

## Nevus of Hoffmann-Zurhelle: A Case Around the Right Parotid Duct

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**Abstract.** *Nevus lipomatosus cutaneus superficialis (NLCS) is an extremely rare hamartomatous disorder comprising of ectopic mature adipose tissue. The lesions take the form of large, slow-growing, sessile or pedunculated tumors. We report the case of an 11-year-old boy with NLCS, dating back many months, that was growing as a cuff around the parotid duct. The lesion was successfully removed by diode laser excision (980 nm) and a diagnosis of NLCS was rendered. At one-year follow-up the tumor has not recurred. To our knowledge this is the first report of NLCS in the oral cavity; the characteristic clinical and morphological features aided in the diagnosis of this hamartoma. The peculiarity of this case is its location; the extreme rarity of NLCS, in general, probably makes this case unique.*

Nevus lipomatosus cutaneus superficialis (NLCS) is an idiopathic hamartomatous disorder characterized by an ectopic mature adipose tissue in the dermis, which was first described by Hoffmann and Zurhelle in 1912 (1). The lesions usually appear at birth, or during the first 2-3 decades of life. Neither gender preference nor familiarity has been reported (2).

Clinically, two variants have been described: classical (Hoffman-Zurhelle) and solitary. The classical variant consists of multiple, asymptomatic, soft, yellowish lesions that may coalesce to form a plaque with smooth, wrinkled, orange-peel appearance, or with cerebriform surface, with or without the presence of comedones and follicular orifices (3, 4).

The solitary type appears between the third and fifth decade of life and manifests as an isolated, domed, or sessile papule. The base is broader than of the common skin tag (acrochordon).

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Lesions are usually unilateral with a linear or zosteriform pattern, and can occur anywhere on the body, with a predilection for the abdomen, back, buttocks, hips, upper posterior thigh and pelvic girdle (1-12). Atypical sites include the face, auricle and neck (13). Lesions can occasionally reach 20×30 cm in size, forming linear plaques along the skin folds (8). They may be associated with café-au-lait spots, hypopigmented spots, presence of hair, with ulceration and necrosis; Fordyce's angiokeratomas have also been reported (14). To our knowledge, this is the first report of NLCS in the oral cavity. The clinical and histopathological profile is described.

### Case Report

The patient, an 11-year-old boy, was referred to San Martino Hospital, Genoa, Italy for a nodular lesion measuring 1x1 cm in the buccal mucosa. It was yellowish in color and of round shape, surrounding the right parotid duct; it was sessile, mobile on underlying layers, not painful, and of pinkish-yellow color (Figure 1). It had been present for some time and had been gradually enlarging; the patient was unable to specify for how long.

The patient was in good general health, with blood chemistry in the normal range, anesthesiological risk ASA 1. The patient was treated with laser surgery with diode laser at 980 nm. Excision of the lesion was under conscious sedation through nasal inhalation of a mixture of nitrous oxide and oxygen (50%-50%, Matrix – Digital DMD; Reinhold, Italy), and the surgical piece was sent for histological examination.

*Surgical procedure.* Surgery was performed with a diode laser at 980 nm, under conscious sedation, with a continuous flow of nitrous oxide and oxygen of 8 l/min. In the surgical area, local anesthesia was achieved with carbocaine 2% and adrenaline 1:100,000, using a 30 G needle. Using a Zeiss x 3.6 binocular microscopic enlarger, an excisional biopsy was taken with a diode laser at 980 nm (DM 980, DMT, Lissone, Italy), 320 micron fiber, power setting 2.2 W in continuous wave mode (CW), under guidance of a button probe



Figure 1. Clinical photograph shows a solitary, sessile, tumor on the right parotid duct. It is pinkish-yellow in color, with a diameter approximately 1 cm.



Figure 2. Excision of the lesion with diode laser, 980 nm, 320 micron fiber, 2.2 W power, in continuous wave mode, under guidance from a button probe introduced into the parotid duct, note that the operative field is blood-free, giving optimal visibility during surgery.

introduced into the parotid duct; this provided a fundamental landmark during surgery (Figure 2). Detachment was achieved under laser light, and following the cleavage plane in the connective tissue around the outlet of the right parotid duct, the neof ormation was excised in a blood-free operating field. Hemostasis was achieved, and the patency of the duct was checked by means of a distal compressive maneuver and release, which showed normal return to salivary flow.

Conscious sedation lasted for approximately 20 minutes, and the patient suffered no side-effects; oxygen saturation, checked by means of a pulse oximeter, remained above 98% for the entire duration of surgery. Having achieved hemostasis, no suture stitches were placed; a 0.2%

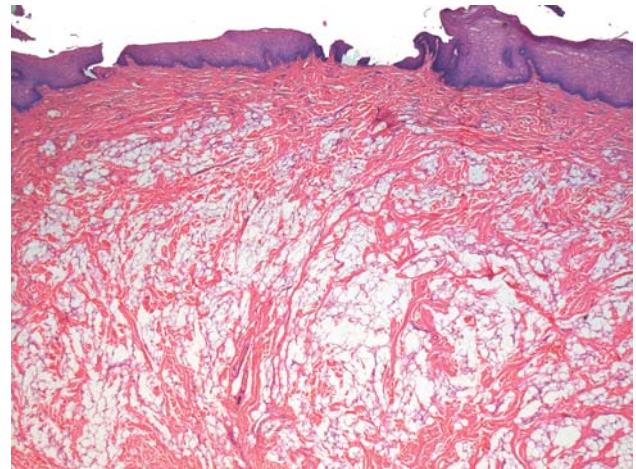


Figure 3. Thinning of the papillary and reticular dermis with a marked increase of adipose tissue. Scattered lobules of fat cells entrapped between bundles of dermal collagen fibers (hematoxylin and eosin stain; original magnification  $\times 100$ ).

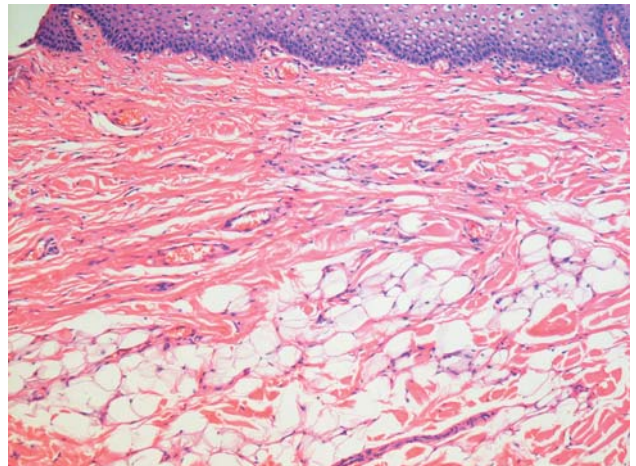


Figure 4. High-power view of the tumor exhibiting enlargement and the increase of adipose tissue (hematoxylin and eosin stain; original magnification  $\times 200$ ).

chlorhexidine gel was applied, and the patient instructed to repeat applications twice daily for 10 consecutive days after surgery.

**Histology.** Surgical pathology specimens were sent to San Martino Hospital, Genoa, Italy. Sections were cut and prepared by conventional routine, and stained with hematoxylin and eosin and by other appropriate methods. Histologically, the specimens showed a slightly acanthotic epidermis with focal flattened rete ridges. Both the papillary

and reticular dermis contained scattered lobules of fat cells entrapped between bundles of collagen fibers and around blood vessels (Figures 3 and 4). Based on these findings, a diagnosis of lipofibroma was established. The nodule was completely excised and there has been no recurrence after one year.

## Discussion

The NLCS is an extremely rare lesion; we report on a case that, to our knowledge, is unique in the literature. Diagnosis can be made by clinical examination based on the characteristic appearance of NLCS, and confirmed by histopathology. Histopathological examination in NLCS cases generally shows isolated groups of ectopic mature adipocytes within the upper reticular dermis, characterized by the presence of ectopic mature adipose tissue in close proximity to blood vessels in the dermis. Additional features are changes in the connective tissue, blood vessels, and skin appendages (1-3).

The proportion of adipocytes in NLCS lesions is reported to vary from 10% to 70%. Electron microscopic findings support the vascular origin of fat cells and differentiation into mature fat cells that are comparable to those seen in fetal lipogenesis (15).

Differential diagnoses includes lipoma, localized scleroderma and dermal nevus. However, histopathological examination clearly distinguishes these three entities. Lipomas appear as well-circumscribed subcutaneous neoplasms of mature adipocytes that lack the typical cerebriform macroscopic appearance (16, 17). Localized scleroderma can be distinguished from NLCS by the markedly thickened collagen bundles that replace the eccrine glands with their surrounding adipose tissue, and the associated moderate lymphocytic infiltrate. The giant congenital intradermal nevus is composed of proliferating melanocytes along the dermo-epidermal junction or the dermis (19). Ultrastructural studies show adipocytes containing large intracytoplasmic lipid vacuoles, often associated with vascular structures (5, 8).

The pathogenesis of NLCS remains unknown, and several theories have been proposed: Hoffman and Zurhelle (1) postulated that fat deposition in the dermis is secondary to degenerative changes (metaplasia) in the connective tissue. Other researchers have suggested that the adipocytes originate from the pericytes of dermal vessels (18), or that the fat cells comprise a true nevus resulting from the focal heterotopic development of adipose tissue (19). At present, it is assumed that NLCS develops from dermal primitive perivascular lipoblasts that subsequently proliferate and progress to mature adipocytes (20-23).

Recurrence and progressive growth of NLCS after surgery are rare, but occasionally NLCS may recur and continue to extend for many years (6).

The treatment of choice is surgical excision, but cryotherapy also may be employed (8). Recently, some authors have reported on cases of classic NLCS treated by CO<sub>2</sub> laser (24, 25). Our case was treated with diode laser at 980 nm and at 12 months there has been no recurrence.

## Conclusion

This is the first report of NLCS in the oral cavity, the characteristic clinical and morphological features aided in diagnosis of this extremely rare hamartoma. The laser (Diode 930) treatment is effective and safe for treating lesions of NLCS without recurrence.

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