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Case Series

Case Series

Syringocystadenoma papilliferum: case series and review of the literature

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Abstract

Syringocystadenoma papilliferum (SCAP) is a rare, hamartomatous tumor of the apocrine glands, which typically manifests in the head and neck region. We present a case of 60-year-old male with a several-year history of the lesion located on the abdominal wall and a second case of a 58-year-old male with a history of a slow-growing lesion located on the tragus. Despite varying presentations and locations, both patients were identified to have SCAP on pathological evaluation. Appropriate treatments of SCAP range from CO2 laser treatment to surgical excision; we recommend surgical excision due to the risk of malignant transformation.

INTRODUCTION

Syringocystadenoma papilliferum (SCAP) is a rare, hamartomatous tumor of the apocrine glands that predominantly occur in the head and neck but may occasionally appear in other regions of the body. The majority of these lesions are present at birth or develop during adolescents. Presentation and development in adults have also been reported but are significantly rarer. Due to its rarity, SCAP can often be misdiagnosed and overtreated. However, it is prudent to be aware that SCAP lesions do carry the risk of malignant transformation [1].

CASE REPORTS

Case 1

A 60-year-old male presented with a long-term history of a left lower quadrant cyst located at the belt line associated with spontaneous drainage. The cyst slowly increased in size and recently demonstrated drainage episodes prompting the patient to pursue a surgical evaluation. The patient's past medical history was remarkable for diabetes and gastroesophageal reflux disease. Furthermore, the patient's previous surgical history included an appendectomy and an inguinal hernia pending intervention. The lesion was initially presumed to be infected secondary to its drainage and treated with antibiotics; however, on surgical evaluation, the lesion did not possess the qualities consistent with an abscess. Intraoperatively, a 3.2 cm × 1.2 cm × 0.9 cm lesion was removed utilising an elliptical incision, ensuring that the cyst capsule was removed in its entirety. On a pathology review, the section revealed a pasty yellow-white substance and was deemed to be SCAP, which was confirmed with an expert review.

Case 2

A 58-year-old male was diagnosed with a 3-month history of a friable, tender right tragus mass measuring approximately 1 cm. The patient's past medical history was remarkable for a 26-pack-year smoking history. The patient's surgical history included an appendectomy and a bone growth removal, for which pathology was not available to us. Intraoperatively, a 0.5 cm × 0.5 cm lobulated right ear canal mass was removed. On pathological review, the mass appeared to have verruciform squamous proliferation with hyperkeratosis and underlying papillomatous proliferations that communicated with the overlying skin. The mass also demonstrated a bilayered columnar appearance, with mononuclear inflammatory infiltrates consisting predominantly of plasma cells. The lesions were diagnosed as a SCAP.

DISCUSSION

SCAP is a rare, hamartomatous tumor that arises from the apocrine glands with an underlying risk of malignant transformation. Malignant transformation typically results in basal cell carcinoma, but reports of squamous cell carcinoma have also been made [1–3]. There are three different classifications of SCAP: solitary nodular, linear and plaque. The solitary type presents as pedunculated nodules ranging from 5 to 10 mm in size and manifests near the proximal joints of the upper extremities. The linear type presents as multiple erythematous papules ranging from 1 to 10 mm in size. Lastly, the plaque type is evidenced by an alopecic area with nodular or crusted plaques [4, 5]. A majority of reported cases occur in the head and neck region, with approximately 30% of cases occurring in the trunk and extremities demonstrating brown, erythematous skin lesions of various sizes. SCAP frequently demonstrates cystic invaginations with a background of fibrous tissue. The superior and inferior portions of the cystic invaginations each possess unique characteristics, for the superior aspect is lined with squamous epithelium and the inferior aspect possesses granular epithelial projections [5]. SCAPs present as single or multiple nodular plaque lesions, 40%

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of which are reportedly associated with a subcutaneous nevus. In sporadic cases, BRAF and HRAS mutations have been identified [6, 7]. Approximately 50% occurs at birth, up to 30% in adolescence, and the remainder in adulthood. The infantile disease presents as alopecic yellow plaques, the adolescent disease is governed by androgen release resulting in hyperkeratosis and sebaceous gland formation, and the adult disease manifests as hamartomatous apocrine glands accompanied by epidermal and sebaceous glands [7]. SCACP is a rare neoplastic differentiation of the apocrine glands, a limited number of cases are available and have been reported in the literature ranging from carcinomas in situ to the malignancy associated regional lymph node metastases [2, 3]. SCACP should be suspected in lesions with ulcerations and rapid progression. Although successful management of SCAP has been reported with Mohs surgery and CO2 laser treatment, surgical excision is ultimately advised due to the risk of malignant transformation [8].

CONCLUSION

SCAP typically presents as a slow-growing, skin-colored lesion, which often begins oozing serous fluid or blood. Although SCAP is a largely benign hamartomatous tumor, it does possess the risk of malignant transformation. As a result, we recommend surgical excision in order to prevent the lesion from evolving into a more insidious pathology. These lesions often present in the infantile stage on the head and neck region; however, we present two cases of a sporadic presentation on the abdomen and tragus of adult males.

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CONFLICT OF INTEREST STATEMENT

None declared.

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INFORMED CONSENT

All attempts have been exhausted in trying to contact the patient, next of kin and/or parent/guardian for informed consent to publish their information, but consent could not be obtained.

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