Syringocystadenoma papilliferum

Authors: Doctor Alexander C. Katoulis¹, Doctor Evangelia Bozi
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¹Department of Dermatology and Venereology, A. Sygros Hospital, National and Kapodistrian University of Athens, Athens, Greece.

Abstract

Syringocystadenoma papilliferum (SP) is a skin hamartoma, which is believed to be derived from apocrine sweat glands or, less often, from eccrine sweat glands. It is an exceedingly rare neoplasm, appearing at birth or arising during infancy, around the time of puberty, or later in life. Three clinical types have been described. The plaque type presents mostly as a hairless area in the scalp, often in association with a sebaceous nevus of Jadassohn; it may also appear de novo. The linear type, which is usually seen on the neck or face and the Solitary nodular type, which shows predilection for the trunk. Association with hamartomas of follicular or sebaceous gland origin is common. In about one-third of the cases, SP is associated with a nevus sebaceous. Basal cell carcinoma development has been reported in up to 10% of the cases. Squamous cell, verrucous or ductal carcinoma may also develop, but much less frequently. The diagnosis is clinically suspected and histologically confirmed. Due to the risk of malignant degeneration, prophylactic surgical excision is advisable, wherever possible. Complete surgical excision followed by detailed histological examination is the treatment of choice.

Keywords

skin hamartoma, apocrine sweat gland, sebaceous nevus, basal cell carcinoma

Synonyms

Syringadenoma papilliferum, naevus syringocystadenomatous papilliferus

Definition and Histogenesis

Syringocystadenoma papilliferum (SP) is a hamartomatous malformation of the skin, which is believed to derive from apocrine sweat glands. However, a few may derive from eccrine sweat glands. In the past, it was thought to represent an apocrine gland adenoma. Nevertheless, the histogenesis remains controversial (Yamamoto et al, 2002). According to the classical concept, SP is a hamartoma arising from pluripotent cells.

Epidemiology

SP is an exceedingly rare neoplasm. Almost half are present at birth or appear during infancy (plaque and linear variety). It commonly arises around the time of puberty, or less commonly, during adolescence or adult life (solitary nodule variety) (Pinkus, 1954).

Clinical description

Three clinical types have been described. It may occur as a linear group of nodules, a nodular plaque or as a solitary nodule (Pinkus, 1954).

The plaque type most often presents as a hairless area in the scalp. At puberty it may become larger, nodular, verrucous or crusted, resembling nevus sebaceous. Such a lesion
often develops in association with a sebaceous nevus of Jadassohn or may appear de novo without preexisting lesion (Koga, 1999).

The linear type is usually seen on the neck or face. Lesions consist of multiple pink to red to brown, firm papules or nodules, 1-10 mm of diameter, that occur in groups. The papules may be umbilicated, mimicking molluscum contagiosum. Pinpoint vesicle-like inclusions filled with clear fluid are usually seen.

The solitary nodular form shows predilection for the trunk, especially for the shoulders, axillae and the genital area (Patterson, 2001). The solitary nodule is up to 1 cm in diameter, it is domed, umbilicated or pedunculated, often with a friable or crusted surface. Several such nodules may be scattered in the same area. Occasionally, extensive verrucous or papillary plaques are present. Association with hamartomas of follicular or sebaceous gland origin is common (Pinkus, 1954). In about one-third of the cases, SP is associated with a nevus sebaceous. Multiple tumours of adnexal origin (such as trichoblastoma, hidradenoma papilliferum, poroma folliculare, trichilemmoma etc.) have been reported to arise on a sebaceous nevus, among which a SP may be included (Stavrianeas et al, 1997). Basal cell carcinoma (BCC) development has been reported in up to 10% of the cases (Helwig et al, 1955). In the majority of such cases, there is a coexistent nevus sebaceous. It has been suggested that allelic loss at 9q22 is consistent with the clinical observation of transition of SP to BCC (Boni et al, 2001). Squamous cell carcinoma (SCC) may also be developed, but much less frequently. Since now, only two cases of verrucous carcinoma in conjunction with SP have been published (Monticciolo et al, 2002). Ductal carcinoma arising from SP has been reported as well (Hugel et al, 2003). Ulceration or rapid enlargement is indicative of malignant transformation. Association with extra-cutaneous abnormalities may be seen in the contact of epidermal nevus syndrome. Syringocystadenocarcinoma papilliferum is the malignant counterpart of SP (Ishida-Yamamoto et al, 2001).

Diagnostic Methods
Histology
The epidermis shows papillomatosis. One or several cystic invaginations extend downward from the epidermis. Cystic changes of the sweat glands can be observed, i.e. dilated glands and ducts with numerous villus papillary projections extending into the lumen of cystic invaginations. Both the invaginations and the papillary projections are lined by a glandular epithelium consisting of two rows of cells: an outer layer of small cuboidal cells with strongly basophilic nuclei; and an inner layer of high columnar cells demonstrating “decapitation” secretion (characteristic for the secretory cells of the apocrine sweat glands). Apocrine glands are prominent deep in the dermis below the invaginations to which they are connected. An inflammatory infiltrate composed almost entirely of plasmacytes is present, especially in the stroma within the papillary projections (Niizuma K, 1976).

Ultrastructurally, the constituent epithelial cells are divided into three types: luminal tumour cells bearing features of the secretory or ductal luminal cells of sweat glands, but being somewhat immature in appearance; basal tumour cells that were basaloid in nature; and undifferentiated clear cells suggesting stem or progenitor cell properties (Yamamoto et al, 2002).

Immunohistochemistry
The tumor stains positively for carcinoembryonic antigen. Histochemical studies of individual cases showed evidence (large amount of glycogen, absence of large secretory granules, presence of amylaphosphorylase) of eccrine rather than apocrine secretory cells presence.

Diagnosis and Differential diagnosis
The diagnosis is clinically suspected and histologically confirmed. Syringoma (non grouped lesions) and basal cell carcinoma (especially around the eye) must be excluded. The plaque and linear forms on the head and neck may be difficult to distinguish from the sebaceous nevus to which they may be related. Umbilicated nodules may be confused with Molluscum contagiosum.

Treatment
Due to the risk of malignant degeneration, prophylactic surgical excision is advisable, wherever possible. Complete surgical excision followed by detailed histological examination is the treatment of choice. Radiotherapy is ineffective. Destructive methods are wise to be avoided.

References
Helwig EB, Hackney VC. Syringadenoma papilliferum: lesions with and without naevus

http://www.orpha.net/data/patho/GB/uk-Syringocystadenoma-papilliferum.pdf


