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Syringocystadenoma Papilliferum Vulve – Tumor with Very Rare Localization on the Vulva

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Abstract

Syringocystadenoma papilliferum is a benign cutaneous adnexal tumor of eccrine and apocrine glands, with a warty appearance that is usually found on the scalp, neck and face, much less frequently appears in the chest or abdomen and extremely rarely on the female genital organs, i.e. the vulva. We present a case of Syringocystadenoma papilliferum on the vulva of a 64-year-old woman. This case illustrates the atypical location of this rare disease and adds to the differential diagnosis of lesions on the vulva.

Keywords: Syringocystadenoma Papilliferum, Vulva, Wart, Adnexal Skin Tumor, Nevus Sebaceous, Tumor Excision

Background

Syringocystadenoma papilliferum (SCAP) is a rare benign adnexal neoplasm of unclear pathogenesis, originating from apocrine and eccrine glands [1, 2, 3]. It occurs significantly more often in children and adolescents than in adults. The most common localization is the head and neck (up to 75%), trunk (20–53.5%), on the extremities (5.0–33.8%), with only 12.5% on the genitals [4, 5, 6, 7, 8, 9]. The size of this neoplasm varies from 5 to 160 mm [10], and it appears in the form of single or multiple, papillary, cystic-nodular lesions that can be brown or erythematous in color [9, 10]. This paper presents a case of SCAP, and due to its unusual localization, only a few cases of syringocystadenoma papilliferum vulvae have been recorded in the literature.

Personal Content

A 64-year-old female presented with two tumorous changes on the lower third of the left labia majoris, measuring 10×6 mm and 5×5 mm. She noticed the described changes six months ago, which increased over the past three months, and there was noticeable redness accompanied by itching in the area of the described changes. In the anamnestic data, the patient states that she had cancer of the larynx, and that the right lobe of the thyroid gland was removed. He states that he has hypertension, diabetes mellitus type 2 and mixed anxiety and depression disorder, and insomnia non-organica. Use therapy for all previous illnesses.

Problem

A colposcopy examination revealed two swellings in a hyperemic zone without enlargement of the regional lymph nodes. The changes were clearly limited from the rest of the tissue with increased abnormal shape and branching of the capillary drawings (green optical light filter was used during the examination). Tumor formations had smooth surfaces, fibroelastic consistency, discreetly erythematous, well circumscribed. With the patient's written consent, a punch biopsy of the described changes was performed under colposcope control with 2% local lidocaine anesthesia. The sample obtained by punch biopsy was sent for pathohistological analysis. The histological diagnosis showed that it was syringocysadenoma papilliferum of the vulva.

Pathological-anatomical analysis indicated that these are tissue particles whose surface is partially covered by unevenly thickened, multi-layered epithelial epithelium. In the underlying stroma, mutually anastomosing adenoid formations were seen, surrounded by cylindrical cells, surrounded by an inflamed stroma with a dominance of plasma cells. The patient was presented at the gynecological surgery council, where it was decided that the patient should undergo surgical excision of the described changes. The operation was performed in our clinic following the guidelines of asepsis and antisepsis, in the form of surgical excision of the tumor in toto under 2% lidocaine local anesthesia.

Solution

The lesion was thoroughly and extensively excised, and the sample was sent for pathohistological analysis. The wound is closed with a pair of single sutures and sterile bandaged. Postoperatively, cefazolin 2 g i.v. was prescribed. in a single dose. The resection margins are without visible pathological changes. The patient recovered well and was discharged home the same day. At the follow-up examination, on the tenth postoperative day, the patient was free of symptoms and complaints, with an orderly healing of the operative wound. The patient was given guidelines for a three-month follow-up. At the follow-up examination, the patient had no complaints and the wound had healed well.

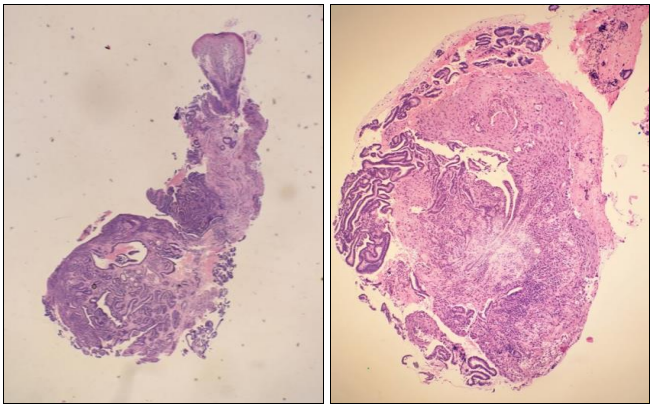

Fig 1: Microscopic views of changes of SCAP on the vulva

Discussion

Syringocystadenoma papilliferum is a rare benign hamartomatous adnexal tumor of apocrine or eccrine sweat glands. Most patients have a solitary lesion in the head and neck area [11]. Presentation outside the head and neck area is still quite rare. In our case report, we presented the appearance of Syringocystadenoma papilliferum with a papulonodular lesion on the vulva of a 64-year-old woman. According to the available data, this case was diagnosed for the first time at the Clinic for Gynecology and Obstetrics Clinical University Center Sarajevo, and through this case report we would like to highlight our experience with the diagnosis and treatment of syringocystadenoma papilliferum vulva. Epidemiological data in the literature show that 50% of cases are diagnosed in people at birth or in early childhood, while 15-30% of cases develop during puberty. It occurs more often in women than in men [12, 13]. In the vulvar region is an extremely rare condition in gynaecological practice with a few cases identified in the literature. These cases were reported in women between 27 and 64 years old.

Clinically, it presents as a papule or plaque and increases in size after puberty to form a papillomatous and crusted lesion. It is usually less than 4 cm in diameter. It is extremely rarely manifested as a multiple lesion. Macroscopically, it manifests as a pink plaque or lump without hair. It can be manifested as a solitary papule or a smooth plaque, up to 4 cm in size, or a papule with a linear arrangement, rarely with the appearance of a verrucous change and often in the environment of nevus sebaceus [17]. A hemorrhagic crust may develop on the surface of syringocystadenoma papilliferum.

Microscopically, it is visualized as cystic invaginations of the infundibular epithelium extending into the dermis with a double layer of inner squamous and outer cuboidal cells [18]. The pathophysiology of syringocystadenoma papilliferum indicates that it is a benign hamartomatous adnexal tumor of unclear origin. It can come from apocrine glands, eccrine glands or both. Immunohistochemical studies support an apocrine origin, while ultrastructural analysis favors an eccrine derivation. It can arise from pluripotent stem cells [19, 20]. Syringocystadenoma papilliferum is often observed in combination with other benign adnexal neoplasms, such as nevus sebaceus (8-19%), apocrine nevus, tubular apocrine adenoma, apocrine hydrocyst, apocrine cystadenoma and clear cell syringoma. In one out of ten cases, it is associated with basal cell carcinoma. Differential diagnosis syringcystadenoma papilliferum should be distinguished from hidradenoma papilliferum which is usually seen in the perineal region. Syringocystadenoma papilliferum should also be distinguished from adenocarcinoma, in which there is pronounced nuclear atypia, an infiltrative growth pattern, and a lack of myoepithelial layer. Syringocystadenoma papilliferum can be associated with malignant tumors such as verrucous carcinoma, basal cell carcinoma, sebaceum carcinoma and ductal carcinoma [21] Excisional biopsy has proven to be the treatment of choice for this disease. Guidelines for therapy have not yet been systematically developed due to the rare occurrence of this lesion. We believe that the changes should be completely surgically removed due to the risk of malignant alteration to adenocarcinoma.

Lessons Fot the Field

Syringocystadenoma papilliferum rarely occurs on the genitals. Due to such a rare occurrence, a broad differential diagnosis should be taken into account in the treatment of the patient and a histopathological examination of the sample should be performed to rule out malignancy. Excisional biopsy is the treatment of choice.

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