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Solitary Neurofibroma of the Lateral Canthal Region of the Face Masquerading as a Lipoma – A Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. Authors KM, SG, TMD designed the study, performed the statistical analysis, wrote the protocol and wrote the first draft of the manuscript. Authors KM and TMD managed the analyses of the study. Authors SG and KM managed the literature searches. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Background: Solitary neurofibromas of the maxillofacial region are extremely rare and there are no cases reported in literature so far, overlying the lateral canthus region of the face. This case report aimed to evaluate the clinical and radiological presentation of this lesion and our experience in its management.
Methods: We illustrate a rare solitary neurofibroma in a 50 year old female overlying the left lateral canthus region of her face. A computed tomography described the lesion to be well-circumscribed without causing any bony changes, suggestive of a benign soft tissue pathology. The lesion was enucleated in toto under local anesthesia.
Results: Histopathological analysis revealed interlacing bundles of elongated cells with wavy and dark-stained nuclei, wire-like strands of collagen and immuno-histochemical positivity with S-100 protein, which are the hallmarks of a neurofibroma.

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Conclusions: Based on our experience, the importance of a thorough clinical and radiological investigation in the management of such lesions cannot be over-emphasized. The patient remained free of recurrence on a one year follow up period.

Keywords: Neurofibroma; nerve sheath neoplasms; s-100 protein; solitary; Tumor.

1. INTRODUCTION

Neurofibromas are benign peripheral nerve sheath tumors caused by a combined proliferation of varied elements of a peripheral nerve [1]. These lesions can arise as a solitary tumor, not associated with any syndrome, as multiple tumors, or as a component of neurofibromatosis type 1. [2]. Solitary neurofibromas of the maxillofacial region are slow-growing, soft, asymptomatic, and well circumscribed rare benign neoplasms; diagnosed by the absence of other clinical characteristics associated to systemic disease, seen in the 1-2 decade of life. [3]. The aim of this report was to present a novel case of a facial solitary neurofibroma at the lateral canthal region of the face.

2. CASE PRESENTATION

A 50 year old female patient presented to our out-patient Department of Oral and Maxillofacial Surgery in January 2020, for the evaluation and management of a painless nodule overlying her left lateral canthus region of the face. It was first noticed 10 years ago, when it appeared to be a small papule and it gradually increased to its present size. There was a history of a local trauma associated with the region ten years back and her medical and family history were non-contributory. On inspection and palpation, there was a bilobed, ovoid, soft in consistency, freely movable, non-tender, non-pulsatile, non-pigmented, sessile nodule; measuring approximately 1-1.5 cm diameter Fig. 1. Locally, there was no sensory or motor deficit and the surrounding skin was normal. Her general physical examination examination revealed no similar pigmentation or nodules. A provisional diagnosis of a lipoma was made and further investigations advised.

A contrast enhanced multi-detector Computed Tomography [CT] scan was done that revealed a well-defined, hypodense, non enhancing lesion adjacent to the outer canthus of the left eye measuring 1.4 x 1 x 1.1 cm wherein, intermittent fat density was noted within the lesion and no associated bone erosion Fig. 2.

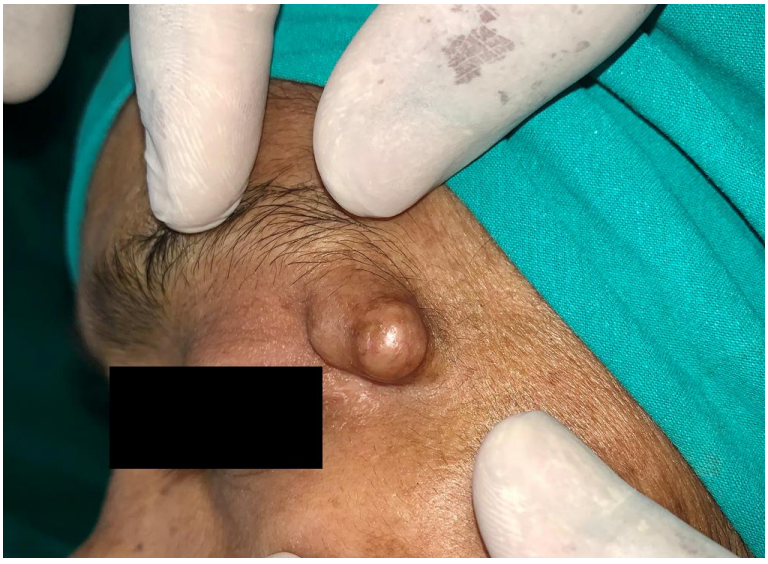

Fig. 1. Pre-operative clinical photograph of the lesion. Note its bilobed and white shiny appearance

Owing to its limited size on inspection and confined anatomic boundaries on Computed Tomography evaluation, we decided not to perform a fine needle aspiration cytology of the lesion. Therefore, an excisional biopsy of the lesion was carried out under local anesthesia as a day care procedure, as per standard operative protocol. Prior to skin incision, a ring field and hydro-dissection nerve block was given using 2% lignocaine and 1:100000 solution. A linear incision was placed on the summit of the nodule, a blunt sub-dermal dissection was carried out to the base of the lesion followed by enucleation in toto with no peri-operative complications and the specimen was submitted for a histopathological analysis Fig. 3.

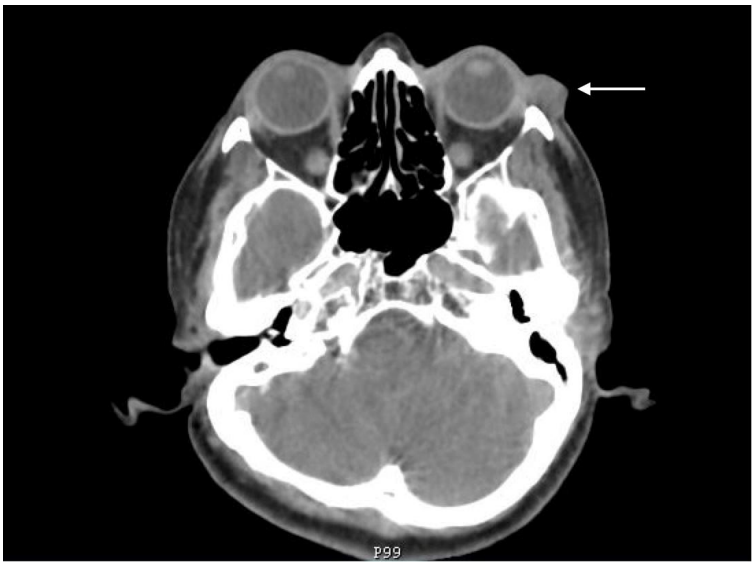

Fig. 2. A contrast enhanced axial CT Scan section demonstrating a well-defined, hypodense, non-enhancing, solitary lesion [arrow] adjacent to the outer canthus of the left eye, with no underlying bone involvement

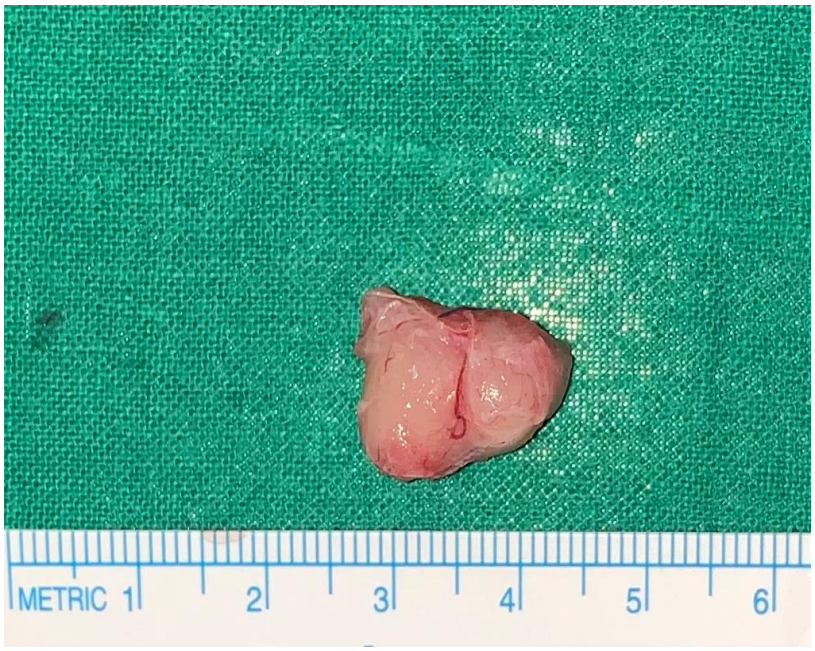

Fig. 3. The specimen was 1.5 cm x 1 cm, flesh coloured, soft nodule

Excessive skin was trimmed and a tension free, cosmetically acceptable, primary closure was achieved with 6-0 non resorbable nylon sutures Fig. 4.

Microscopic examination showed bi-lobed, completely encapsulated fibro-cellular lesional tissue with proliferation of randomly arranged spindle shaped cells, interspersed with delicate intertwining collagen fibre bundles and many blood vessels Fig. 5. At the periphery, the cells were arranged in fascicles. The spindle shaped cells showed elongated, wavy nuclei.

Also, immuno- histochemical analysis of the spindle-shaped cells revealed positive reaction with S-100 protein Fig. 6. The histopathological diagnosis was a solitary neurofibroma. Surgery resulted in a satisfactory cosmetic or functional outcome without neuro-sensory changes and no recurrence at a 1 year follow up period.

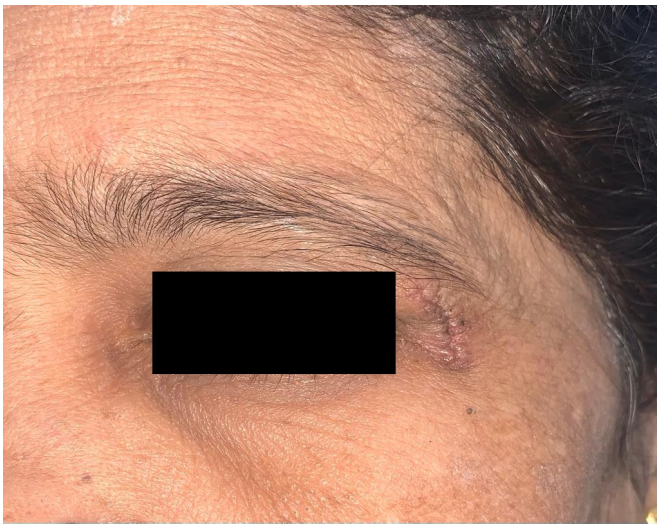

Fig. 4. Post-operative clinical photograph on Day 7 after the removal of sutures

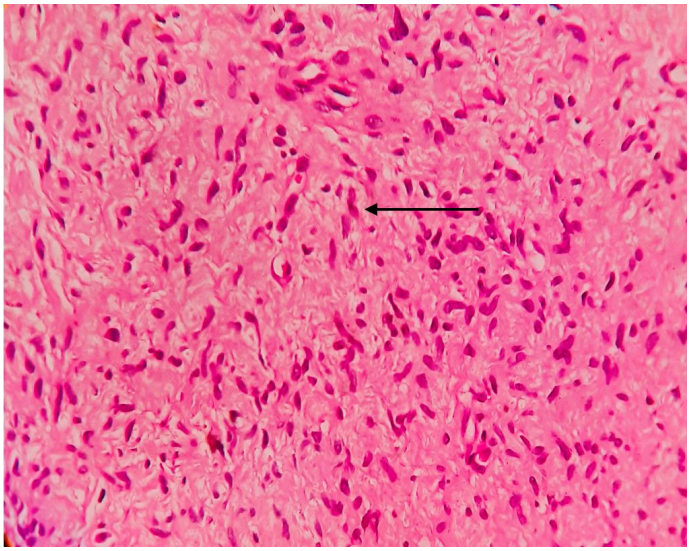

Fig. 5. Hematoxylin and eosin stained section under 40x magnification showed the classical appearance of well delineated haphazardly arranged delicate spindle shaped cells [arrow]with elongated, wavy nuclei intertwining with connective tissue fibrils

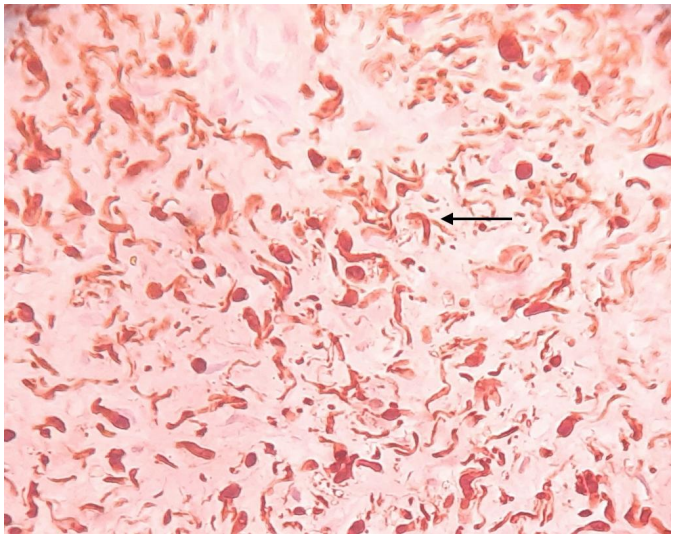

Fig. 6. Immunohistochemical analysis of the spindle shaped tumour cells showed strong positivity for S-100 protein

3. DISCUSSION

Neurofibromas represent a focal proliferation of neural tissue within the dermis, as soft, protruding papules and nodules; and less often, as deep, firm, subcutaneous nodules [4]. Superficial, cutaneous lesions of the face cause a visible mass that usually assumes three growth patterns: solitary, diffuse, or plexiform.[5] of which solitary neurofibromas commonly arise from cutaneous nerves [6]. They are usually well-delineated, rubbery and firm lesions with a white and shiny appearance [7]. The etiology of solitary neurofibroma is attributed to it being a hyperplastic hamartomatous malformation rather than a neoplastic disease [8]. The pathogenesis of solitary neurofibroma is still not well established, however Da Rosa et al. [9], suggested the association between local trauma being a possible etiologic factor for solitary neurofibromas which is in accordance with our case where history of local trauma was reported by the patient 10 years back.

In a review conducted by Maracchio et al. [8] for a total 43 cases of head and neck neurofibroma; where in 22 (51.2%) presented as isolated lesion, involving tongue, buccal mucosa, palate, gingiva, maxillary bones, mandible, chin, neck, and salivary gland. None of the papers documented in literature describe this entity in the lateral canthal region of the face.

A contrast enhanced Computed Tomography imaging was advised in our study to evaluate the extent of the lesion and investigate orbital bone erosion. Since the radiological features described the lesion to be well circumscribed, we were able to perform a simple excisional biopsy of the lesion under local anesthesia as an out-patient procedure. The best surgical approach for resection is dependent on the extent and the location of the tumor [10]. Also, functional and cosmetic considerations should be taken into account, especially those in locations like the peri orbital region. The lesion in this case was confined to the sub dermal plane and was resected en bloc. Various studies describe a successful surgical enucleation of a solitary neurofibroma with no recurrence on a long term follow up [3,6,7,9] [11-15].

Microscopic features of interlacing bundles of elongated spindle shaped cells with wavy and dark-stained nuclei, wire-like strands of collagen and immuno reactivity for S-100 protein, are pathognomonic features of a neurofibroma [14]. The standard of care for benign peripheral nerve tumors is surgical excision [13]. Incomplete resection may result in a recurrence and approximately 10% cases undergo malignant transformation [2].

4. CONCLUSION

The clinico- radiological profile of neurofibroma observed in our case is in congruence with what is commonly reported in the literature. Diagnosis of a neurofibroma is characteristically histopathological. Hence, such clinically

suspicious lesions must always be subjected to an excisional biopsy followed by the gold standard of a histopathological examination.

CONSENT

A verbal and written consent was taken from the patient for the publication of data.

ETHICAL APPROVAL

Not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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