Exogenous ochronosis: a case report

Ocronose exógena: relato de caso

ABSTRACT

Introduction: Exogenous ochronosis is characterized by asymptomatic black-bluish hyperpigmentation, typically located in the face, neck, dorsum and the extensor surfaces of the extremities. It most frequently results from using hydroquinone.

Objective: To highlight the use of several therapeutic modalities in order to obtain satisfactory treatment results.

Method: The treatment began with the application of Nd:YAG laser. Since there was no response, ultrapulse CO2 laser was indicated. Still without satisfactory results, a combination of intense pulsed light, microdermabrasion and chemical peels was carried out.

Results: The total resolution of the lesions was verified.

Conclusion: Exogenous ochronosis is difficult to treat, sometimes requiring the combination of several treatment methods.

Keywords: ochronosis; chemical process; treatment outcome.

INTRODUCTION

Ochronosis, a brownish-gray pigmentation of connective tissues, can be classified as endogenous or exogenous. The endogenous form (also known as alcaptonuria) is a rare congenital condition of recessive autosomal inheritance, resulting from the absence of oxidase – the enzyme that converts homogentisic acid in acetoacetic and fumaric acids – in homogentisic acid. Affected individuals develop an accumulation of homogentisic acid, an insoluble pigment that is deposited in several tissues such as cartilages, cardiac valves and the skin.

Exogenous ochronosis is clinically and histologically similar to the endogenous type, however it is not a systemic condition. It is characterized by asymptomatic bluish-black or grayish hyperpigmentation, typically located in the face, neck, dorsum and extremities’ extending surfaces. It is most commonly caused by the use of hydroquinone, however resorcinol, phenol, mercury, picric acid and oral antimalarials can also contribute to...
the condition. The highest incidence occurs in blacks from South Africa, where the prevalence is estimated at between 28 and 35% of the population. Exogenous ochronosis was previously believed to be caused only by the use of high concentrations of hydroquinone for prolonged periods. However, recent reports indicate that using 2% hydroquinone for less than 3 months can trigger the condition.

The hyperpigmentation mechanism induced by hydroquinone remains uncertain. High concentrations of hydroquinone may activate tyrosinase and thus stimulate the synthesis of melanin. Many cases are linked to sun exposure, with reports of ochronosis not spreading into areas of vitiligo, which suggests that melanocytes might be involved. Other authors suggest that hydroquinone inhibits the activity of the homogentisic acid’s oxidase enzyme in the skin, resulting in the accumulation of that acid.

Three phases can be identified in exogenous ochronoses. Phase I involves only erythema and minor pigmentation of the face and neck. Hyperpigmentation, “caviar-like” papules, and atrophy appear progressively in phase II. Phase III includes papular nodular lesions that are sometimes surrounded by inflammation.

Exogenous ochronosis is quite resistant to treatment. There are reports of several therapeutic modalities, most of which have produced discouraging results. In this study we describe a case of exogenous ochronosis treated with a combination of procedures that completely treated the lesions.

CASE REPORT

A 40-year-old female patient presented with dark macules on the face. She reported continuous use of hydroquinone for 8 years. A physical examination showed hyperchromia of the malar region, with the presence of dark micropapules. The histopathology of the lesion revealed banana-shaped, brownish-yellow ochronotic filaments in the papillary dermis, compatible with a diagnosis of exogenous ochronosis (Figure 1). The patient tested negative for endogenous ochronosis.

The treatment began with four sessions of the Nd:YAG 1064 nm laser (Q-YAG 5® Palomar Medical Technologies, Inc., Burlington, MA, US), using the 4 mm spot and a fluence ranging between 2.9 and 3.05 J/cm². As a satisfactory clinical response was not achieved, the ultrapulse CO₂ laser (Luxar 20 LUMENIS Ltda., Yokeam, Israel) was applied with a fluence of 5 watts, in six sessions with 30 days intervals. Although the lesions improved after this treatment, the right malar region did not improve as much as the left side. Intense pulsed light (IPL) (G handpiece, Starlux® platform, Palomar Medical Technologies, Inc. Burlington, MA, USA) (36 J – 10 ms) was applied in the left malar region to further lighten the area. Since the patient still presented light hyperchromia in the malar regions after those procedures, microdermabrasion and chemical peeling (5% hydroquinone + 5% retinoic acid + 14% salicylic acid) were carried out. Afterwards, the patient still presented hyperchromia and some darkened papules in the malar region. After one month, 3 sessions of IPL (G handpiece, Starlux®) (36 J – 10 ms) were carried out, with intervals of 30 days, immediately followed by 20% trichloroacetic acid (TCA) peels; the lesions were then completely resolved. (Figure 2)

DISCUSSION

Exogenous ochronosis is difficult to treat. Several treatments have been already used, usually with frustrating results. Avoiding the use of a substance that causes discoloration (e.g., hydroquinone) is beneficial, however noticeable improvement may take several years. Retinoic acid was effective in some patients, yet caused temporary hyperpigmentation in others. The results of treatments with sunscreens and low potency corticosteroids were variable. A case of sarcoid-like ochronosis was treated with tetracycline, with improvement observed after 1 month and total whitening after 3 months.

One case described in the literature reports the removal of hyperpigmentation in a patient with fair skin using dermabra-
Exogenous ochronosis

Diven and others used a combination of dermabrasion and CO2 laser with satisfactory results in the periorbital and nasal regions in a female patient with a high phototype. In the case described in this study, the ochronosis lesion improved using this combination, yet was not resolved completely.

The use of Q-switched lasers (QS) for the treatment of tattoos and pigmented lesions is well documented in the literature. The efficacy of tattoo removal results from the use of high energy and ultrashort pulses, with the formation of microparticles that are removed by lymphatic drainage or through transepidermal elimination. Based on the recently discovered fact that the exogenous ochronosis' pigment accumulates in the dermis in a similar way as tattoo pigment, the QS Nd:YAG laser showed only modest success in treating the lesions after four sessions, demonstrating the variability in response of exogenous ochronosis to different types of lasers.

IPL has also been reported to be effective in the treatment of pigmented lesions. The advantages of IPL include the ability to adjust the width of the pulse and the wavelength according to skin type and depth of the pigment deposits. The use of IPL contributed to the whitening of ochronosis lesions in that patient. No reports on the use of IPL in the treatment of ochronosis were found in the literature.

TCA peels in different concentrations have been used for several years in the treatment of photoaging, acne scars and pigmentation disorders. The use of TCA in hyperpigmentation is related to the coagulative necrosis of the epidermal cells' proteins, followed by cellular death. There are reports in the literature that this acid is ineffective in the treatment of ochronosis. Nevertheless, we used TCA peels as an adjuvant therapy, applied immediately after IPL sessions, and were able to observe that this method most effectively caused the regression of the lesions.

CONCLUSION

Exogenous ochronosis is a pathology that is difficult to treat; the literature reports several ineffective therapies. In the case presented, the association of several methods (QS Nd:YAG laser, CO2 laser, microdermabrasion, IPL and 20% TCA peel) was necessary to obtain a satisfactory result.

REFERÊNCIAS
