Extradural Granulocytic Sarcoma of the Thoracic Spine in Acute Myelogenous Leukemia with 8 ; 21 Chromosome Translocation

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AIZAWA, T., KOKUBUN, S., HATORI, M., KASHIMOTO, O. and ICHINOHASAMA, R. *Extradural Granulocytic Sarcoma of the Thoracic Spine in Acute Myelogenous Leukemia with 8 ; 21 Chromosome Translocation.* Tohoku J. Exp. Med., 1996, **178** (4), 431-436 — We report a case of a 19-year-old male with an extradural granulocytic sarcoma at T5/6 to T7 demonstrated by MRI. He recovered from paraplegia that had progressed rapidly after excision of the tumor as an emergency operation. Histopathological studies disclosed granulocytic sarcoma. Both peripheral blood and bone marrow examinations revealed acute myelogenous leukemia. The karyotype of the bone marrow showed 45, X, -Y, t (8 ; 21) (q22 ; q22). ——— granulocytic sarcoma; acute myelogenous leukemia; epidural tumor; chromosome abnormality; 8 ; 21 translocation

Granulocytic sarcoma is a localized tumor composed of myeloid precursor cells (Chan et al. 1986), and is associated with leukemia or myeloid proliferative disorders in most cases (Petursson and Boggs 1981). Many granulocytic sarcomas have a translocation between chromosomes 8 and 21 (Swirsky et al. 1984; Abe et al. 1986; Tallman et al. 1993). But among those with epidural manifestations, only 5 cases with this translocation have been reported (Abe et al. 1986; Wodzinski et al. 1988; Watanabe et al. 1990). We surgically treated a patient with paraplegia due to an epidural granulocytic sarcoma having the same chromosomal abnormality.

CASE REPORT

A 19-year-old male felt a dull pain in the lower back in December, 1993, which subsided gradually without any treatment. On February 14, 1994, he was

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referred to our hospital because of difficulty in walking due to muscle weakness of the lower limbs and progressive urinary disturbance.

Physical examination revealed incomplete spastic paraplegia with marked ankle clonus and weakness of Grade fair to good in the lower limbs. Light touch sensation and pinprick were decreased on the 9th thoracic and lower dermatomes. Knee and ankle jerks were accelerated and the Babinski sign was bilaterally observed. A plain A-P radiograph of the thoracic spine depicted a large paraspinal mass without apparent vertebral erosion (Fig. 1). Magnetic resonance imaging (MRI) demonstrated an epidural tumor compressing the spinal cord at T5/6 to T7. The tumor had high signal intensities on both T1- and T2-weighted images, and was slightly enhanced with gadolinium-diethylenetriamine-pentaacetic acid (Gd-DTPA) (Fig. 2). Axial view of MRI did not show distinct connection between the paraspinal and the epidural tumor.

The white blood cell count was $7,300/\mu\text{l}$ with 17% segmental neutrophils, 1% bands, 15% myelocytes, 1% promyelocytes, 32% myeloblasts, 3% monocytes and 30% lymphocytes. Auer rods were found in the myeloblasts. The red blood cell count was $561,000/\mu\text{l}$ and the platelet count was $202,000/\text{mm}^3$. The hemoglobin was 16.3 g/100 ml and the hematocrit 48.3%. The possible diagnosis from these haematologic findings was acute myelogenous leukemia.

His paraplegia progressed rapidly and the leg muscles weakened to Grade trace or poor on February 15. An emergent operation for decompression of the

Fig. 1. A-P radiograph of the thoracic spine showing a large paraspinal mass on the left side without bone erosion (arrows).

Fig. 2. (A) T1-weighted, (B) T2-weighted, (C) Gd-enhanced T1-weighted MR images. The tumor was detected in the epidural space at T5-7. The tumor shows high signal intensities both on T1- and T2-weighted images. Slight enhancement is displayed (arrows).

spinal cord was performed. Through laminectomy from T5 to T7, a soft and grayish tumor compressing the spinal cord was disclosed. The tumor was removed en bloc, which measured $3 \times 2 \times 1.5$ cm.

Histopathological examination revealed that the tumor was composed of medium- to large-sized cells having round or indented nuclei with fine chromatin (Fig. 3). Electron micrographs demonstrated that the tumor cells had round to

Fig. 3. Photomicrograph showing leukemic cells of tumor resected which have round or indented nuclei with fine chromatin (H-E stain, $\times 400$).

Fig. 4. Electronmicrograph showing various dense granules in leukemic cells (arrows) ($\times 11,000$).

oval euchromatic nuclei with large nucleoli. Most of them possessed diverse dense granules (Fig. 4). The tumor was diagnosed as granulocytic sarcoma.

Bone marrow aspiration was carried out after surgery. It revealed a nucleated cell count 19.4×10^4 cells/mm³, 41.6% myeloblasts, 5.6% promyelocytes, 9.6%

Fig. 5. Karyotype of the bone marrow cell. The karyotype is 45, X, -Y, t(8;21)(q22;q22).

myelocytes, 2% eosinocytes, 4.8% stab neutrophils, 9.2% segmental neutrophils, 1.2% monocytes 18% lymphocytes, and 0.4% plasmocytes. Forty-two percent of the leukemic cells of the bone marrow showed positive staining with myeloperoxidase and chloracetate esterase. He was classified as M2 acute myelogenous leukemia according to the French-American-British (FAB) classification based on this bone marrow and peripheral blood findings. The karyotype of the bone marrow cells showed a translocation between chromosomes 8 and 21, 45, X, -Y, t(8; 21) (q22; q22) in 15 cells out of 20 cells examined (Fig. 5).

Chemotherapy was started 2 days after surgery and led to haematologic remission. Neurologic recovery was achieved 6 months after surgery.

DISCUSSION

Granulocytic sarcoma is a solid tumor composed of immature cells of the myeloid series (Chan et al. 1986). It occurs in 3% of patients with myelogenous leukemia (Krause 1979). Fifty to 60% of granulocytic sarcoma cases are composed of acute myelogenous leukemia and 10 to 20% of acute lymphocytic leukemia (Kurita 1982; Petursson and Boggs 1981). It can occur anywhere in the body, but epidural presentation is rare (Chan et al. 1986; Ripp et al. 1989). To our knowledge, only 23 patients, nineteen males and four females, with an epidural granulocytic sarcoma have been reported (Wilhyde et al. 1963; Mason et al. 1973; Neiman et al. 1981; Petursson and Boggs 1981; Kellie and Waters 1984; Abe et al. 1986; Chan et al. 1986; Wodzinski et al. 1988; Ripp et al. 1989; Watanabe et al. 1990).

Epidural granulocytic sarcoma should be differentiated from hematoma, malignant lymphoma, lipoma and liposarcoma when no destructive changes in the spine are found on radiographs and paralysis progresses rapidly. Immature myeloid cells in peripheral blood are diagnostic. In the present case, myeloblasts and Auer rods were detected, facilitating preoperative diagnosis.

Recently, a translocation between chromosomes 8 and 21 has been reported specifically to be associated with granulocytic sarcoma (Swirsky et al. 1984; Abe et al. 1986; Tallman et al. 1993). Regarding epidural granulocytic sarcoma, only 5 patients with this translocation have been reported (Abe et al. 1986; Wodzinski et al. 1988; Watanabe et al. 1990). The bone marrow cells of this case also had this abnormality.

Surgical decompression of the spinal cord is the treatment of choice for an epidural granulocytic sarcoma, especially before establishment of complete paralysis (Wodzinski et al. 1988). Recent chemotherapy is often effective for acute myelogenous leukemia and frequently leads to haematologic remission. It could also be indicated for epidural granulocytic sarcomas. Paralysis, however, may persist if chemotherapy fails to reduce the tumor mass quickly. Radiation is also one of the treatments. But it may cause delayed myelopathy and may be ineffective for quickly diminishing the tumor size (Nagao et al. 1975; Chan et al.

1986; Wodzinski et al. 1988). In the present case, emergency decompression was performed soon after paraplegia had occurred, and then chemotherapy followed, resulting in neurologic recovery.

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