scale is a numerical classification system for human skin color. It is a recognized tool for dermatological research into human skin pigmentation. While there is no known increased affinity for specific Fitzpatrick skin types, a brief search of NLCS articles yielded case reports largely from countries bordering the South China Sea or countries around the Mediterranean Sea.

While the tendency for NLCS to ulcerate remains low, instances where it has ulcerated have resulted in a symptomatic presentation (i.e., with pain or with pustular exudate). This, in turn, prolongs surgical treatment and affects the healing process. However, surgical management appears to be the hallmark of effective management. Other methods of removal, such as CO₂ laser ablation—although non-invasive—have shown to be a temporary fix at best. The same evidence also purports that a recurrence can sometimes come with a proliferation of more lesions.

This case is highly unique as there are a mere handful of reports of such lesions detected in neonates in the world. The histopathology suggested signs of chronicity secondary to the presence of calcium deposits. This implies the lesion may have formed in the early part of
fetal development. Considering the above, it could resemble calcinosis cutis, resulting in an auto-immune or metabolic disorder. The likelihood is low given the lack of any other systemic conditions.

The lesion was infarcted due to it twisting on its stalk in utero. This most likely halted its growth. Usually, the classical NLCS lesions present in clusters with a pedunculated appearance with a predilection for the pelvic regions. In our case, the pedunculated appearance and location in the pelvic region strongly suggest it is likely to be of the Hoffman–Zurhelle subtype.

4 | Conclusion

There are no current official guidelines on how to manage such lesions. It seems appropriate that a systematic review be conducted for developing a universal management guideline. Perhaps, an attempt to run this may shed light on the causes of this condition. It may also yield a starting point for control variables to be measured, such as race, gender, or skin type.

However, at this point, we would recommend, as best clinical practice, the resection of similar-looking lesions in the pediatric population to prevent any chance of ulceration or calcinosis cutis. Surgical management, thus far, has resulted in a very small rate of recurrence. Lesions should also be routinely sent for histopathological evaluation for the accurate diagnosis of NLCS from other, more serious conditions in which a skin lesion may play a sentinel role.

Acknowledgement

None.

Conflict of interest

No conflicts of interest for any authors to report.

Author contributions

Susmit Prosun Roy involved in supervising, writing up, editing and reviewing contents and images for the report. Caleb Ting involved in writing up and editing of images for the report. Tony Dill involved in interpretation of pathological images and diagnosis. Gerard Roy served as treating surgeon and supervisor of writing up of the report.
CONSENT
Written consent has been obtained from the patient’s mother for publication of the case report and sharing of the images in a de-identified manner.

DATA AVAILABILITY STATEMENT
The data that support the findings of this study are available on request from the corresponding author. The data are not publicly available due to privacy or ethical restrictions.

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