Aggressive Syringomatous Carcinoma of the Orbit

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ABSTRACT

A 68-year-old woman with an unusual tumor involving the right orbit presented with painful exophthalmos of the right eye. Excision biopsy of her right eyelid was performed. The specimen showed ductal differentiation with comma-like extensions identical to syringoma. Cellular atypia, an invasive growth pattern, and remarkable perineural invasion led to the diagnosis of syringomatous carcinoma. Magnetic resonance imaging showed that the tumor exhibited maxillary involvement, invading the supraorbital fissure and bone of the middle skull base. The patient underwent craniofacial resection and has been alive with no sign of recurrence for 2 years. This case suggests that wide excision of these tumors with a clear surgical margin using skull base surgical techniques may offer a good prognosis.

KEYWORDS: Syringomatous carcinoma, eccrine sweat glands tumor, orbital tumor, skull base surgery

Syringomatous carcinomas are rare tumors derived from eccrine sweat glands. They were first described by Goldstein et al¹ in 1982 as “microcystic adnexal carcinoma.” These tumors most frequently occur on the scalp^{2–4} and face^{5–7}; occasionally they involve the periorbital area (Table 1).^{5,8–17} We describe one case with a syringomatous carcinoma of the orbit that demonstrates the locally aggressive nature of these tumors.

CASE REPORT

In February 2003, a 68-year-old woman sought treatment after experiencing a painful lesion in her right orbit for 3 months. Her physical examination revealed exophthalmos of the right eye. Lymphadenopathy was not observed in the neck. Magnetic resonance imaging showed a tumor with maxillary involvement and invading the supraorbital fissure

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Table 1 Syringomatous Carcinoma of the Eyelid: Reported Cases

Authors	Age/Sex	Orbital Involvement	F/U	Results
Cooper et al ⁵	20/F	—	3 yr	ANED
Glatt et al ⁸	20/F	—	3 yr	ANED
Glatt et al ⁸	18/M	—	2 yr	ANED
Hoppenreijns et al ⁹	50/F	—	—	ANED
Hoppenreijns et al ⁹	83/F	+	—	AWD
Hoppenreijns et al ⁹	54/M	+	—	DOD
Khalil et al ¹⁰	70/M	+	4 yr, 6 m	DOD
Rosen et al ¹¹	47/M	—	4 yr	AWD
Urso et al ¹²	84/F	—	—	—
Ishikawa et al ¹³	73/M	+	1 yr	DOD
Hesse et al ¹⁴	35/F	—	2 yr	ANED
Hunts et al ¹⁵	61/M	—	—	ANED
Leboit and Sexton ¹⁶	33/F	—	2 m	ANED
Mayer et al ¹⁷	82/M	+	2 yr, 8 m	DOD
Present Case	68/F	+	1 yr, 6 m	ANED

ANED, alive with no evidence of disease; AWD, alive with disease; DOD, died of disease; F/U, follow up; yr, year; m, month.

(Fig. 1). Computed tomography showed that the tumor infiltrated the bone of her right skull base (Fig. 1). An excisional biopsy of her right eyelid was performed at the Department of Ophthalmology and the histopathological diagnosis was a syringomatous carcinoma.

The patient underwent craniofacial resection and reconstruction using a rectus abdominis musculocutaneous free flap. Briefly, after a semicoronal skin incision was made, the zygomatic arch and coronoid process were removed (Fig. 2). A frontotemporal craniotomy was performed and the brain was elevated from the lateral base to the frontal base of the skull. Cranial nerves II, III, IV, VI, VI and the ophthalmic artery were resected at the supra-orbital fissure. Osteotomy of the skull base bone was performed through the right foramen rotundum, supraorbital fissure, and frontal skull base (Fig. 3). After a modified Weber's incision was made, the tumor was resected en bloc with a clear margin (Fig. 4). The skull base and facial skin defects were reconstructed using a rectus abdominis musculocutaneous free flap (Fig. 5).

The tumor, which measured by $3 \times 3 \times 2$ cm, showed ductal differentiation with comma-like ex-

tensions identical to syringoma (Fig. 6). Cellular atypia, invasive growth pattern, and remarkable perineural invasion were suggestive of malignancy. These features led to the diagnosis of syringomatous carcinoma. The carcinoma extended deep into maxillary bone (Fig. 6).

The patient's postoperative course was uneventful. She has been followed for 2 years at our outpatient clinic with no signs of a recurrence.

DISCUSSION

Syringomatous carcinomas have many synonyms such as carcinomas with eccrine differentiation, sclerosing sweat duct syringomatous carcinomas, syringoid eccrine carcinomas, microcystic adnexal carcinomas, basal cell tumors with eccrine differentiation, eccrine epitheliomas, sweat gland carcinomas with syringomatous features, and malignant syringomas.^{1-3,5,8} These carcinomas are now thought to share the same neoplastic process and should be termed "syringomatous carcinomas." This term has been suggested by Abenoza and

Figure 1 Preoperative imaging. (A) Axial MRI with gadolinium contrast showed an enhancing mass invading the supraorbital fissure. (B) Axial contrast computed tomographic scan showed a large mass destroying maxillary bone by direct extension.

Ackerman⁷ to avoid complex classifications. Based on their proposal, syringomatous carcinomas can be classified into three groups: well differentiated, moderately differentiated, and poorly differentiated. The well-differentiated form is found more frequently than the other two forms.

Histopathologically, syringomatous carcinomas are characterized by (1) cords and nests of cytologically uniform keratinocytes, (2) keratotic and cyst-like structures, (3) variable ductal differentiation, (4) sclerotic collagenous stroma, (5) infiltration of subcutaneous tissue, and (6) perineural invasion.⁵

Syringomatous carcinomas are thought to occur in the middle-aged and elderly and to show

Figure 2 Illustration showing skin incision and craniotomy. After a semicoronal skin incision was made, the zygomatic arch and coronoid process were removed. Then, a frontotemporal craniotomy was performed. Asterisk indicates resected facial skin.

a predilection for the scalp²⁻⁴ and face.⁵⁻⁷ The gender distribution is about equal. Many patients are aware of their neoplasm for many years because most syringomatous carcinomas are indolent and grow slowly. Surgical excision is the most reliable method of eradicating syringomatous carcinomas, as is the case with other eccrine sweat gland carcinomas. The rate of tumor recurrence is high reflecting extensive perineural invasion, but regional or distant metastases are rare.^{5,9}

Figure 3 Illustration showing skull base osteotomy. The osteotomy (shading) was performed through the right foramen rotundum, supraorbital fissure, and frontal skull base.

Figure 4 Illustration showing en bloc resection (shading) of the maxilla with orbital contents and skull base.

Figure 5 Postoperative computed tomographic scan. The skull base and facial skin defects were reconstructed using a rectus abdominis musculocutaneous free flap.

Figure 6 Photograph of the pathological specimen. (A) Low power. (B) High power. The tumor showed ductal differentiation with comma-like extensions.

Radiotherapy has been reported as insufficient to treat this carcinoma.¹⁷⁻¹⁹ In two cases treated with radiotherapy for primary microcystic adnexal carcinoma, the tumor recurred within 3 years.^{18,19} One patient who underwent postoperative radiotherapy and chemotherapy consisting of tamoxifen also had recurrence despite the intensive treatment.²⁰

To date, 14 cases of syringomatous carcinoma arising in the eyelid have been reported.^{5,8-17} In 5 of these cases, the orbital content was involved. All relapsed after surgical treatment. These outcomes suggest that wide surgical excision using skull base surgical techniques, as in our case, should be

considered for the treatment of syringomatous carcinomas arising in the eyelid.

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