

Periorbital syringomas – Excision with Castroviejo scissors. Experience in 38 patients and literature review*

*Siringomas periorbitários – Excisão com tesoura de castroviejo. Experiência em 38 pacientes e revisão da literatura**

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Abstract: BACKGROUND - Syringomas are benign adnexal tumors from intraepidermal eccrine ducts, treated by diverse surgical modalities with variable results.

OBJECTIVES - 1. To report our experience in surgical treatment of multiple periorbital syringomas excised by Castroviejo scissors followed by healing by secondary intention. 2. To present a literature review with emphasis on therapeutic aspects.

MATERIAL AND METHOD - During 68 months we treated 38 cases of periorbital syringoma. All of them were submitted to surgical excision using Castroviejo scissors, followed by healing by secondary intention. Evaluation was done 7, 30 and 90 days after surgery. The mean follow-up period was 33 months, ranging from 3 to 62 months.

RESULTS - The results were excellent in 24 patients (63.1%), good in 12 patients (31.6%) and regular in two patients (5.3%). The only immediate complication observed was edema; late complications were hypochromia (12 patients), depressed scar (one patient) and hypertrophic scar (one patient). Recurrences were not observed during the follow-up period.

CONCLUSIONS - We concluded from our results that excision of periorbital syringomas by Castroviejo scissors followed by healing by secondary intention is a low cost, straightforward outpatient procedure. Results are good, with no recurrences, although transient hypochromia may occur.

Keywords: Adenoma, sweat gland; Eccrine glands; Eyelids; Skin neoplasms; Syringoma; Treatment outcome

Resumo: FUNDAMENTO - Siringoma é tumor anexial benigno do ducto sudorífero écrino cujo tratamento é realizado por diversas modalidades cirúrgicas com resultados variáveis.

OBJETIVO - 1. Relatar a experiência no tratamento cirúrgico de siringomas periorbitários mediante excisão com a tesoura oftalmológica de Castroviejo, seguida pela cicatrização por segunda intenção. 2. Apresentar revisão da literatura com enfoque no aspecto terapêutico.

MATERIAL E MÉTODOS - Em 68 meses foram tratados 38 pacientes com siringomas periorbitários. Realizada a exérese cirúrgica com tesoura oftalmológica de Castroviejo seguida pela cicatrização por segunda intenção. Realizadas avaliações sete, 30 e 90 dias após a cirurgia. O seguimento variou entre três e 62 meses, com média de 33 meses.

RESULTADOS - Dos 38 pacientes tratados, 63,1% tiveram resultado ótimo, 31,6% bom e 5,3% regular. A única complicação imediata observada foi edema, e as tardias foram: hipocromia (12 casos), cicatriz deprimida (um caso) e hipertrófica (um caso). Não ocorreram recidivas no período observado.

CONCLUSÕES - O tratamento dos siringomas periorbitários mediante exérese cirúrgica com tesoura oftalmológica de Castroviejo, seguida pela cicatrização por segunda intenção, constitui procedimento ambulatorial de baixo custo e fácil execução. Proporciona resultados satisfatórios, sem recidivas, apesar da possibilidade de ocorrer hipocromia transitória.

Palavras-chave: Adenoma de glândula sudorípara; Glândulas écrinas; Neoplasias cutâneas; Pálpebras; Resultado de tratamento; Siringoma

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Conflict of interests: None

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INTRODUCTION

Syringoma is a benign adnexal tumor of the intra-epidermal eccrine sweat duct. It is characterized by skin-colored papules—generally multiple although sometimes isolated—ranging from one to 5mm that are asymptomatic and occur most frequently in adult women.^{1,2} It occurs predominantly in white individuals, but it has been described in black female patients.³ The most commonly affected area is the face, particularly the eyelid and periorbital regions.^{1,2} Atypical presentations on the face were reported in the form of unilateral multiple lesions forming plaques.⁴ Other locations reported are: thorax, neck, gluteal regions, pubis and vulva. On the vulva, lesions may be multiple or solitary, and cause vulvar pruritus.^{1,5} There are studies proving the presence of nuclear receptors for progesterone in the eccrine glands of syringomas of the vulva, as well as in normal glands of the deep dermis of this region.⁶

Syringoma occurs sporadically but there are familial forms with autosomal dominant heredity affecting both sexes equally and which seem to represent a form of mosaicism.^{7,8}

Histologically,⁹ syringoma is a proliferation of multiple small ducts whose walls are usually lined by two rows of flattened epithelial cells in a fibrous stroma, located in the papillary and upper reticular dermis. The lumina of the ducts contain amorphous debris. Some ducts have small comma-like tails, which gives them a tadpole-like appearance. There are also solid strings of basophilic epithelial cells that are independent of the canals. There may be cystic canalicular lumina full of keratin, lined by milium-like cells containing granules of keratohyalin. These structures can burst and produce a foreign body-type reaction. Histochemistry and electron microscopy show that syringoma is an adenoma of intra-epidermal eccrine canals. Calcium may be deposited in the ductal lumina, which is also seen in mitochondria under electron microscopy, suggesting the role of the syringeal structure in the pathogenesis of cutaneous calcinosis. An immunohistochemical study¹⁰ based on the expression of cytokeratins shows that syringoma differentiates from the transition region between the acrosyringeal region and the dermal duct of the eccrine sweat gland. Solitary tumors must be histologically differentiated from microcystic adnexal carcinoma,¹¹ in order to avoid therapeutic mismanagement.

The eruptive form generally begins suddenly in adolescence with a large number of lesions that become widespread, appear in episodes and are a therapeutic challenge.^{12,13}

Localized or eruptive syringoma were reported in association with Down syndrome.²

There is a proposal for classifying the clinical

variants of syringoma¹⁴ into four forms: localized, familial, generalized (including the eruptive form), and associated with Down syndrome. Other variants have been reported, which suggests a need to broaden this classification.²

Diagnosis of syringoma is clinical and histological. No other laboratory investigations are necessary. Clinically, a differential diagnosis must in some cases be made against the lesions of xanthelasma and milia. However it is common for syringoma, xanthelasma and milium lesions to be associated in the periorbital regions.¹⁴

The reason to treat syringoma is esthetic. In rare cases there is a need to treat the pruritus that can be present in the eruptive forms¹⁵ and vulvar forms. The indicated therapeutic modalities aim to achieve careful and effective destruction or removal of the lesions and to avoid relapse and/or unnecessary unsightly scars. The literature thus includes a range of techniques including electrocautery or electrodissection,¹⁶⁻¹⁹ surgical excision,²⁰⁻²² the use of different types of laser,²³⁻²⁸ or techniques combining the use of trichloroacetic acid and CO₂ laser.^{29,30}

MATERIAL AND METHODS

Over 68 months, 38 patients with histopathologically confirmed multiple periorbital syringomas were treated: 36 female and two male patients, with ages ranging from 19 to 72, mean age of 39.6 years. The lesions predominated in all patients in the lower periorbital region – 27 had lesions only in the lower eyelid, nine presented lesions in the upper periorbital region also, and in two patients the lesions were scattered around their faces, principally in the periorbital region and the forehead (Table 1).

The method employed was careful surgical excision followed by healing by second intention. This means that it is possible, with training and using Castroviejo ophthalmological scissors, to remove syringoma lesions individually, respecting their limits. In the technique suggested by the present authors, after a very small initial incision (a “pinch”) and raising of the skin with a fine-toothed forceps, the tumor is easily visualized since the tissue is different from normal. Carefully, in other words, without haste and after training of the hands, one can detach and remove the whole lesion, which avoids relapse, and without going in too deep, which might lead to depressed scar. It is not shaving, but actual excision, except performed extremely carefully.

It is possible to remove the syringoma without causing a sequela because it begins in the intraepidermal region in the eccrine sweat gland. However one must be attentive because there may be progression to

TABLE 1: Location of syringoma lesions on the face

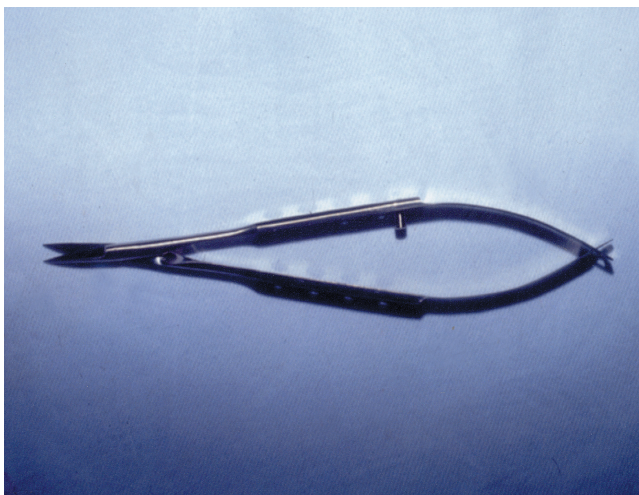
Location	Number of patients	%
Lower eyelids	27	71.1
Lower and upper eyelids	9	23.7
Scattered on the face	2	5.2
Total	38	100

the intradermal portion, which is when a slightly deeper excision is necessary; nonetheless only the tumor itself is removed, which avoids relapse and produces satisfactory esthetic results.

Thus, after cleaning the area with 0.9% saline solution, we applied infiltrative local anesthesia with 2% xylocaine and proceeded to excise the tumor using a Castroviejo ophthalmological scissors (Figure 1), with hemostasis by local compression and wound dressing with a micropore strip, removed seven days post-surgery by the physician. Depending on the number of lesions, the treatment was carried out in one or more stages, the average being two stages. All patients underwent prior testing consisting of the removal and healing of a lesion, and were observed after 30 days.

Patients were assessed seven days after surgery, when the micropore bandage was removed, and after 30 and 90 days.

Assessment of results included the patient's opinion, examination by the physician and photographic control before and after the treatment (seven, 30 and 90 days). Results were deemed: a. excellent – the resulting scar was virtually imperceptible; b. good – there was mild hypochromia, without changes in relief; and c. average – depressed scar or hypertro-

**FIGURE 1:** Castroviejo ophthalmological scissors

phic scar and/or accentuated hypochromia.

Follow-up to detect relapses ranged from three to 62 months, with an average of 33 months, after final assessment of the esthetic result.

RESULTS

In the population that was treated, white adult female patients predominated (94.7%). No familial cases were observed.

Diagnosis of syringoma was previously confirmed by histopathological examination of the lesion removed *a priori*, also as a test for the chosen method of treatment. Additionally, a sample of other lesions excised *a posteriori* underwent histologically confirmed diagnosis.

In five patients (13.1%) there were associated lesions in the same location as the syringomas, with the following histopathological diagnoses: trichoepithelioma (one case), milia (two cases) and xanthelasma (two cases).

As to the results of the treatment employed, the final overall assessment including the impressions of both physician and patient and the observation of the photographs by the investigator coincided. The results of treatment in the 38 patients (Table 2) were: excellent in 24 (63.1%) (Figures 2 and 3, A and B); good in 12 (31.6%) (Figure 4) and average in two (5.3%) (Figure 5). The only immediate complication observed was edema; late complications were: hypochromia (12 cases), depressed scar (one case) and hypertrophic scar (one case). Residual hypochromia did not fully regress but did significantly improve over an average period of 31 months, without treatment; depressed scar was treated surgically by excision and suture with 6.0 thread, and hypertrophic scar was treated by intralesional filtration with corticosteroids, both resulting in improved esthetic appearance. All patients declared themselves very satisfied at the end of the observation period and there were no relapses.

TABLE 2: Esthetic result of treatment, after 90 days, in 38 patients with multiple periorbital syringomas

Result	Number of patients	%
Excellent	24	63.1
Good	12	31.6
Average	2	5.3
Total	38	100



FIGURE 2: Female patient with syringomas in the lower periorbital regions before (A) and after (B) treatment, with excellent result

DISCUSSION

Syringoma can be a major esthetic problem since the lesions generally occur on the faces of adult female patients and are multiple.¹

In the present study there was a predominance of multiple periorbital syringomas in adult white women (36 cases or 94.7%), which is in line with the literature.² Solitary lesions or cases of eruptive syringoma were excluded from this observation. Familial cases are described,^{7,8} but did not occur in our sample.

All five patients (13.1%) with other associated lesions were treated by the same technique. The occurrence of milia and xanthelasma in association with



FIGURE 4: Female patient with syringomas in the upper eyelid after treatment, with good result (hypochromia)

syringoma lesions is reported relatively frequently.

Prior excision of one lesion was carried out for assessment after 30 days of the esthetic results of the method employed. A diagnosis of syringoma was confirmed by histopathological examination of this lesion. A sample of the other lesions excised afterwards were also diagnosed by the characteristic histological findings.⁹

There are actually few studies analyzing in detail the aspects mentioned above, in other words, that report the epidemiological and clinical features of the disease, the therapeutic outcome and the follow-up in a larger number of cases. It is undeniably difficult to follow up the therapeutic outcome when the lesions are benign and the chosen treatment modality reduces the possibility of relapses.

Underscoring the treatment of multiple perior-



FIGURE 3: Female patient with syringomas in the lower periorbital regions before (A) and after (B) treatment, with excellent result



FIGURE 5: Female patient with syringomas in the lower periorbital regions after treatment, with average result (hypertrophic scar)

point,¹⁶⁻³⁰ the results of the observation are similar to those reported in the literature, whatever the chosen technique. The destruction of the lesions by electrosurgery may produce scars and/or relapses with greater frequency, given the tumor's histological features, above all the location of its differentiation.^{9,10} Some authors advocate electrosurgery, whether electrocautery or intralesional electrodissection, with a fine-tipped electrode or an epilation needle introduced to the level of the reticular dermis, without curettage.¹⁶⁻¹⁹ They deem it a safe, effective and affordable treatment, but one which requires experience and attention, above all in the treatment of lesions on the eyelids, so as to avoid sequelae. They stress the risk of this simple method being abandoned in favor of laser, which is more expensive since there are no clinical studies comparing these two techniques. The authors' experience with different modalities of electrosurgery in very few patients (unpublished data) has not been satisfactory, since despite taking all necessary care, permanent depressed hypochromic scars occurred as well as relapses in some cases.

Conventional surgical removal with suture and blepharoplasty are effective methods that give excellent results, particularly when the lesions are clustered and in a linear arrangement, provided a skilled professional performs them.²⁰⁻²²

A recent study²¹ of surgical excision followed by healing by second intention in xanthelasma lesions in 28 patients who were followed over 18 months presented similar conclusions to the present study in regard to effectiveness and satisfactory esthetic results.

Healing by secondary intention²⁰ provides esthetic outcomes similar to or better than those given by approximation of the surgical boundaries by suture. It is a simple, straightforward, affordable method and therefore useful in carrying out procedures in an outpatient setting, specially for a large number of lesions. The results of this observation agree with those of other authors^{21,22} in regard to the effectiveness of the method, in the treatment of xanthelasma. In the present experiment, among the 38 patients who had lesions excised and allowed to heal by secondary intention, results were deemed excellent in 24 (63.1%), good in 12 (31.6%) and average in only two cases (5.3%).

One should point out the possibility of transitory hypochromia occurring as a sequela, which did in fact occur in 12 (31.6%) patients and regressed spontaneously after an average of three years' follow-up.

Recent literature on the treatment of multiple

syringoma most often addresses the use of different types of laser to destroy the lesions.²³⁻³⁰ The first reports referred to continuous CO₂ laser,²³ with disastrous results owing to the increased risk of scarring. The most frequently mentioned type is pulsed CO₂ laser, which reduces the risk of scarring and gives excellent esthetic results, and may even be performed without anesthesia or with only topical anesthesia.^{25-27,29,30} The use of a 1-mm-handpiece provides additional advantage, further reducing the risk of scarring.²⁷ The literature also refers to the use of erbium laser²⁴ and alexandrite laser which requires prior tattooing.²⁸ Treatments combining application of 50% trichloroacetic acid before or after the use of pulsed CO₂ laser^{29,30} are reported as advantageous. The combined technique enables the number of passes of the laser to be reduced, thereby minimizing the risk of thermal damage both at the lesion site and to the surrounding skin, as well as removing deeper syringoma cells, thus avoiding scarring and relapse.

Using laser is advantageous but the cost is high, the method is virtually unavailable in public institutions, and requires well-trained professionals working in suitable settings and following correct protection measures, since the lesions predominate in the periocular region, which greatly restricts the use of the laser.

This report of the present experiment, involving simple and careful surgical excision using a Castroviejo ophthalmological scissors followed by healing by secondary intention, is thus considered an important contribution to the solution of the problem of multiple periorbital syringomas, particularly in public hospitals.

CONCLUSIONS

The treatment of periorbital syringomas by careful surgical exeresis with the Castroviejo ophthalmological scissors is an outpatient department procedure that may require one or more stages depending on the number of lesions. Healing by secondary intention facilitates and expedites the treatment of multiple lesions. This straightforward, safe, effective and accessible method gives satisfactory outcomes ranging from good to excellent, without relapse, despite the possibility of the occurrence of transitory hypochromia. We highlight the need for clinical and histopathological diagnosis, in addition to prior assessment of the esthetic outcome by test, in other words, by the removal and healing of at least one lesion in all patients, particularly in dark-skinned people or those with periorbital hyperchromia. □

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