Case Report

Long-term Survival of Askin Tumor for 10 Years with 2 Relapses

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An abnormal shadow was noted on a chest X-ray of a 32-year-old female in a medical check-up in March 1995, 3 months after she had given birth. Thoracic CT detected a tumor in contact with the left thoracic wall, and tumorectomy was performed in May 1995. The tumor was diagnosed as a primitive neuroectodermal tumor (PNET). After surgery, the thoracic wall to which the tumor adhered was treated with irradiation at 50 Gy. Chemotherapy was considered, but the patient did not wish to undergo this treatment. Lung metastasis occurred 5 years after the first surgery, and the left lower lobe of the lung was partially resected. Four years later, lobectomy of the left lower lobe of the lung was performed, due to further lung metastasis. The patient remains healthy as of April 2005. (Ann Thorac Cardiovasc Surg 2006; 12: 137–40)

Key words: Askin tumor, Ewing sarcoma, primitive neuroectodermal tumor, small round cell tumor

Introduction

Askin tumor is a rare malignant tumor that develops in the thoracic region in young people. The prognosis is generally poor. Here, we report a patient with Askin tumor who has survived for 10 years after two occurrences of lung metastasis; 5 and 9 years after initial surgery and postoperative radiotherapy. The tumor was diagnosed as a primitive neuroectodermal tumor (PNET), and this tumor type and Ewing sarcoma have been integrated into a single item (Ewing sarcoma/PNET) in the new 2002 WHO classification. However, different therapeutic methods are currently used for the two tumor types, and literature reports of treatment of these tumors are discussed below.

Case Report

The patient is a 32-year-old female who visited our hospital in March 1995 because of an abnormal shadow found in a chest x-ray photograph in a medical check-up 3 months after she had given birth. No clinical symptoms were observed, and the patient had no particular past medical events or familial medical history. Hematology and blood chemistry were within the normal ranges, and tumor markers (CEA, NSE, SCC, CA19-9 and HCG-β) were lower than the standard values. No metastasis to other organs was found. On needle biopsy, small round cells were noted histologically. Malignant lymphoma and neuroendocrine tumor were suspected, and a tumorectomy was performed in May 1995. The tumor was pediculated and disk-shaped with a clear boundary measuring 10×10 cm, and was attached to the thoracic wall through a 3×3 cm pedicle (Fig. 1). The pedicle was easily detached from the costal surface. Since no tumor invasion was evident macroscopically, no resection of the costa was performed, and only the periosteum at the tumor attachment site was dissected. Since differentiation into the nervous system was noted in several immunostaining tests and in electron microscopy on histopathological examination, the tumor was diagnosed as a PNET. Radiotherapy at 50 Gy was applied to the costal surface following surgical dissection of the periosteum.

During follow-up, a mass shadow in the left lung was

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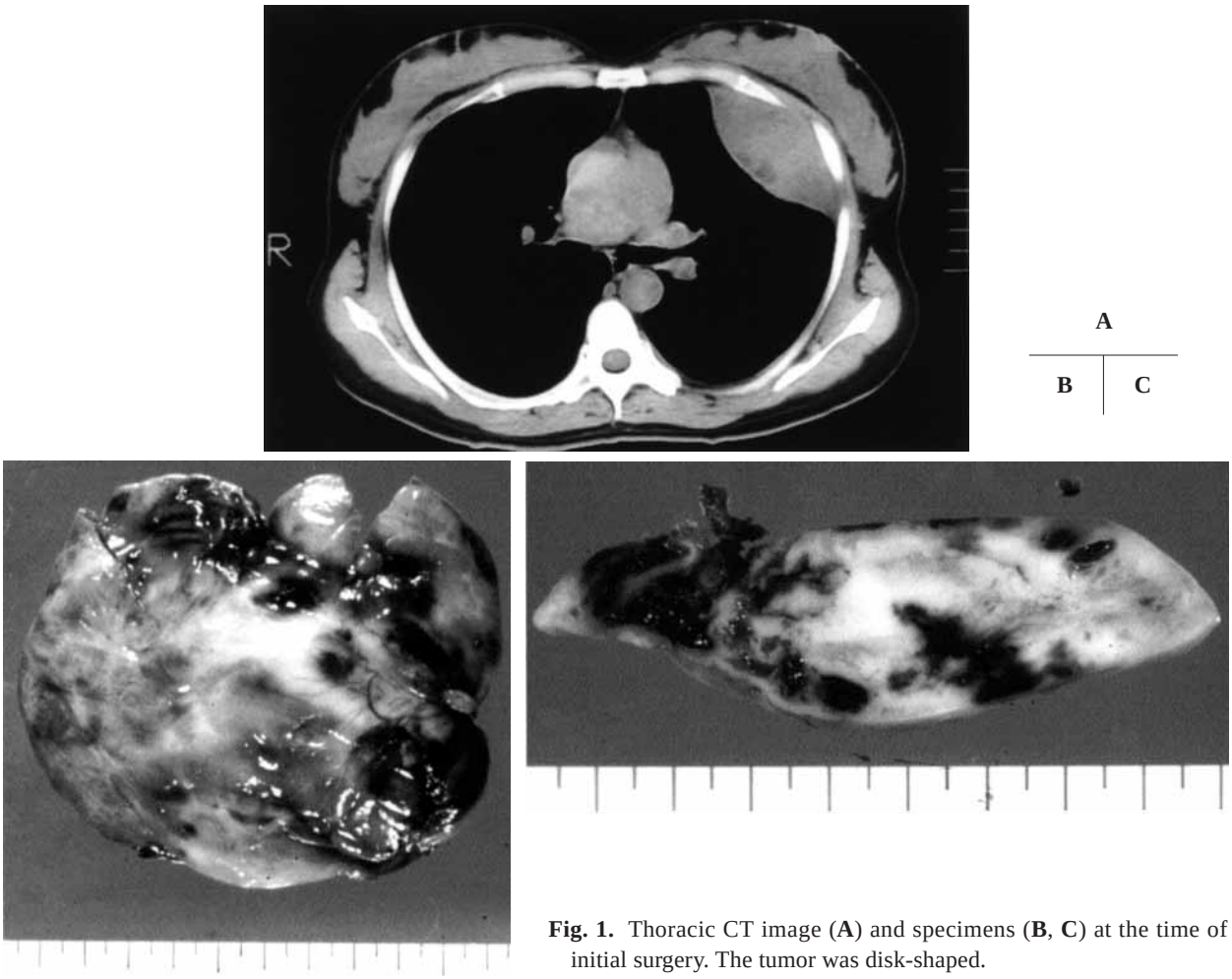

Fig. 1. Thoracic CT image (**A**) and specimens (**B**, **C**) at the time of initial surgery. The tumor was disk-shaped.

detected by thoracic CT in March 2000, and partial thoracoscopic resection of the left lung was performed (Fig. 2). Based on a histopathological examination, a diagnosis of left lung metastasis was made. In June 2004, a tumor shadow in the left lung was detected by thoracic CT, and lobectomy of the left lower lobe of the lung was performed (Fig. 3). A diagnosis of metastasis of the left lung was made based on a histopathological examination. The patient is still being followed as of April 2005.

Discussion

PNET is a nerve-derived small round cell tumor, and is a rare disease that develops in soft tissues in young individuals. Among cases of PNET, tumors of thoracopulmonary origin were first reported as "malignant small cell tumors of the thoracopulmonary region in childhood" by Askin et al. in 1979, and thus these tumors are referred to as Askin tumor.[1] PNET is included as a tumor of bone soft tissue in the WHO classification. PNET and Ewing sarcoma were individually described as separate items in the small round cell tumor group in the 2nd version of the WHO classification, published in 1993.[2] However, they have subsequently been integrated into a single item (Ewing sarcoma/PNET) in the new WHO classification, published in 2002,[3] in which the two tumor types are defined as identical tumors with different differentiation in the neuroectoderm, because a t(11;22)(q24;q12) translocation specific to both Ewing sarcoma and PNET has been found by chromosomal analysis, and the EWS-FLI-1 fused gene was identified at the cleavage site of this translocation,[4] suggesting that these tumors belong to the same group in histogenetics.

Ewing sarcoma is considered to be highly sensitive to chemotherapy and radiotherapy, and the prognosis is relatively good, while the prognosis of PNET and Askin tumor is considered to be poor. Chemotherapy is the first choice for treatment of Ewing sarcoma, and a subsequent

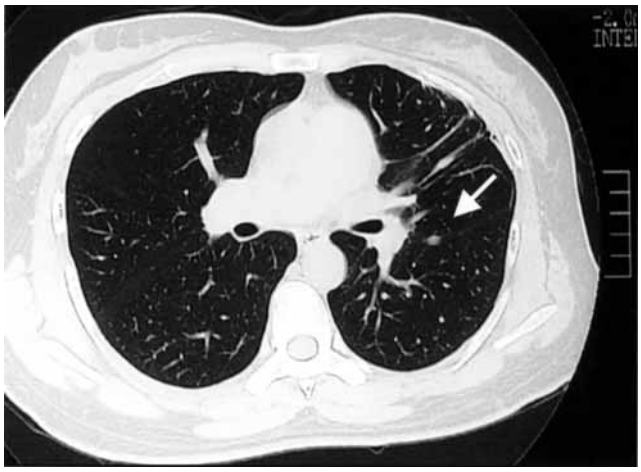

Fig. 2. Thoracic CT image at the time of recurrence. A mass shadow measuring 7×5 mm was observed in S5 of the left lung (arrow).

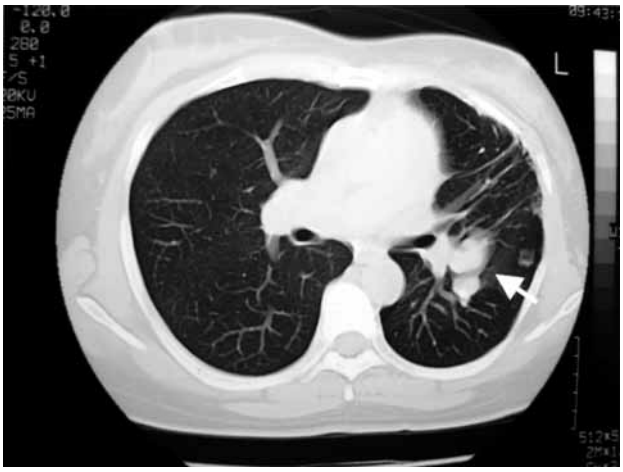

Fig. 3. Thoracic CT image at the time of second recurrence (arrow).

combination of surgery and radiation is the standard therapy.[5] In contrast, no standard therapy is available for Askin tumor, because only a few cases have been reported and the prognosis is poor. In past reports, extended surgery, including resection of the thoracic wall, has been performed as the first choice, followed by postoperative chemotherapy and radiotherapy in many cases.[6] However, it has recently been reported that preoperative chemotherapy for Askin tumor, with reference to the treatment of Ewing sarcoma, can result in a good outcome.[7,8] The grouping of the two tumors into the same WHO classification also suggests that preoperative chemotherapy may become the main treatment approach for Askin tumor.

Since a definite diagnosis could not be made before surgery in the patient in the current case, diagnostic tumorectomy was performed. Additional resection of the costa was considered after diagnosis of PNET, but detachment of the tumor from the costal surface was straightforward, as described above, and the surgical margin was pathologically negative. In addition, bone scintigraphy suggested no uptake in the region. Therefore, no additional resection was performed, but post-operative irradiation of the costal surface was conducted at 50 Gy. Postoperative chemotherapy was also considered, but the patient did not wish to undergo this treatment. In past case reports, chemotherapy for Askin tumor has almost always been performed and, according to Veronesi et al.,[8] preoperative chemotherapy is also effective because shrinkage of the lesion can be achieved, allowing complete surgical resection.

Since Ewing sarcoma develops in bone in many cases and the symptoms are easily detected, the tumor is likely to be discovered early. In contrast, PNET develops in soft tissues, and the symptoms are largely detectable only after enlargement of the tumor; thus, the tumor may be discovered only at an advanced stage, which is one of the reason for the poor prognosis of PNET. Regarding the prognosis of Askin tumor, Askin reported that 14 of 18 patients with known prognosis died 4-44 months after diagnosis, and the mean survival period was 8 months.[1] Contesso et al. reported that the 2-year survival rate was 38%, and the 6-year survival rate was 14%.[9] For the patient in the current case, surgery and postoperative radiotherapy has allowed long-term survival for 10 years after the first surgery, despite no chemotherapy being performed. The reason for the long-term survival may be discovery at a medical check-up before development of symptoms, which allowed complete surgical resection at a non-advanced stage. Also, relapse was solitary, and was found and treated surgically while the tumor was small. However, considering that lung metastasis occurred 5 and 9 years after the first surgery, chemotherapy may be necessary, even for non-advanced cases, and since the tumor relapsed after 9 years, careful long-term follow-up is also necessary.

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