

Dermoscopic aspects of syringocystadenoma papilliferum associated with nevus sebaceus*

Aspectos dermatoscópicos do siringocistoadenoma papilífero associado a nevo sebáceo

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Abstract: Syringocystadenoma papilliferum is a rare benign adnexal tumor that frequently shows apocrine differentiation. It usually develops on the scalp and is associated with a nevus sebaceus in 40% of cases. Although the clinical presentation may differ, its histology is characteristic. Reports have been made of dermoscopy used in cases of adnexal tumors such as eccrine poromas, hidradenomas and angiohistiocytomas; however, up to the present moment there have been no reports of dermoscopy in a case of syringocystadenoma. This paper describes the dermoscopic features found in a case of syringocystadenoma associated with a nevus sebaceus, revealing a polymorphous vascular pattern including a horseshoe-shaped arrangement of vessels.

Keywords: Dermoscopy; Diagnosis; Evaluation; Neoplasms, adnexal and skin appendage; Skin neoplasms

Resumo: O siringocistoadenoma papilífero é uma neoplasia anexial benigna rara, com frequente diferenciação apócrina. Localiza-se preferencialmente no couro cabeludo e está associado ao nevo sebáceo em 40% dos casos. Apesar da variabilidade clínica, a histologia é característica. Há relatos da dermatoscopia de tumores anexiais, como poroma écrino, hidradenoma e angio-histiocitoma; porém, até o momento, não há descrição da dermatoscopia do siringocistoadenoma. Apresentamos aspectos dermatoscópicos de um caso de siringocistoadenoma associado a nevo sebáceo, visualizando-se padrão vascular polimorfo e vasos em ferradura.

Palavras-chave: Avaliação; Dermoscopia; Diagnóstico; Neoplasias de anexos e de apêndices cutâneos; Neoplasias cutâneas

INTRODUCTION

Syringocystadenoma papilliferum is a rare, benign adnexal tumor that in 50% of cases is present at birth, while in another 15-30% of cases it develops during puberty.^{1,2} It is often located on the scalp and face.^{3,4} It originates from undifferentiated cells and has the potential for apocrine or eccrine differentiation, although presence of the apocrine component is more common.⁴⁻⁶ In the majority of cases, syringocystadenoma papilliferum is associated with other

benign tumors, nevus sebaceus being present in 40% of cases.^{3,6} Syringocystadenoma papilliferum and trichoblastoma are the most common tumors developing in nevus sebaceus; however, others are also found: nodular hidradenoma, syringoma, sebaceous epithelioma, chondroid syringoma, trichilemmoma, trichoadenoma, sebaceous carcinoma, basal cell carcinoma, trichilemmal cyst and basaloid epithelial proliferation, which may be confused with basal cell carcinoma.

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noma.⁵

Clinically, syringocystadenoma papilliferum presents with a range of nonspecific lesions. Diagnosis may be confirmed by histopathology, a reliable tool for this purpose since histology of this condition is characteristic.⁵ Dermoscopy or surface microscopy is a method used to visualize structures situated under the stratum corneum.⁷ Its principal indication is the diagnosis and follow-up of pigmented skin lesions, with the objective of detecting melanoma at early stages. However, its use has been extended to the study of non-melanocytic lesions, pathologies of the scalp and nails, and evaluation of the vascular pattern of skin tumors.⁷ Reports have been published on the dermoscopic pattern of adnexal tumors such as eccrine poromas, hidradenomas and angiohistiocytomas.⁸⁻¹⁰ Nevertheless, up to the present moment, there has been no description of dermoscopy carried out to evaluate syringocystadenoma papilliferum. The present report describes the clinical investigation, dermoscopy and histopathology of a case of syringocystadenoma papilliferum associated with a nevus sebaceus and syringoma in a 50-year old woman.

CASE REPORT

A healthy 50-year old female civil servant with skin photo type III presented with an asymptomatic lesion on her scalp that had been present since birth. Fifteen years prior to consultation she had noticed a change in the lesion, with occasional episodes of bleeding resulting from trauma incurred while brushing her hair. At clinical examination, an alopecic papular, yellowish-orange plaque was found in the left parietal region of the scalp. Around this plaque an erythematous, lobular, exophytic lesion with a moist appearance



FIGURE 2: Papular, yellowish-orange plaque and a lobular, exophytic lesion with a moist appearance

was found (Figures 1 and 2).

Dermoscopy of the papular, yellowish-orange plaque showed round-shaped or oval structures, presenting either singly or in clusters, varying in size and of a yellowish-white color (Figure 3). Dermoscopy of the exophytic lesion showed an erythematous background apparently divided by whitish linear structures that demarcated lobules of different sizes and contained vascular structures of different forms: irregularly linear vessels, glomerular vessels and some vessels arranged in a horseshoe-shaped pattern (Figure 4). The entire lesion was resected.

Histopathology showed a hamartomatous lesion characterized by acanthosis, papillomatosis and various superficial, mature sebaceous glands compatible with a diagnosis of nevus sebaceus (Figure 5). In



FIGURE 1: Papular, yellowish-orange plaque and a lobular, exophytic lesion with a moist appearance



FIGURE 3: Dermoscopy of the sebaceous nevus showing round-shaped or oval, yellowish-white structures presenting either singly or in clusters

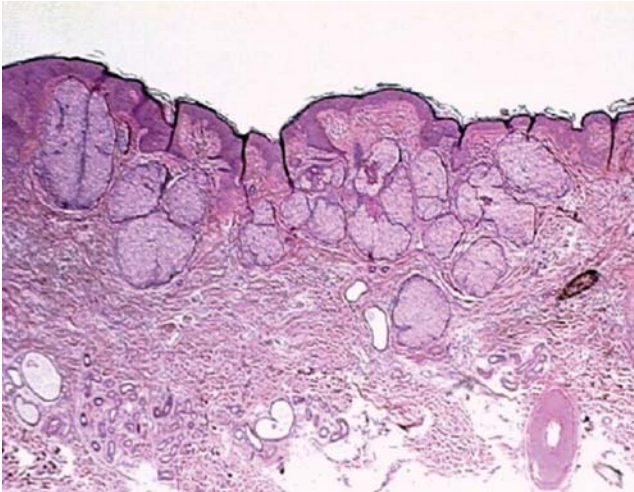


FIGURE 4: Dermoscopy of the syringocystadenoma papilliferum showing a whitish-pink background. Irregularly linear and tortuous vessels, glomerular vessels and some vessels arranged in a horseshoe-shaped pattern. Detailed image showing vessels arranged in a horseshoe-shaped pattern

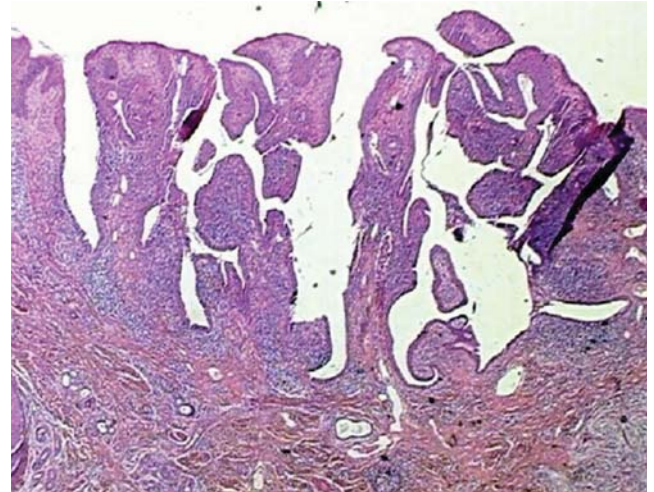


FIGURE 6: Central area of the epidermis showing pronounced papillomatosis associated with invaginations and papillary projections. Presence of sparse plasmacytic infiltrate in the tumor stroma. (HE, 20x magnification)

addition, papillomatosis was more pronounced in a central area of the epidermis and was associated with invaginations and papillary projections, the lining of which consisted of a double layer of cuboidal cells. The luminal layer consisted of cells with an oval nucleus, eosinophilic cytoplasm, the presence of decapitation secretion and evidence of cell debris in the lumen. The external layer consists of cuboidal cells with clear, sparse cytoplasm. In the tumor stroma, the presence of sparse plasmacytic infiltrate. These alterations are characteristic of syringocystadenoma papilliferum (Figures 6 and 7). Finally, tubular structures consisting of two lines of clear cells with amorphous debris compatible with syringoma were

found in the lumen. The final diagnosis consisted of nevus sebaceus, syringocystadenoma papilliferum and syringoma occurring in the same lesion.

DISCUSSION

The use of dermoscopy to aid diagnosis in cases of cutaneous non-melanocytic tumors is relatively new compared to its application in malignant melanomas of the skin.¹⁰ There are few cases published in the literature on dermoscopy of adnexal tumors and these refer to eccrine poromas, hidradenomas and angiohistiocytomas.⁸⁻¹⁰ Therefore, up to the present time little is known on dermoscopy in cases of syringocystadenoma papilliferum and nevus sebaceus.



FIGURE 5: Superficial, mature sebaceous glands. (HE, 20x magnification)

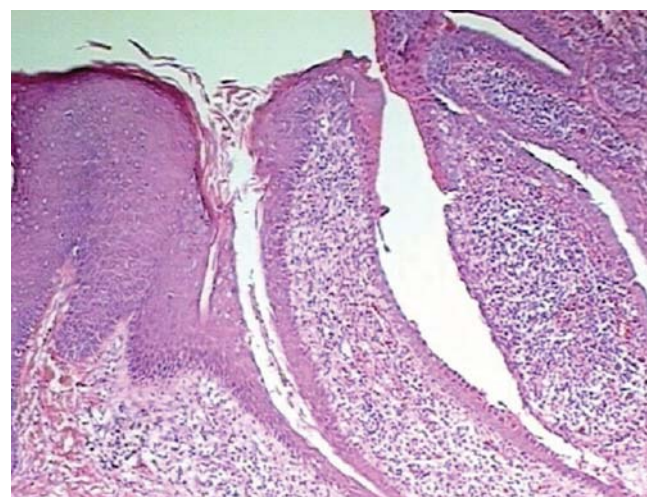


FIGURE 7: Detailed image of the papillary projections showing a double-layer cuboidal cell lining. (HE, 40x magnification)

Dermoscopy of a nevus sebaceus shows round-shaped structures of a yellowish-white color, grouped together or presenting singly, which may correspond to clusters of mature, superficial sebaceous glands. Dermoscopy of a syringocystadenoma highlights a polymorphous vascular pattern on a pinkish-white background. This polymorphous vascular pattern shows irregularly linear and glomerular vessels, some surrounded by a whitish halo and others grouped in a horseshoe arrangement.

Dermoscopy is a noninvasive tool that aids diagnosis and monitors the progression of dermatological lesions, principally melanomas.⁷ New concepts in dermatology based on dermoscopy are discovered and defined daily. The actual value of this technique in the diagnosis of adnexal tumors, which tend to be uncharacteristic at clinical examination, is not yet known; therefore, in the future it is hoped that dermoscopy may also contribute towards identification of this group of lesions. □

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