

Do you know this syndrome?

Você conhece esta síndrome?

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CASE REPORT

A 52-year old female patient was referred to the dermatology clinic complaining of unsightly lesions on her chest that had been present for the past 15 years. She described the sudden onset of asymptomatic erythematous papules, confined to the left side of her chest. Over the following years, the patient noticed an increase in the number of lesions, which spread to her left arm, now developing a mild burning sensation when exposed to low temperatures. The patient had had a hysterectomy 15 years previously due to uterine

fibroids. On dermatological examination, firm, reddish-brown nodules-papules measuring 0.3 cm in diameter, presenting singly or coalescing into plaques, were found on the left side of her chest (Figures 1 and 2). A single papule with the same characteristics was found on her left arm. Biopsy revealed proliferation of bundles of muscle cells with spindle-shaped nuclei in the papillary and reticular dermis (Figure 3). Tomography of the abdomen and pelvis was normal.



FIGURE 1: Firm, reddish-brown nodules-papules measuring 0.3 cm in diameter, presenting singly or coalescing into plaques, on the left side of the chest



FIGURE 2: The same nodule-papules showed at greater magnification

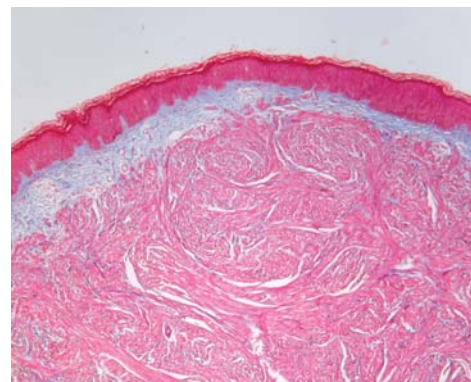


FIGURE 3: Well-circumscribed, increased smooth muscle fibers with spindle-shaped nuclei, located in the papillary and reticular dermis and interspersed with collagen, which is stained in blue and mostly seen in the papillary dermis. Masson's trichrome stain, 100X magnification

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DISCUSSION

Leiomyoma, a benign tumor originating in the smooth muscle tissue, is classified into three types according to its site of origin: piloleiomyoma in which the arrector pili muscle is affected; genital and nipple leiomyoma, which develop in these regions, and vascular leiomyoma in which the smooth muscle of the middle layer of the vessels is affected.^{1,3,4} Piloleiomyoma is the most common of these, affecting both sexes equally and affecting patients of any age, although it is more common between 10 and 30 years of age¹⁻⁶. It presents as a single lesion, more common in males, or as multiple lesions, which are more common in women. It is characterized by reddish-brown papules-nodules of less than 1.5 cm in diameter with a smooth surface and firm consistency, located asymmetrically on the extensor muscles of the limbs, chest and face. The lesions are tender or painful, particularly when presentation is multiple, with patients referring to a burning sensation, tightness or pain that may appear spontaneously or induced by cold temperatures, touch, pressure or excitement. The pain is known to be caused by compression of the cutaneous nerves, by injury or by simple contraction of the muscle fibers of the tumor.^{1,3,4,6} The occurrence of multiple cases of lei-

omyomas in families has been described as autosomal dominant inheritance with incomplete penetrance. The finding of cutaneous leiomyomas with concomitant erythrocytosis has been reported and is explained by an effect of the "erythropoietin-like" behavior of the tumor.¹ The association between piloleiomyoma and uterine leiomyomas, referred to as Reed syndrome or *familial leiomyomatosis cutis et uteri*, is considered a rare presentation that may be associated with renal cell carcinoma.^{1,2,3,5} In this syndrome, the defect occurs in chromosome 1q.42.3-43, which acts as a tumor suppressor.² In these cases, investigation by imaging tests is mandatory.^{1,2} As for treatment, surgical excision is recommended when lesions are few; however, there is a recurrence of the lesions in 50% of cases. In cases of numerous and painful piloleiomyomas, a pharmacological approach is recommended; however, effectiveness is limited. Drugs used for this treatment include gabapentin, oral or topical nitroglycerin, lidocaine, nifedipine, verapamil, phenoxybenzamine, phentolamine, hyoscine, painkillers and antidepressants. Cryotherapy and electrocoagulation have been used, but with little success.¹⁻⁶ In this case, a decision was made to monitor the patient clinically, since her symptoms were mild. □

Abstract: Piloleiomyoma, a benign smooth-muscle tumor arising from the arrectores pilorum muscles of the skin, affects males and females in the third decade of life. It presents as asymmetrical, reddish-brown nodules or papules with a firm consistency, predominantly located on the limbs. When multiple lesions are present, they may be tender or painful. Their association with uterine fibroids, referred to as Reed syndrome or *familial leiomyomatosis cutis et uteri*, is rare and may be associated with renal cell carcinoma. The approach consists of surgical excision in cases presenting few lesions and pharmacological treatment if symptomatic. The present paper describes a case of Reed syndrome in which a decision was made to monitor the patient in view of the absence of symptoms.

Keywords: Collagen; Cryotherapy; Leiomyoma

Resumo: Tumor benigno de tecido muscular, o piloleiomioma tem origem no músculo eretor do pelo, atingindo ambos os sexos geralmente na terceira década de vida. Apresenta-se como nódulo-pápulas assimétricas nas extremidades, de cor eritemato-acastanhada e de consistência firme. As lesões, quando múltiplas, podem ser sensíveis ou dolorosas. Sua associação com miomas uterinos, denominada de síndrome de Reed ou leiomiomatose cutis et uteri, é apresentação rara, podendo estar associada a carcinoma de células renais. A abordagem é cirúrgica em casos isolados e medicamentosa se houver sintomas. Relatamos um caso de síndrome de Reed em que se optou por acompanhamento pela ausência de sintomatologia.

Palavras-chave: Colágeno; Crioterapia; Leiomioma

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