

Endocrine Mucin-Producing Sweat Gland Carcinoma of the Eyelid with Progression to Invasive Mucinous Adenocarcinoma

A Rare Entity with Breast Carcinoma Analogy — Case Report

Yaren Guven, MD · Fatma Sumer, MD · Cigdem Ozturk, MD
Department of Ophthalmology, Recep Tayyip Erdogan University, Rize, Turkey

POSTER #61

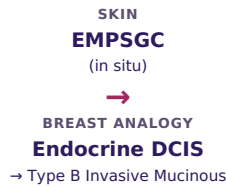
EMPSGC = cutaneous equivalent of endocrine ductal carcinoma in situ of the breast — neuroendocrine differentiation, ER/PR positivity, potential for invasive progression

BACKGROUND — EMPSGC

WHAT IS EMPSGC?

<100 cases worldwide

Low-grade neuroendocrine cutaneous neoplasm · Strong eyelid predilection · Dual IHC phenotype



Key question: True progression or de novo invasive carcinoma?

CASE PRESENTATION

PATIENT

72-year-old male

Left upper eyelid · Slowly enlarging × 8 months · Firm, non-tender

INITIAL IMPRESSION

Epidermal inclusion cyst

Classic misdiagnosis — progressive enlargement in elderly should prompt excision

MANAGEMENT

Wide excision — 4 mm margins

Advancement flap · Full IHC panel · PET-CT systemic staging

HISTOPATHOLOGY — BIPHASIC PROGRESSION PATTERN

SUPERFICIAL — EMPSGC (IN SITU)

Solid nests + papillary structures of **low-grade epithelial cells** with intracytoplasmic mucin · Characteristic neuroendocrine architecture



DEEP — INVASIVE MUCINOUS ADENOCARCINOMA

Extracellular mucin pools with floating tumour cell clusters · Transition zone clearly identifiable — supports progression model

✓ **Surgical margins: CLEAR on permanent section histopathology**

IMMUNOHISTOCHEMISTRY

NEUROENDOCRINE

- ✓ Chromogranin
- ✓ Synaptophysin

HORMONE

- ✓ ER (+)
- ✓ PR (+)
- ✓ CK7 (+)

NEGATIVE

- ✗ CK20
- ✗ CDX2

Chromogranin/synaptophysin positivity distinguishes EMPSGC from metastatic breast mucinous carcinoma — even with ER/PR positivity.

SYSTEMIC WORKUP & OUTCOME

✓ **PET-CT** — No breast, GI or distant primary

✓ **Male breast** — No uptake (mandatory even in males)

→ **PRIMARY cutaneous origin confirmed**




No recurrence
24 months



No metastasis



Full lid function

 This case documents **in situ EMPSGC progressing to invasive mucinous adenocarcinoma** — the rarest progression in an already rare tumour. The **visible transition zone** and neuroendocrine marker positivity in both components support a progression model paralleling breast carcinoma biology. **Clonality studies** (NGS, LOH) would confirm shared origin in future cases.

TAKE-HOME MESSAGES

- 1** EMPSGC is a **rare neuroendocrine eyelid carcinoma** — any firm, slowly enlarging upper eyelid nodule in an elderly patient should be excised and submitted for histopathology with full IHC panel including neuroendocrine markers
- 2** EMPSGC **can progress to invasive mucinous adenocarcinoma**, directly paralleling endocrine DCIS in breast cancer — a unique progression with biphasic histopathological pattern
- 3** **Dual IHC phenotype** is the diagnostic key: neuroendocrine markers (chromogranin + synaptophysin) PLUS hormone receptors (ER/PR) — neuroendocrine positivity is absent in metastatic breast mucinous carcinoma
- 4** **Complete excision with clear margins is curative.** Invasive component warrants enhanced surveillance. Systemic workup mandatory even in male patients when ER/PR positive mucinous carcinoma identified