Case report

Long-term survival after repeated resections of Askin tumor recurrences

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Abstract

The case of a 16-year-old boy is reported who underwent surgery for the excision of an Askin tumor and has subsequently undergone six excisions of local Askin tumor recurrences, with follow-up postoperative chemo- and radiotherapy over a 7-year period. The patient continues to survive. As far as can be determined, this patient appears to be the first reported case of long-term survival after repeated resections of local Askin tumor recurrences. It thus may be that postoperative chemo- and radiotherapy after repeated excisions of these local Askin tumor recurrences plays a role in prolonging survival. © 1998 Elsevier Science B.V. All rights reserved.

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1. Introduction

In 1979, Askin et al. first identified a primitive neuroectodermal tumor (PNET) of the thoracopulmonary region in children and young adults as a specific pathological entity [1] and since that time, such a tumor is often referred to as being an Askin tumor.

Clinically, a local recurrence in an Askin tumor has frequently occurred, and the survival rate is poor [1]. However, little exists in the literature about the effectiveness of a surgical resection of a locally recurred Askin tumor when combined with chemo- and radiotherapies. Herein, we report an Askin tumor case of long survival after being given this combined treatment, said patient having thus far undergone five resections for re-occurring Askin tumors.

2. Case report

In August 1990, a 16-year-old boy was admitted to hospital because of an expanding mass without pain in the left posterior chest wall. Although his chest radiography appeared normal, chest CT scans revealed a solid mass sited in the left posterior chest wall. On August 27, 1990, the tumor was resected with the 10th and 11th ribs, and measured 10 × 9 × 9 cm.

In November 1991, the patient was again admitted to hospital because of another mass with pain that had developed over a 3-month period in his left posterior wall.

A chest radiograph showed smooth bordered mass occupying the left thoracic wall (Fig. 1). Chest CT scans revealed two masses: one that involved the ribs and one that was subcutaneously sited on the latissimus dorsi muscle. The patient underwent a wide local tumoral resection: one tumor, which was excised with the
9th and 10th ribs, measured $10 \times 9 \times 8$ cm, and the other tumor, $4 \times 5 \times 5$ cm. Postoperatively, a chemotherapeutic regimen of three cycles of cyclophosphamide, adriamycin, vincristine and actinomycin-D was initiated.

In September 1993, the patient again presented two subcutaneous nodules along the previous surgical wound. Chest CT scans also revealed several nodules in the musculature under the wound. The patient underwent a $25 \times 10$ cm-wide resection of the skin and latissimus dorsi muscle along the site of the previous surgery, after which he was given the same cycles of the postoperative chemotherapy as well as radiotherapy.

In March, 1994, in spite of no complaint, follow-up chest CT scans again revealed the local recurrence of a solitary mass in his left pleural space, and the patient underwent yet another resection. The excised mass, which had a thin neck that showed that it had originated in the pericardium, measured $55 \times 35 \times 25$ mm. Postoperatively, the patient received the same cycles of chemotherapy.

In November, 1995, he was asymptomatic, but chest CT scans again detected a solitary pleural mass in his left intrathoracic cavity. This tumor, which was sited in the posterior parietal pleura adjust to the neck of the 10th rib, was resected. This time the postoperative chemotherapy consisted of five cycles of pirarubicin and ifosfamide.

At a follow-up exam in December 1996, the patient complained of swelling and tenderness of the chest along the site of the previous surgery, and a subcutaneous tumor with a rib involvement was found under the wound. So again this tumor, which measured $10 \times 8 \times 5$ cm, was resected with the 8th, 9th, and 10th ribs.

In February 1997, the patient complained of a severe back pain and a chest CT revealed a solitary intrathoracic tumor, extending from the neck of the 12th rib to the body of the 12th vertebra. This time, however, the patient refuse further surgery and chemotherapy. Thus, using a heavy-ion accelerator in Chiba, Japan, the patient underwent heavy-ion therapy; the tumor was reduced to half its size and his chest pain disappeared.

In conclusion, since the initial primary lesion was excised 7 years earlier, the patient has undergone five tumoral resections for local recurrences and has received chemo- and radiotherapy after each bout of surgery. At 7 years postoperatively, he remains alive with the disease.

3. The histological and immunological findings

Specimens of the primary tumor and the subsequent locally recurred lesions taken at the time of each operation were histologically and immunologically evaluated. Histologically, the tumor cells of the primary consisted of small, round or oval-shaped nuclei and abortive rosette-like figures (Fig. 2).

The immunohistochemical findings revealed that groups of cells were negative for S-100 protein, neuron-specific enolase (NSE), glial fibrillary acidic protein (GFAP), epithelial membrane antigen (EMA), keratin, and KL-1. In contrast, the tumor cells were fairly positive for vimentin, and the tumor cell membranes were weakly positive for HBA-71 (MIC 2) (Fig. 3).
Excised tumor. There is the predilection for a PNET to reappear in approximately the same area as the primary neoplasm [1].

Survival for patients with a malignant, small, round cell tumor (MSRCT), which includes an Askin tumor or Ewing's sarcoma, is poor, the 2-year survival rate is reported to be 38% and the 6-year rate is 14%; also, survival after a local MSRCT recurrence or a metastasis from a primary MSRCT has reported to be 11 months [5]. Further, Askin has reported that no PNET patients with a local recurrence have survived longer than 4 years [1]. As far as we are able to determine, our patient is the first reported case of a long-term survival after repeated resections of locally recurring lesions.

As we included postoperative chemothera- and radiotherapy after each of these resections, it may be that this combined method of treatment has helped to extend our patient’s life.

References


4. Discussion

A PNET is not easy to diagnose, since it consists of small, round cells that bear a resemblance to a neuroblastoma, Ewing’s sarcoma, and a rhabdomyosarcoma [2]. HBA-71, which recognizes the surface protein p30/32mic2encoded by the pseudoautosomal MIC2 gene reacts in PNET tumors and Ewing’s sarcoma [3]. The distinction between Ewing’s sarcoma and a PNET can become blurred [4]. However, in our patient, most of the clinical and morphological findings support a PNET diagnosis. This Askin tumor locally recurred six times in 7 years and whenever the tumor recurred, the metastatic status was investigated. Bone scan, as well as brain and liver CT, always showed no abnormalities, and no signs of a distant metastasis were noted. Though we undertook no partial biopsy, but relative wide excision of local Askin tumor recurrences, the tumor had repeatedly risen at sites adjacent to the

Fig. 3. Tumor cell immunocytochemical HBA-71 reaction. Tumor cell membranes are weakly positive for HBA-71( × 200).

Based on above and the clinical findings, the diagnosis was a PNET.