Malignant Trigeminal Schwannoma Associated with Xeroderma Pigmentosum

—Case Report—

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Abstract

A 46-year-old male with xeroderma pigmentosum developed an intracranial malignant schwannoma originating from the second branch of the left trigeminal nerve. The tumor was subtotally removed and postoperative radiation therapy given, but the tumor recurred twice over 3 years, and extended to the third branch of the left trigeminal nerve and the ipsilateral facial nerve. Radical surgery and radiation therapy finally achieved a cure. This is the first case of malignant trigeminal schwannoma with xeroderma pigmentosum, although various other internal neoplasms including central nervous system tumors have been reported in xeroderma pigmentosum patients. Radical surgery and radiation therapy are effective for treating intracranial malignant schwannoma. Radiation therapy is considered safe for xeroderma pigmentosum patients.

Key words: malignant schwannoma, xeroderma pigmentosum, radiation therapy, radical surgery

Introduction

Xeroderma pigmentosum is an autosomal recessive disease characterized by hypersensitivity to sunlight, freckling, premature skin aging, and multiple skin neoplasias. Approximately 80-90% of all sufferers manifest dysfunction of deoxyribonucleic acid (DNA) repair after damage by ultraviolet radiation. Skin cancer develops in 45% of xeroderma pigmentosum patients, and 1.4-2.5% develop various associated internal neoplasms including central nervous system tumors. Five cases of brain tumors with xeroderma pigmentosum have been reported: astrocytoma, glioblastoma, medulloblastoma, and meningeal sarcoma (2 cases).

We report a case of malignant trigeminal schwannoma associated with xeroderma pigmentosum, which recurred twice and was finally cured by radical surgery and radiation therapy.

Case Report

A 46-year-old male presented with left facial pain and swelling of the left cheek in September, 1987. Biopsy of the tumor projecting into the oral cavity demonstrated malignant schwannoma. He had previously undergone removal of the lentigo maligna below the right palpebra under a diagnosis of xeroderma pigmentosum at a different institution. He was admitted to our neurosurgery service in March, 1988 because of left facial pain corresponding to the distribution of the first and second divisions of the left trigeminal nerve. There were no clinical signs of von Recklinghausen's disease. Magnetic resonance (MR) imaging showed that a mass lesion was located in the left lateral cavernous region (Fig. 1) and the second branch of the left trigeminal nerve was abnormally enlarged. The first operation was performed by the left subtemporal approach. The tumor was located mainly at the second branch of the left trigeminal nerve, and extended into the foramen rotundum anteriorly. The tumor was subtotally removed, leaving the posterior part of the tumor at the third branch of the trigeminal nerve.
running toward the foramen ovale. The tumor arising from the extracranial portion of the left trigeminal nerve was treated surgically by the otolaryngology service. External irradiation (50 Gy) was administered to the intra- and extra-cranial tumors.

He remained well until January, 1989 when he developed visual and gait disturbances. MR imaging showed a well-demarcated mass lesion in the left posterior fossa (Fig. 2). A second operation using the left subtemporal approach demonstrated tumors involving the left Gasserian ganglion and the proximal root of the left trigeminal nerve. Only partial removal of the tumor was possible because of tight adhesion to the brain stem. A course of radiation therapy (50 Gy) was given postoperatively.

There were no signs or symptoms of recurrence until soft swellings developed in the left preauricular area (1990) and the mental foramen (March, 1991). The tumor had recurred, extending to the third branch of the left trigeminal nerve and the ipsilateral facial nerve (Fig. 3). He was treated surgically in collaboration with dental and oral surgeons in April, 1991. The tumor arising from the left mental and the mandibular nerves was removed after left mandibulectomy. The third division of the left trigeminal nerve, which was enlarged by the neoplasms, was cut at the level of the foramen ovale. The left facial nerve and its small branches were involved and so were cut at the level of the styloid foramen. Histological examination of the specimens of tumor
tissues from the enlarged trigeminal and the facial nerves showed fascicular arrangements consisting of atypical spindle-shaped cells with hyperchromatic and pleomorphic nuclei including a few mitotic figures (Fig. 4). Immunohistochemical staining showed the tumor cells were positive for anti-S-100 protein antibody. The final diagnosis for all specimens was malignant schwannoma. Postoperative radiation therapy (45 Gy) to the left facial region was given. His postoperative progress was satisfactory except for left facial nerve paralysis and hypesthesia. There has been no evidence of recurrence as of April, 1994.

### Discussion

Our patient is the first reported case of intracranial malignant schwannoma associated with xeroderma pigmentosum.

Intracranial malignant schwannomas arising from the trigeminal nerve are rare, with only 15 previous cases reported (Table 1). Recurrence is frequent and spread easily occurs proximally or distally along the nerve sheath. Therefore, radical surgery and radiation therapy are necessary to prevent recurrence. Our patient has developed no recurrence in the 3 years since radical surgery during the third hospitalization. Incomplete removal in the previous operations did not prevent tumor recurrence and extension. Radical surgery is mandatory for the treatment of intracranial malignant schwannoma even in xeroderma pigmentosum patients.

In xeroderma pigmentosum patients, x-rays and many chemotherapeutic agents which may damage DNA are too toxic and mutagenic for use as anticancer therapy because of dysfunction in DNA repair mechanisms. However, Giannelli et al. found clinically and experimentally that radiation therapy was safe in a xeroderma pigmentosum patient with medulloblastoma. Our patient had a satisfactory clinical course after radiation therapy. Radiation therapy is therefore an appropriate treatment for brain tumors in xeroderma pigmentosum patients.

The prognosis for xeroderma pigmentosum patients with brain tumors is not known in detail. The mortality among such patients with other cancers seems to be higher than that of the general population. However, radical surgery and radiation therapy for the treatment of tumors can provide a good quality of life.

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**Table 1 Initial treatment and outcome in patients with malignant schwannomas**

<table>
<thead>
<tr>
<th>Author (Year)</th>
<th>Age/Sex</th>
<th>Tumor removal</th>
<th>Radiation therapy</th>
<th>Tumor origin → recurrence and extension</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cuneo and Rand (1952)</td>
<td>37/M</td>
<td>subtotal</td>
<td>none</td>
<td>Lt V3 → Lt V2</td>
<td>alive 3 mos</td>
</tr>
<tr>
<td>Dinakar et al. (1971)</td>
<td>30/M</td>
<td>partial</td>
<td>performed</td>
<td>Lt GG</td>
<td>alive 2 mos</td>
</tr>
<tr>
<td>Hedeman et al. (1978)</td>
<td>53/M</td>
<td>subtotal</td>
<td>51.75 Gy</td>
<td>rt GG</td>
<td>alive 18 mos</td>
</tr>
<tr>
<td>Karmody (1979)</td>
<td>70/M*</td>
<td>partial</td>
<td>60 Gy</td>
<td>rt V2–3 → rt V1–3</td>
<td>died 3 yrs of pulmonary embolism</td>
</tr>
<tr>
<td>Liwnicz (1979)</td>
<td>49/M</td>
<td>partial</td>
<td>66 Gy</td>
<td>rt V3 → Lt V1–3, Lt VII, pons</td>
<td>died 4 yrs of pneumonia</td>
</tr>
<tr>
<td>Levy et al. (1983)</td>
<td>61/M</td>
<td>partial</td>
<td>60 Gy</td>
<td>rt V1–3, rt root</td>
<td>died 10 mos</td>
</tr>
<tr>
<td>Robertson et al. (1983)</td>
<td>42/M</td>
<td>performed</td>
<td>none</td>
<td>rt GG</td>
<td>died 3 days of sepsis</td>
</tr>
<tr>
<td></td>
<td>45/M</td>
<td>performed</td>
<td>none</td>
<td>lt inferior dental nerve → local</td>
<td>alive</td>
</tr>
<tr>
<td></td>
<td>24/M</td>
<td>partial</td>
<td>none</td>
<td>lt mental nerve → Lt GG</td>
<td>alive</td>
</tr>
<tr>
<td></td>
<td>46/M</td>
<td>partial</td>
<td>performed</td>
<td>rt V2 → rt cervical plexus, rt vagus nerve, rt superior laryngeal nerve</td>
<td>died 9 yrs</td>
</tr>
<tr>
<td></td>
<td>62/M</td>
<td>total</td>
<td>none</td>
<td>rt parotid gland → rt temporal region</td>
<td>alive</td>
</tr>
<tr>
<td>Maroun et al. (1986)</td>
<td>49/F</td>
<td>partial</td>
<td>60 Gy</td>
<td>Lt V2</td>
<td>alive</td>
</tr>
<tr>
<td>Horie et al. (1990)</td>
<td>18/M</td>
<td>partial</td>
<td>none</td>
<td>rt root, rt GG</td>
<td>died 2 mos</td>
</tr>
<tr>
<td>Present case</td>
<td>46/M</td>
<td>subtotal</td>
<td>50 Gy</td>
<td>Lt V2 → Lt root, Lt V3, Lt VII</td>
<td>died 3 yrs</td>
</tr>
</tbody>
</table>

References


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