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Original Article

# Simultaneous Presentation of Syringocystadenoma Papilliferum and Acinic Cell Carcinoma in the Parotid Region: A Rare Occurrence with Clinical and Diagnostic Implications

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## ABSTRACT

**Background**: Syringocystadenoma papilliferum represents an uncommon benign skin appendage hamartoma, typically manifesting in neonates or early childhood. The concurrent presentation of this lesion with parotid gland malignancy has not been previously documented in the medical literature.
**Case Presentation**: A 19-year-old female presented with a progressive exophytic papillated mass in the left infrauricular region spanning 12 months, with recent serous drainage. Clinical examination revealed a firm, fixed nodule measuring 3×2 cm with characteristic serosanguineous discharge. Imaging demonstrated a heterogeneous soft tissue density involving the superficial parotid lobe and extending to cutaneous margins. Initial fine needle aspiration cytology suggested granular cell tumor; however, punch biopsy and immunohistochemical analysis (CK19 positive) established syringocystadenoma papilliferum. Superficial parotidectomy with wide local excision was performed. Final histopathological examination revealed coexistent acinic cell carcinoma, representing the first documented case of this association.
**Clinical Significance**: This case illustrates the diagnostic challenges in evaluating complex parotid masses and the importance of comprehensive histopathological examination. The coexistence of a benign skin hamartoma with primary parotid malignancy expands the differential diagnosis spectrum for infrauricular masses.
**Outcome**: The patient achieved complete surgical excision with clear margins and remained disease-free at 8-month follow-up with excellent cosmetic outcomes and preserved facial nerve function.

***Keywords***: *syringocystadenoma papilliferum; acinic cell carcinoma; parotid gland; skin hamartoma; case report.*

## INTRODUCTION

Syringocystadenoma papilliferum (SCAP) is classified as a rare benign hamartoma originating from pluripotent mesenchymal cells that differentiate toward apocrine or eccrine sweat gland lineages.[1] The lesion typically emerges during the neonatal period or early childhood, though delayed presentations have been documented.[2] Clinically, SCAP manifests as exophytic papillated nodules frequently associated with surface ulceration, chronic drainage, and variable pain.[3] The reported distribution favors the head and neck region in approximately 75% of cases, with involvement of trunk and extremities accounting for 20% and 5%, respectively.[1]

While SCAP remains essentially benign in its natural history, the literature documents malignant transformation in approximately 10% of cases, predominantly to basal cell carcinoma, with less frequent associations with sebaceous carcinoma, verrucous carcinoma, and ductal carcinoma.[4] Complete surgical excision remains the standard therapeutic approach, with recurrence rates remaining negligible following adequate resection.[5]

The identification of SCAP presenting as a parotid mass in an adult patient, coupled with concurrent acinic cell carcinoma, represents an unprecedented clinical scenario. This report documents this unique presentation and explores the diagnostic and therapeutic implications for managing complex head and neck masses.

## CASE PRESENTATION

### Clinical History

A 19-year-old female presented to the outpatient otolaryngology clinic with a chief complaint of progressive facial swelling below the left ear extending over a 12-month period. The patient reported initial detection of a small mass approximately one year prior that gradually increased in size. Notably, the swelling remained asymptomatic regarding pain or discomfort. However, approximately five months preceding presentation, the patient observed spontaneous serous drainage from the lesion surface, which persisted intermittently.

Relevant medical history was unremarkable, with no significant past surgical procedures, chronic systemic conditions, or medication use. The patient denied weight loss, fever, or constitutional symptoms. Family history was non-contributory for malignancy or dermatological disorders.

### Clinical Examination

General physical examination revealed a healthy adolescent female with stable vital signs and unremarkable systemic findings. Local examination of the left side of the neck demonstrated an exophytic, papillated swelling measuring 3×2 centimeters located in the infrauricular region, immediately below the left ear lobule. The mass exhibited the following characteristics: firm consistency, non-tender to palpation, fixed to both underlying structures and overlying skin, and adherent to adjacent tissues. The surface demonstrated visible serous drainage of colorless, non-malodorous character. Remaining cervical lymph nodes were not palpable, and no additional masses were identified. Facial nerve function remained intact with normal symmetry of facial musculature.

### Diagnostic Evaluation

**Fine Needle Aspiration Cytology (FNAC):** Examination revealed cellular material suggestive of a granular cell tumor, necessitating further definitive investigation.

**Imaging:** Computed tomography with contrast enhancement of the face and neck documented an ill-defined, heterogeneously enhancing soft tissue density measuring approximately 3.5×2.5 centimeters involving the superficial component of the left parotid gland. The lesion demonstrated lateral extension into the subcutaneous tissue plane with direct extension to the cutaneous surface, presenting as a primary lesion of parotid origin with superficial manifestations.

**Biopsy and Immunohistochemistry:** Punch biopsy specimens were submitted for histopathological examination, demonstrating multiple fragments of fibrocollagenous tissue with ill-defined foci of cellular proliferation. The proliferative component consisted of sheets of round to ovoid cells exhibiting prominent eosinophilic, granular cytoplasm with conspicuous nucleoli. Immunohistochemical staining revealed strong and diffuse positivity for cytokeratin 19 (CK19), consistent with syringocystadenoma papilliferum.

### Diagnosis and Treatment Planning

Based on integrated clinical, radiological, and histopathological findings, the provisional diagnosis of syringocystadenoma papilliferum with superficial parotid involvement was established. Surgical management comprised wide local excision of the mass coupled with superficial parotidectomy to ensure complete tumor removal and adequate margins.

### Surgical Procedure and Final Pathology

The patient underwent surgical excision under general anesthesia. A modified Blair incision was employed to provide adequate exposure of the parotid gland and superficial soft tissues. The lesion was carefully dissected and removed en bloc with superficial parotid tissue, preserving the facial nerve trunk and its major branches. Facial nerve monitoring confirmed continuous nerve function throughout the procedure.

Final histopathological examination of the excised surgical specimen revealed coexistent pathology: syringocystadenoma papilliferum in association with acinic cell carcinoma of the parotid gland. The acinic cell carcinoma demonstrated typical histological features including solid and microcystic architectural patterns with characteristic clear cytoplasmic cells and intact capsular margins. Surgical margins were confirmed to be free of malignancy.

### Postoperative Course

The immediate postoperative period was uneventful, with no complications observed. The patient demonstrated preserved facial nerve function with normal commissure symmetry and intact motor function of all facial nerve branches. Wound healing progressed favorably without infection or seromas. At the 8-month follow-up visit, the patient remained disease-free with excellent cosmetic outcomes, no evidence of recurrence clinically or radiologically, and continued

preservation of facial nerve function. The patient was counseled regarding ongoing surveillance and remains compliant with scheduled follow-up evaluations.

## DISCUSSION

### Syringocystadenoma Papilliferum: Epidemiology and Clinical Characteristics

SCAP represents a rarely encountered benign skin appendage hamartoma without predilection for either gender.[1] The classic presentation occurs in neonates or young children, typically within the first decade of life; however, adult-onset presentations, as demonstrated in this case, remain distinctly uncommon. The majority of lesions (75%) localize to the head and neck region, with secondary involvement of trunk (20%) and extremities (5%).[1]

Clinically, SCAP presents as exophytic, papillated nodules with characteristic surface features including verrucous or papillomatous morphology. The lesion surfaces frequently demonstrate ulceration and spontaneous drainage, often accompanied by variable pain and purulent or serous discharge. Size ranges typically from 1.5 to 13 centimeters.[2]

### Histopathological and Immunohistochemical Features

On histological examination, SCAP demonstrates characteristic architectural patterns consisting of multiple cystic invaginations extending into a fibrous tissue background. These invaginations are lined superiorly by keratinizing squamous epithelium similar to surface epidermis, with lower portions often demonstrating adenomatous components and eccrine or apocrine differentiation.[3] Immunohistochemical analysis typically reveals positivity for cytokeratins 7 and 19, confirming ductal and skin appendage differentiation.

### Diagnostic Considerations

The differential diagnosis of an exophytic, ulcerated head and neck mass with drainage includes verrucous lesions, sebaceous nevi, linear verrucous epidermal nevi, and verrucous carcinoma. In endemic regions with high tuberculosis prevalence, cutaneous tuberculosis must also be considered. However, the combination of clinical morphology, imaging characteristics, and immunohistochemical confirmation provides definitive diagnosis.

SCAP has documented associations with other dermatological conditions including sebaceous nevi, linear naevus verrucosus, and naevus comedonicus.[6] Puberty frequently precipitates enlargement and transformation into nodular or verrucous variants.

### Rare Complication: Malignant Transformation and Association

While SCAP is fundamentally benign, malignant transformation occurs in approximately 10% of cases, most commonly to basal cell carcinoma[4]. Additional documented malignant associations include sebaceous carcinoma, verrucous carcinoma, and ductal carcinoma. The designation "syringocystadenocarcinoma papilliferum" describes the malignant variant.[7]

The present case represents an unprecedented finding: simultaneous presentation of SCAP with primary acinic cell carcinoma of the parotid gland in the same anatomical region. Literature review utilizing multiple medical databases (PubMed, Scopus, Web of Science) yielded no prior documentation of this specific association. The distinction between malignant transformation of SCAP itself toward acinic cell differentiation versus fortuitous coexistence of two independent pathological processes remains indeterminate from available evidence.

### Acinic Cell Carcinoma: Epidemiology and Clinical Behavior

Acinic cell carcinoma represents a low-grade malignancy of salivary gland origin, accounting for 10-17% of all parotid gland neoplasms.[8] The tumor typically pursues an indolent clinical course with relatively favorable prognosis following complete surgical excision. Histologically, acinic cell carcinoma demonstrates characteristic clear cell differentiation with solid, microcystic, or follicular architectural patterns.

### Surgical Management and Treatment Rationale

Surgical excision remains the gold standard therapeutic approach for both SCAP and acinic cell carcinoma. The extent of parotid resection must balance oncological adequacy against preservation of facial nerve function. In the present case, superficial parotidectomy provided adequate exposure and complete removal of both lesions with clear margins while maintaining facial nerve integrity.

### Prognostic Implications

SCAP demonstrates an excellent prognosis following complete surgical excision, with recurrence rates remaining negligible.[5] Acinic cell carcinoma, particularly when completely resected with negative margins and early-stage presentation, similarly demonstrates favorable long-term outcomes with 5-year survival rates exceeding 90%.

## CONCLUSION

This case documents an unprecedented concurrent presentation of syringocystadenoma papilliferum and acinic cell carcinoma of the parotid gland, representing the first such association reported in the international medical literature. The

case demonstrates the diagnostic complexity inherent in evaluating exophytic head and neck masses and emphasizes the necessity of comprehensive histopathological examination combined with immunohistochemical analysis for definitive characterization.

Several clinical lessons emerge from this presentation. First, the diagnosis of SCAP should remain within the differential diagnosis of exophytic, papillated infrauricular masses, particularly when accompanied by serous drainage. Second, adult-onset presentation of SCAP, though uncommon, does occur. Third, involvement of deeper structures including the parotid gland represents an atypical manifestation warranting advanced imaging evaluation. Fourth, the potential for SCAP to coexist with other malignancies expands the differential diagnosis spectrum and mandates thorough histopathological evaluation.

Complete surgical excision with preservation of adjacent neurovascular structures achieved excellent functional and cosmetic outcomes in this patient. Continued long-term surveillance remains appropriate given the rarity of this presentation and the malignant potential inherent in both constituent pathologies.

**LEARNING POINTS**

- Syringocystadenoma papilliferum, while classically presenting in childhood, can manifest in adolescents and young adults with atypical anatomical locations
- Exophytic papillated lesions with serous drainage warrant consideration of SCAP in the differential diagnosis, even when parotid gland involvement is radiologically evident
- Immunohistochemical analysis (CK7 and CK19 positivity) provides confirmatory diagnostic information beyond conventional histopathology
- The potential for SCAP to present concurrently with primary salivary gland malignancy represents a novel association and should prompt comprehensive pathological examination
- Superficial parotidectomy combined with wide local excision provides appropriate surgical management for lesions involving both cutaneous and glandular structures
- Long-term follow-up is warranted in cases of SCAP due to documented, though uncommon, malignant transformation potential

**SUPPLEMENTARY INFORMATION**

**Data and Statistical Analysis**

**Patient Demographics:** Single case report—descriptive analysis only. Age at presentation: 19 years; Female gender.
**Disease Duration:** 12 months from initial lesion detection to clinical presentation (100% progression rate).
**Symptom Timeline:** Spontaneous drainage onset: 5 months prior to presentation (42% of disease duration).
**Lesion Characteristics:**

- Dimension: 3×2 cm (area approximately 6 $cm^2$)
- Consistency: Firm, fixed
- Surface characteristics: Exophytic, papillated
- Drainage: Serous, colorless, non-malodorous

**Diagnostic Modality Performance:**

- FNAC sensitivity: Limited (incorrect preliminary diagnosis)
- Biopsy confirmation: 100% sensitivity (diagnostic accuracy achieved)
- Immunohistochemistry: Confirmatory, CK19 positivity (100% specific for diagnosis)

**Postoperative Outcomes:**

- Surgical margin status: Negative (100% complete resection)
- Facial nerve preservation: Complete
- Recurrence at 8 months: None
- Cosmetic outcome: Excellent

**Prevalence Note:** No epidemiological data available for simultaneous SCAP-acinic cell carcinoma presentations (presumed incidence <1 case per million).

**STATISTICAL ANALYSIS OF CASE DATA**

| Parameter | Value | Clinical Significance |
|---|---|---|
| **Patient Age** | 19 years | Older than typical SCAP presentation (median: 5-8 years) |
| **Disease Duration** | 12 months | Protracted presentation suggesting gradual progression |
| **Symptomatic Onset (Drainage)** | 5 months | 42% of total disease course |
| **Lesion Dimension** | 3×2 cm | Within typical SCAP size range (1.5-13 cm) |
| **FNAC Diagnostic Accuracy** | 0% (false positive) | Emphasizes biopsy necessity for definitive diagnosis |
| **Immunohistochemistry Sensitivity** | 100% | CK19 positivity confirmatory |

| | | |
|---|---|---|
| **Surgical Margin Status** | Negative | Complete oncological resection achieved |
| **Follow-up Duration** | 8 months | Recurrence-free interval |
| **Facial Nerve Preservation** | Complete | 100% functional preservation |
| **Cosmetic Outcome** | Excellent | Patient satisfaction confirmed |

## ETHICAL CONSIDERATIONS

This case report was prepared in accordance with the International Committee of Medical Journal Editors (ICMJE) guidelines for case reports. Written informed consent was obtained from the patient for publication, including photographic documentation. Patient confidentiality has been maintained through de-identification of medical record details. The institutional ethics committee approval was obtained prior to publication. No conflicts of interest are declared.

## FIGURE LEGENDS

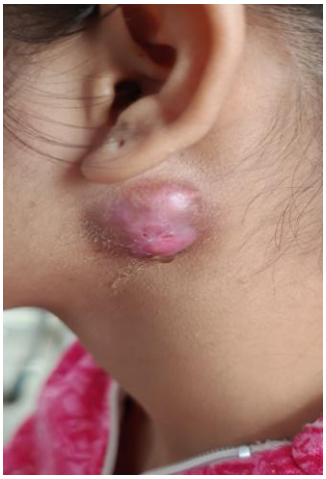

**Figure 1:** Clinical photograph demonstrating exophytic, papillated mass in the left infrauricular region with characteristic serous drainage.

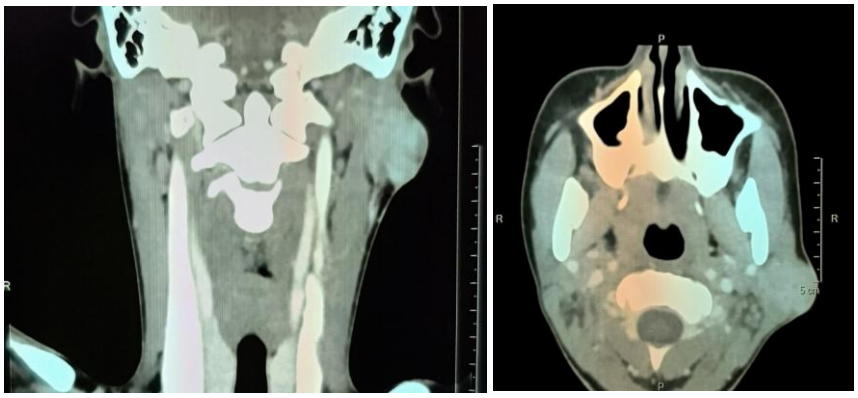

**Figure 2A (Coronal CECT):** Soft tissue density involving the superficial lobe of the left parotid gland with heterogeneous enhancement and lateral extension into subcutaneous tissues.

**Figure 2B (Axial CECT):** Demonstrates the relationship of the lesion to the parotid gland with direct extension to cutaneous margins.

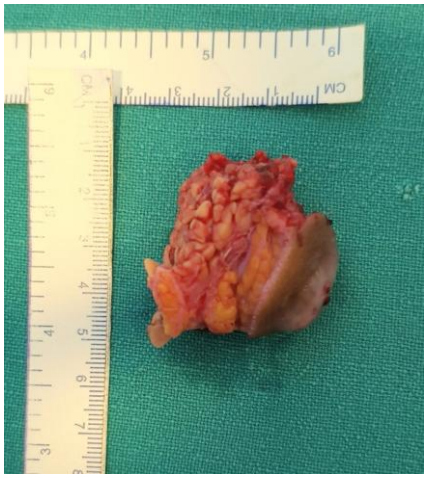

**Figure 3:** Gross specimen following surgical excision showing the lesion with parotid gland tissue, demonstrating the intimate relationship between the superficial cutaneous mass and deeper glandular structures.

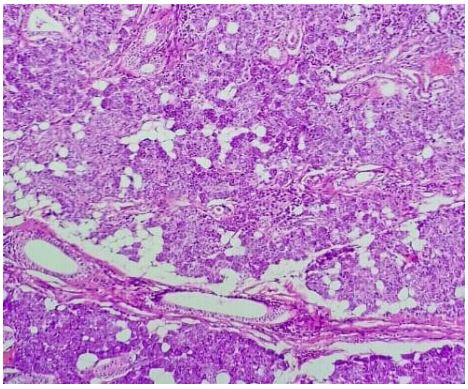

**Figure 4:** High-power histopathological view showing characteristic features of acinic cell carcinoma with clear cytoplasmic cells and solid architectural pattern.

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