

# Unusual Longterm Survival of 3 Patients with Adenoid Cystic Carcinoma of the Lacrimal Gland

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## Background

The nation wide survey concerning malignant tumors of the lacrimal gland in Japan revealed the annual incidence of adenoid cystic carcinoma (ACC) was 0.0034/10<sup>6</sup>. (1, 2) Most of patients with ACC can not enjoy longterm survival without recurrence or metastasis inspite of mutilating surgery combined with radiotherapy as yet. (3) Therefore new approaches to get more satisfactory results are necessary for these patients suffering from ACC. We report our three happy patients who could survive ACC more than 10 years without recurrence or metastasis by undergoing only simple surgery.

## Cases

The author had treated 15 patients with ACC from 1973 to 2000 at National Cancer Center Hospital (NCCH) in Tokyo, Japan. The following 3 cases are among them. Our standard treatment of ACC was complete removal of the tumor by lateral or anterior orbitotomy and resection of the orbital wall invaded by ACC. Exenteration of the orbit was performed after the pathological diagnosis was established unless refusal of the patients or family was encountered.

### Case 1

**57 years, Female**  
**C. C:** Rt. Upper Lid Swelling  
**History:**  
 Apr. 1974 Swelling sensation of rt. upper lid  
 Sep. 1974 Rt. ocular pain & a small mass in the rt. upper lid  
 Sep. 20, 1974 Biopsy at a local hospital revealed ACC.  
 Oct. 15, 1974 Exenteration of the orbit & partial resection of the orbital wall at NCCH  
 Nov. 28, 1974 Free skin grafting  
 Nov. 13, 1998 Death without recurrence or metastasis  
**Past history & Family history:** Noncontributory

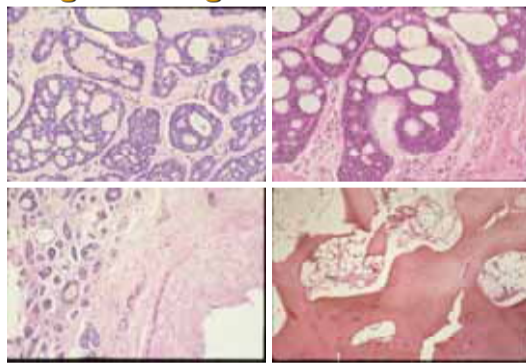
**Clinical Findings at the first visit**  
**Visual Acuity:** VD=0.2 (0.5X-2.0)  
 VS=0.2 (0.8X-2.0)  
**Exophthalmometry:** OD=16mm  
 OS=15mm  
**Ocular Movement & Eye Position:** Within Normal Limits  
**Ocular Fundus:** Within Normal Limits  
**Regional Lymph nodes:** not palpable

### Orbital Tomogram



Rt. Orbital bone invasion was suspected.

### Pathological Findings



Periorbital invasion No bone invasion

### 10 years after the surgery



### Case 2

**7 years, Female**  
**C. C:** Rt. Upper Lid Swelling  
**History:**  
 1 year ago C. C was regognized. 7 eye clinics could not find the cause.  
 Apr. 15, 1994 CT revealed rt. lacrimal mass.  
 May 15, 1994 It was removed with lateral orbitotomy to find ACC.  
 No additional treatment was performed because of desicion of the parents according to Dr. Shields's suggestion.  
 Jun. 10, 2007 No recurrence  
**Past history:** Noncontributory  
**Family history:** Father; American White, Mother; Japanese

**Clinical Findings at the first visit**  
**Visual Acuity:** VD=0.5 (0.7X+0.5, cyl+1.50, 100°)  
 VS=1.2 (n. c)  
**Exophthalmometry:** OD=19mm  
 OS=15mm  
**Ocular Movement:** Limited lateral gaze of OD & Diplopia  
**Ocular Fundus:** Choroidal folds of OD  
**Regional Lymph nodes:** not palpable

### Before the surgery

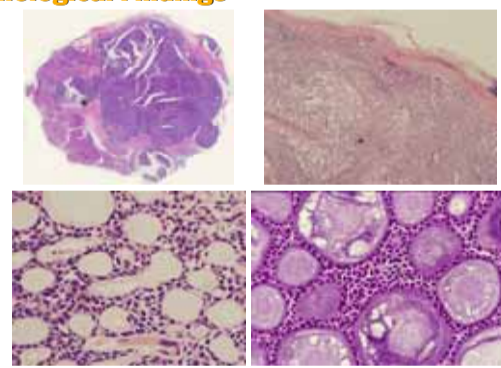


Swelling of the rt. Upper lid entropion of the rt. Lower lid

### CT before the surgery



### Pathological Findings



### 13 years after the surgery



### Case 3

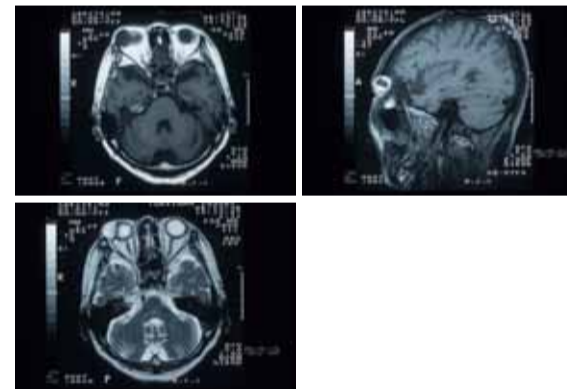
**80 years, Female**  
**C. C:** A Mass in Rt. Upper Lid  
**History:**  
 Mar. mid, 1996 C. C was noticed.  
 The mass was gradually enlarging.  
 Jun. 21, 1996 The first visit at NCCH  
 Sep. 19, 2000 Anterior Orbitotomy revealed ACC.  
 No additional treatment because of the age  
 May 30, 2007 No recurrence observed.  
**Past history:** Hypertention and Ischemic heart disease  
**Family history:** Noncontributory

**Clinical Findings at the first visit**  
**Visual Acuity:** VD=0.08 (0.5X+3.0, cyl+0.75, 1°)  
 VS=0.05 (0.7X+3.75, cyl+0.50, 170°)  
**OU:** Cataracta senilis  
**Ocular Movement:** Limited lateral gaze of OD  
**Ocular Fundus:** Choroidal folds of OD  
**Palpation:** 2.8×1.5cm mass in the rt. upper lid  
**Regional Lymph nodes:** not palpable

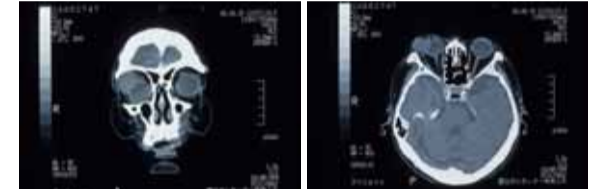
### Before the surgery (Jun. 12, 1996)



### MRI at the first visit



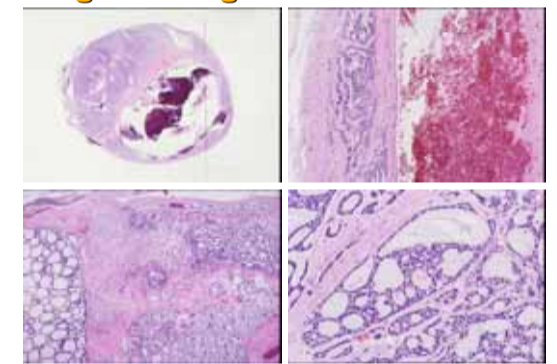
### CT before the surgery



### Echogram of the mass



### Pathological Findings



### One year after the surgery



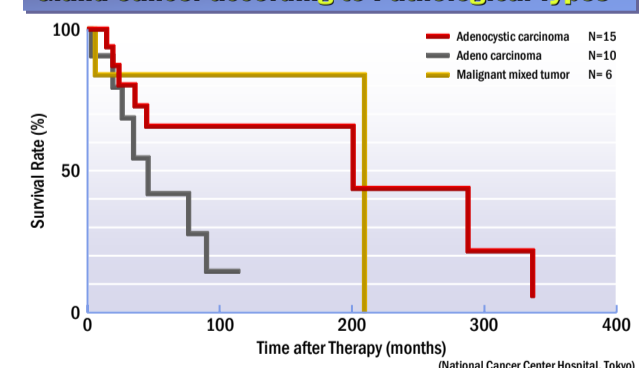
### MRI after the surgery



## Summary of Three Cases

Case	1	2	3
Age (years)	57	7	80
Gender	F	F	F
Time before First Visit (mo.)	5	12	2
Chief Complaint	Swelling of the upper lid	Swelling of the upper lid	Mass in the upper lid
Orbital Pain	(+)	(-)	(-)
Surgery	Exenteration + bone resection	Removal of the mass	Removal of the mass
Pathological Findings of ACC	Periorbital Invasion,	No Invasion	No Invasion
Survival Time After First Visit (years)	24	13	11

## Survival Curves of Patients with Lacrimal Gland Cancer according to Pathological Types



## Discussion

Why these 3 patients survived ACC without recurrence or metastasis in spite of undergoing only simple surgery?

1. Complete resection of the tumor was performed
2. Pathological types of their tumors were cribriform pattern. (4)
3. No perineural invasion is able to be estimated because of not only the histopathology but also no ocular pain except for case 1.
4. The 2 patients were exempted from exenteration of the orbit because of their age.

## Conclusion

1. Most of ACC are usually very invasive and the prognosis is not good. The reasons of good clinical results of these 3 patients are extremely important to find necessary minimum treatments for each patient suffering from ACC. Exenteration of the orbit and extensive bone resection combined with radiation therapy do not warrant good outcome of patients enduring these mutilating surgeries.
2. We need to develop a custom-made therapy of adenoid cystic carcinomas to evade mutilating surgeries by finding important molecular genetic factors deterring prognosis.

## References

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