Suprasellar Malignant Teratoma with 11-year Survival

—Case Report—

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Abstract

Long-term survival of malignant teratoma is rare. A 16-year-old female with a suprasellar malignant teratoma survived for 11 years before succumbing to embryonal carcinoma. A suprasellar immature teratoma with increased serum alpha-fetoprotein level was verified histologically. Radiation therapy and combined chemotherapy were given after partial removal. Eleven years later, the primary lesion recurred as embryonal carcinoma.

Key words: embryonal carcinoma, malignant teratoma, immature teratoma, long-term survival

Introduction

Primary intracranial malignant teratomas such as embryonal carcinoma, yolk sac tumor, and choriocarcinoma are usually fatal, with a mean survival time of approximately 2 years. We report a case of suprasellar malignant teratoma with an unusually long survival of 11 years before recurrence caused death.

Case Report

A 16-year-old female was admitted to our institution in February, 1978, with visual impairment including double vision, headache, nausea, and vomiting. Polydipsia, polyuria, and amenorrhea had developed about 2 years prior to admission.

Physical examination disclosed normal development for her age. Neurological examination found bitemporal hemianopsia, right abducens nerve paresis, decreased visual acuity (right 0.4, left 0.07), and anisocoria (right > left). Precontrast computed tomographic (CT) scans showed a semicircular high-density area in the suprasellar region (Fig. 1 upper). Postcontrast CT scans showed an enhanced, 1.5 × 1.5 cm round mass, with low-density spots, extending to the third ventricle (Fig. 1 lower). Obstructive hydrocephalus was present. Serum alpha-fetoprotein

(AFP) and carcinoembryonic antigen levels were 320 and 2.5 ng/ml, respectively.

The suprasellar tumor was partially removed through a pterional approach *via* a right frontal craniotomy on March 16, 1978. Histological ex-

Fig. 1 Pre- (upper) and postcontrast (lower) CT scans on admission, showing 1.5 × 1.5 cm suprasellar lesion.

amination of the tumor specimen showed malignant teratoma, including cartilage, glandular structures, and dense hyalinized connective tissues (Fig. 2). Immunohistochemical staining for AFP and human chorionic gonadotropin (HCG) were both negative.

A ventriculoperitoneal shunt was installed on the 9th postoperative day. 85 mg adriamycin (10 mg/day $\times$ 4 and 15 mg/day $\times$ 3) was administered alternately from bilateral carotid arteries for 2 months. Two months later, 34.2 Gy irradiation was given in another hospital and followed by combined chemotherapy with nimustine hydrochloride (ACNU) and vincristine. Since CT scans demonstrated no effects from therapy on the lesion, combined chemotherapy with ACNU, procarbazine, and vincristine was given. Thereafter, she was well for 7 years without new neurological deficits or increased serum AFP level. Steroid treatment and correction of hypopituitarism were continued as an out patient.

Fig. 2 Photomicrograph of the surgical specimen, demonstrating malignant immature teratoma. HE stain, $\times$ 60.

Fig. 3 Follow-up postcontrast CT scans, showing an enlarged, $3.0 \times 2.5 \times 3.0$ cm mass extending to the third ventricle.

Fig. 4 *upper*: Photograph of the autopsy brain, showing a large mass development in the suprasellar region. *lower*: Photomicrograph of the autopsy specimen, verifying embryonal carcinoma. HE stain, $\times$ 120.

She was readmitted in March, 1985, after follow-up CT scans revealed that the primary lesion had enlarged to $3.0 \times 2.5 \times 3.0$ cm (Fig. 3). Neurological examination disclosed decreased visual acuity on the right (light perception), but the other deficits remained unchanged. Serum AFP and HCG levels were within the normal ranges.

On April 5, 1985, the suprasellar tumor was partially removed through an interhemispheric approach via a bifrontal craniotomy. Histological examination showed malignant teratoma, with similar features to the first surgical sample. Immunohistochemical staining for AFP and HCG were negative.

The postoperative course was uneventful. No radiation therapy or chemotherapy was given. Medical treatment for hypopituitarism was continued with the same regimen. Her Karnofsky performance status remained at 90%.

She was readmitted on February, 1989, with sudden onset of right hemiparesis. The serum AFP level had increased to 994 ng/ml. CT scans revealed a huge suprasellar mass extending to the midbrain. Combined chemotherapy with cisplatin (30 mg/day

for 5 consecutive days) and etoposide (90 mg/day for 5 consecutive days) was started immediately. After three courses of those chemotherapy within 6 months, her general condition improved. CT scans showed that the lesion size had regressed. However, 2 weeks later her consciousness level fell with speech and memory disturbances and hemiplegia. She died on October 9, 1989.

Autopsy examination showed the suprasellar mass developing toward the skull base with osteolytic changes in the sellar region, and extending to the bilateral thalami, left lenticular nucleus, and mid-brain (Fig. 4 upper). No evidence of extraneural metastasis was seen. Histological examination demonstrated the features of embryonal carcinoma, including proliferated atypical tumor cells exhibiting epithelial arrangements (Fig. 4 lower).

Discussion

128 cases of malignant teratoma, including embryonal carcinoma, yolk sac tumor, and/or choriocarcinoma, have been described in detail and verified histologically,^{2-4,7,9,11-13,16-19,21-23,26,28,29,31,32,35,36)} 76 in various reviews.^{5,8,14,24,27,33,34)} Ten cases including ours survived for over 3 years.^{1,6,9,12,15,16,24,28,31)} Our case was the longest survivor among the malignant teratoma cases.

Table 1 summarizes clinical data for the 10 long-

term survivors. The histology was documented in eight cases: three teratoma and embryonal carcinomas, two yolk sac tumors, one embryonal carcinoma and choriocarcinoma, one choriocarcinoma and teratoma, and one embryonal carcinoma and yolk sac tumor. These results suggest that embryonal carcinoma and yolk sac tumor have a much less satisfactory prognosis than choriocarcinoma. The primary lesion was pineal in seven cases, suprasellar in two, and basal ganglia in one. Most cases received subtotal tumor removal. Such aggressive excision may consequently improve the prognosis.

Six long-term survivors received multimodality therapy consisting of surgery, radiation therapy, and chemotherapy. The effectiveness of radiation therapy except in high doses is still undetermined, because many cases of malignant teratoma excluded from Table 1 died within 2 years after radiation therapy. High-dose irradiation carries a risk of various adverse effects including intellectual impairment.²⁷⁾

More recently, combined chemotherapy using cisplatin and other drugs such as bleomycin, vinblastine, and etoposide has been effective against some malignant teratomas.^{10,12,16-18,20,21,25,27,30)} Our case received combined chemotherapy at the initial admission which might have achieved the 11-year survival. Accordingly, high-dose combined chemotherapy may produce good results, although the

Table 1 Intracranial malignant teratoma cases with more than 3-year survival

Author (Year)	Age	Sex	Histological diagnosis	Lesion site	Tumor removal	Radiation therapy	Chemotherapy	Outcome
Bestle (1968) ⁶⁾	13 yrs	F	yolk sac tumor	suprasellar	yes	2 courses	—	died (5 yrs 6 mos)
Kawakami <i>et al.</i> (1980) ¹⁵⁾	11 yrs	M	choriocarcinoma + teratoma	pineal	subtotal	40 Gy	methotrexate, actinomycin D	alive (4 yrs)
Murovic <i>et al.</i> (1981) ²⁴⁾	20 mos	F	yolk sac tumor	pineal	subtotal	46 Gy local, 26 Gy CS	—	alive (3 yrs 6 mos)
Chang <i>et al.</i> (1981) ⁹⁾	18 yrs	M	teratoma + embryonal carcinoma	pineal	subtotal	58 Gy	—	died (4 yrs 8 mos)
Packer <i>et al.</i> (1984) ²⁸⁾	16 yrs	M	embryonal carcinoma + yolk sac tumor	pineal	not total	55 Gy local, 40 Gy CS	vincristine, actinomycin, cyclophosphamide	alive (7 yrs)
Kida <i>et al.</i> (1986) ¹⁶⁾	24 yrs	M	malignant teratoma	pineal	subtotal	100 Gy	—	died (4 yrs 1 mo)
	9 yrs	M	malignant teratoma	basal ganglia	total	—	cisplatin, bleomycin, vinblastine	alive (4 yrs 11 mos)
Graziano <i>et al.</i> (1987) ¹²⁾	15 yrs	M	embryonal carcinoma + choriocarcinoma	pineal	subtotal	56 Gy	cisplatin, bleomycin, vinblastine	alive (3 yrs 1 mo)
Sakai <i>et al.</i> (1988) ³¹⁾	14 yrs	M	teratoma + embryonal carcinoma	pineal	partial	52.3 Gy	adriamycin	died (4 yrs)
Present case	16 yrs	F	teratoma + embryonal carcinoma	suprasellar	partial	34.2 Gy	multiple*	died (11 yrs 7 mos)

*Adriamycin, ACNU + vincristine, ACNU + procarbazine + vincristine, and cisplatin + etoposide were given. CS: craniospinal.

most efficacious treatment of malignant teratoma still remains to be determined.

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