Case Report

Syringoma – a rare tumour: Case report and review of literature

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Abstract

Syringomas are benign tumours originating from the eccrine ducts. Lower eyelid is the commonest site of origin. Very few cases have been reported in literature till date. Histopathology demonstrates normal compressed eccrine ducts in the fibrous capsule along with tumour cells arranged in tubules and solid islands. This report describes a case of syringoma on the lower eyelid in a female patient.

Keywords: Benign tumour, eccrine duct, syringoma

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Submitted: 14-Sep-2023, **Revised:** 14-Oct-2023, **Accepted:** 27-Oct-2023, **Published:** 20-Dec-2023

INTRODUCTION

Syringoma is a benign adnexal tumour of eccrine origin frequently reported in females.[1] It was first described by Kaposi and Biesiadeki in 1872 as lymphangioma tuberosum multiplex. Syringomas are known to originate from lower intraepidermal and upper dermal portion of the eccrine sweat ducts. They are predominantly observed in the adolescent age group but may manifest between first and sixth decades of life. They appear as small, solitary and firm skin coloured to yellowish papules. The commonest site of occurrence is lower eyelid and malar area of the facial region but can also occur in the chest, neck, axillae, abdomen, upper arms and external genitalia.[2] There are some variations seen between prepubertal and post-pubertal sites of occurrence. The neck and anterior trunk are the most common sites before 15 years of age followed by apocrine localizations. After 15 years of age, the apocrine localization is infrequent.[3]

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Quick Response Code:

Website: https://journals.lww.com/JPAT/

DOI: 10.4103/jomfp.jomfp_415_23

Syringomas affect around 1% population worldwide. The four types of syringomas described are local, eruptive, Down syndrome associated and familial type. The local type is most common, which is identified as symmetrical papules around the eyes. However, it is often ignored by patients due to inconspicuous skin lesions and no symptoms.[4]

This article describes review of the literature and a case report of syringoma in a 32-year-old female patient.

CASE REPORT

A 32-year-old female patient reported with the chief complaint of swelling on the lower right eyelid since six months [Figure 1]. There was no significant past medical and dental history and no positive family history was reported.

Clinical examination revealed a swelling measuring around 1 × 1 cm in dimension near the medial canthus of the lower

How to cite this article: Deshmukh RS, Deo PN, Pawar SR, Karandikar M. Syringoma – a rare tumour: Case report and review of literature. J Oral Maxillofac Pathol 2023;27:768-71.

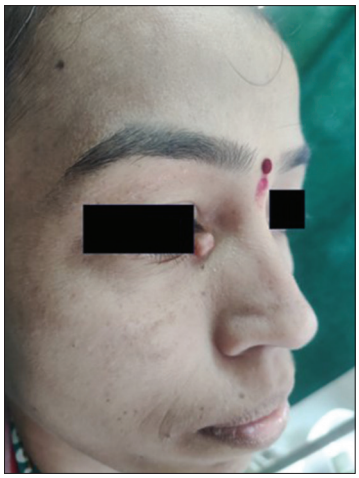

Figure 1: Swelling near the medial canthus of the lower right eyelid

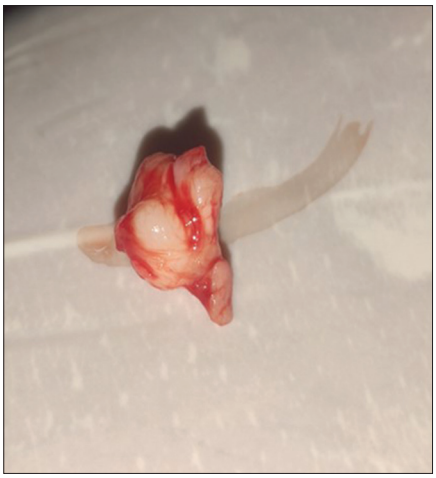

Figure 2: Gross examination – Single bit of tissue measuring approximately 0.8 × 0.8 × 0.6 cm in dimensions, whitish brown in colour with irregular surface and borders

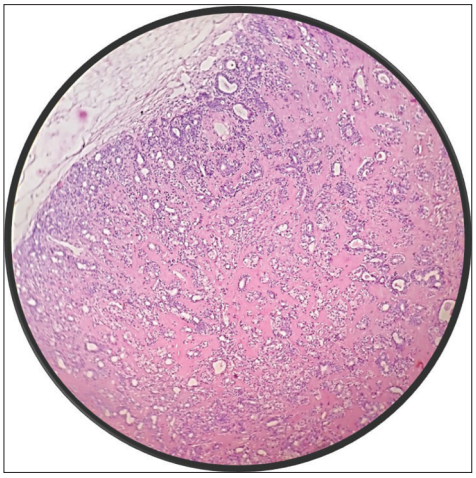

Figure 3: Low power view (10X) – Haematoxylin and eosin-stained section shows a capsulated tumour with eccrine ducts in the fibrous connective tissue

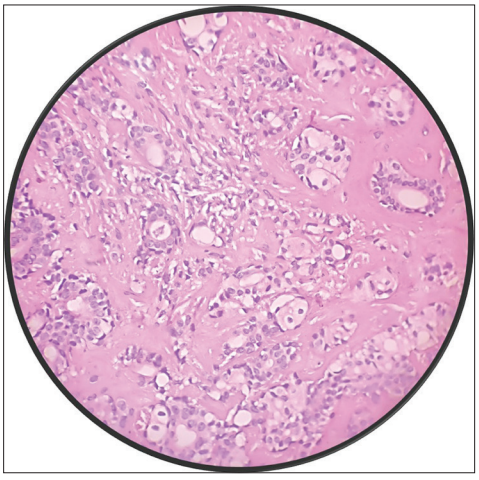

Figure 4: High power view (40X) – Haematoxylin and eosin-stained section shows the eccrine ducts. The walls of the ducts are lined by two layers of epithelial cells, a layer of cuboidal cells and a layer of myoepithelial cells

right eyelid. No complaint of dry eye or excessive formation of tears was noted. The skin over the swelling was intact and normal. There was no ulceration or redness associated with the lesion. On the basis of clinical examination, a provisional diagnosis of sebaceous cyst was made.

Gross examination revealed a single bit of tissue measuring approximately 0.8 × 0.8 × 0.6 cm in dimensions, whitish brown in colour, firm in consistency with regular borders and surfaces [Figure 2].

Microscopically, haematoxylin and eosin-stained section showed a well-circumscribed lobulated and capsulated tumour mass with tumour cells arranged in tubules and solid islands [Figure 3]. The tubules were lined by two layers of epithelium. A layer of cuboidal epithelial cells was surrounded by a layer of myoepithelial cells with vacuolated cytoplasm in most areas. The nuclei were round to oval and showed vesicular chromatin. The intervening stroma was fibro-collagenous and showed hyalinization in a few areas [Figure 4]. The peripheral fibrous capsule showed a few normal eccrine ducts. Another lobule of similar tumour separated by fibro-collagenous stroma was also seen. There was no evidence of atypia. On the basis of histopathological examination, a diagnosis of syringoma was made.

DISCUSSION

The word syringoma is derived from a Greek word syrinx, which literally means a pipe or a tube. Syringomas are benign adnexal neoplasms, which tend to undergo ductal (acrosyringeal) differentiation.[5]

Reports by some authors claimed that it was a hyperplastic response of the eccrine duct to an inflammatory reaction

rather than a true adnexal tumour and proposed the term "syringomatous dermatitis".[6]

Syringomas present as small papules, soft, skin-coloured to slightly yellowish in appearance. The papules range from 1 to 3 mm in diameter and are symmetrically distributed and asymptomatic. The lesions are most often multiple or may be solitary. They may show a localized or generalized distribution. Localized syringomas are the most common clinical variant and are typically observed in the periorbital region, mostly infraorbital. Generalized syringomas are mainly found on the neck and trunk, followed by the forearms.[7]

Female predilection could be due to the influence of oestrogen or progesterone hormones.[8]

Based on Friedman and Butler's classification scheme, many variants of syringoma are recognized: eruptive, linear, familial, vulvar, penile, scalp, acral, and plaque-type syringomas.[9]

The histogenesis of syringomas is most likely related to eccrine elements or pluripotential stem cells. However, due to histological resemblance, distinguishing eccrine versus apocrine ducts remains difficult. Thus, many tumours characterized as eccrine have actually been apocrine in differentiation. The immunohistochemical pattern of cytokeratin expression in eccrine ducts indicates differentiation in both the superficial dermal duct and the deep intraepidermal duct (e.g., sweat duct ridge). Few authors believe that eruptive syringoma represents a hyperplastic response of the eccrine duct to an inflammatory reaction rather than a true adnexal neoplasm.[10] Familial and unilateral syringoma results from mutations either within the zygote or from later post-zygotic somatic mutations, which may or may not affect gonadal mosaicism in the affected patients.[11]

The triggering factor in eruptive syringomas may be hormonal or inflammatory reaction to autoimmune condition, trauma from radiation, waxing or heat stimuli.[12]

Histopathologic examination demonstrates collections of small tadpole-shaped tubular structures, which are lined by a single or double layer of cuboidal epithelial cells in the upper dermis.[13]

Plaque-type syringomas must be histologically differentiated from desmoplastic trichoepithelioma, morpheaform basal cell carcinoma and microcystic adnexal carcinoma so as to avoid unnecessary and extensive surgical procedures.[14]

Chondroid syringoma is another variant with an incidence of less than 0.1%. The tumour is mostly solitary, benign and generally seen in males. However, malignant cases are also reported. It comprises of both epithelial and mesenchymal elements.[15,16]

Clear-cell syringoma is yet another rare variant of syringoma, which is clinically indistinguishable from common types. This variant shows strong association with diabetes mellitus.[17] It results from glycogen deposits in the tumour tissue as a result of phosphorylase deficiency, which is seen in diabetic patients.[18]

Clinically eruptive syringomas may be mistaken for acne vulgaris, sebaceous hyperplasia, lichen planus, milia, eruptive xanthoma, urticaria pigmentosa, hidrocystoma and warts.[19]

Histopathological examination is essential for diagnosis. Most tumours have a high rate of recurrence.[20]

Molecular findings suggest that syringomas may be inherited in an autosomal dominant trait with a locus on chromosome 16q22. Syringomas show positivity for carcinoembryonic antigen, epithelial membrane antigen and CK5.[21]

It has been postulated that syringomas to some extents are influenced by oestrogen or progesterone, as they are more commonly seen in women and aggravate during pregnancy and menstruation. According to Wallace and Smoller, eight out of nine cases of syringomas showed a strong nuclear and cytoplasmic positivity for progesterone receptor.[11]

The reported methods for the treatment of syringomas include surgical methods such as excision, electro dissection, cryotherapy, CO_2 laser ablation or chemical therapies such as topical or systemic retinoids.[22]

CONCLUSION

Syringomas are rare benign, adnexal tumours with a wide range of differentiation and metaplastic changes in its epithelial, myoepithelial and stromal components, the aetiology of which is not distinctive. Advanced studies like enzyme histochemistry and electron microscopy have shown differentiation of the tumour, which may help in better treatment modalities. Syringomas should be considered in the differential diagnosis of swellings of the lower eyelid in post-pubertal women.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have

given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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