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A thoracic-epidural granulocytic sarcoma case that was diagnosed preceding the onset of and that recurred co-incidental to acute promyelocytic leukemia, which developed after surgical treatment.*

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Abstract

Granulocytic sarcoma or chloroma is a tumor seen in myelocytic leukemia. Spinal epidural onset is rare and is generally seen before or together with the onset of myelocytic leukemia. An epidural mass located at the 2nd-5th thoracic levels in an 18-year-old male patient was pathologically diagnosed as granulocytic sarcoma. Radiotherapy was performed after surgical intervention. Ten months later, he was re-admitted with abdominal pain. At this time, an epidural mass at the 6th-9th thoracic levels was detected on magnetic resonance imaging, and acute promyelocytic leukemia was diagnosed. After systemic chemotherapy, partial remission was achieved. We aimed to present this rare case with its remarkable follow-up findings.

KEYWORDS: chloroma, acute promyelocytic leukemia, epidural mass, thoracicspine

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Case Report

A Thoracic-Epidural Granulocytic Sarcoma Case that was Diagnosed Preceding the Onset of and that Recurred Co-incidental to Acute Promyelocytic Leukemia, which Developed after Surgical Treatment

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Granulocytic sarcoma or chloroma is a tumor seen in myelocytic leukemia. Spinal epidural onset is rare and is generally seen before or together with the onset of myelocytic leukemia. An epidural mass located at the 2nd-5th thoracic levels in an 18-year-old male patient was pathologically diagnosed as granulocytic sarcoma. Radiotherapy was performed after surgical intervention. Ten months later, he was re-admitted with abdominal pain. At this time, an epidural mass at the 6th-9th thoracic levels was detected on magnetic resonance imaging, and acute promyelocytic leukemia was diagnosed. After systemic chemotherapy, partial remission was achieved. We aimed to present this rare case with its remarkable follow-up findings.

Key words: chloroma, acute promyelocytic leukemia, epidural mass, thoracic spine

Granulocytic sarcoma or chloroma is a tumor derived from myeloid cell precursors. They are encountered as a clinical sign coincidental to or an alarming sign of future acute myelocytic leukemia (AML) and other myeloproliferative diseases or acute lymphoblastic leukemia [1, 2]. Granulocytic sarcoma starts in the subperiostal regions of the ribs, sternum and orbital bones and spreads to soft tissues. In rare cases, however, it can originate in other regions. One of these regions is the spinal epidural area. Granulocytic sarcoma originating there may cause spinal compression symptoms and signs [3, 4].

Here we report on a case with a mass at the thoracic epidural region. The mass was diagnosed as granulocytic sarcoma and treated accordingly 10 months ago, and on follow-up the patient had developed acute promyelocytic leukemia and granulocytic sarcoma in the nearby region.

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Case Report

An 18-year-old male was admitted to our neurosurgery clinic complaining of back pain, numbness in the legs and fatigue 2 years ago. Under the seventh thoracic level, hypoesthesia and paraparasia were present, and clonic Achilles tests produced a positive result on the right and a negative result on the left. Deep tendon reflexes were hyperactive and the Babinski test was positive bilaterally. Examination by magnetic resonance imaging (MR) (Philips Intera, Best, Holland) on the axial, coronal and sagittal planes produced T1W (400, 10) (TR, TE),

T2W (3500, 120) images and T1W images after contrast medium (gadopentetate dimeglumine, Magnevist, Schering, Germany) administration, 0.1 mmol/kg. The matrix value was 512×512. Conventional SE T1W thoracic MR (Fig. 1) revealed a solid lesion that was intraspinal extradurally located between the second and fifth thoracic vertebral levels with a size of about 5×2×2 cm. The lesion was diffusely stained after intravenous contrast substance injection and isointense with the spinal cord on T1W images without contrast, suggesting the differential diagnosis of a neurogenic mass, lymphoma or metastasis. All hematologic and biochemical tests were within the normal range. Peripheral blood smear and bone marrow examinations were all normal. The total laminectomy procedure was performed at thoracic 2–5 levels. Excision material was a 4×5×5 cm-sized mobile soft extradurally located and easily bleeding solid mass lesion. Though the immunohistochemical examination revealed a LCA+, CD79+, CD3- large cell lymphoma diagnosis, myeloperoxidase staining proved the granulocytic sarcoma diagnosis (Fig. 2). Clinical and laboratory findings were normal during 10 months of follow-up. Radiotherapy was the treatment chosen, and 4 courses of it were administered. The patient received local radiotherapy totalling 1,500 cGy between T2 and T5 using a linear accelerator. After a 10-month asymptomatic period he was readmitted with complaints of abdominal pain, fever and anorexia. On physical examination, a fever of 38.5 °C, pale skin and mild tenderness on palpation of the epigastric region were encountered. One day later; hypoestesia, paraplegia under the seventh thoracic level, a positive clonic Achilles test on the right, a negative Achilles test on the left, hyperactive deep tendon reflexes and a bilaterally positive Babinski test were detected. MR imaging, proving that thoracic levels 2–5 laminectomized, showed a solid lesion, placed intraspinally and extradurally at thoracic levels 6–9, which was isointense with the spinal cord and diffusely stained after intravenous contrast substance injection on T1-weighted images (Fig. 3). Laboratory studies revealed a hemoglobin level of 6.6 g/dL, a leucocyte count of 6,100 per mm^3, a thrombocyte count of 74,000 per mm^3, an erythrocyte sedimentation rate of 150 mm per hour and a uric acid level of 8.0 mg/dL. Prothrombin time, partial thromboplastin time, fibrinogen, and fibrin degradation products were found within the normal range. An upper endoscopy revealed some areas of mild erosion on the duodenal bulbus. A peripheral blood smear examination showed 76% promyelocyte, 18% lymphocyte, and 6% monocyte. Blastic cells were stained peroxidase positive. Erythrocyte morphology showed anisocytosis and poikilocytosis with slight hypochromia. The percentage of APL cells in the bone marrow was about 90%. Computed tomography of the thorax showed an accumulation of some pleural fluid at the posterobasal segment and

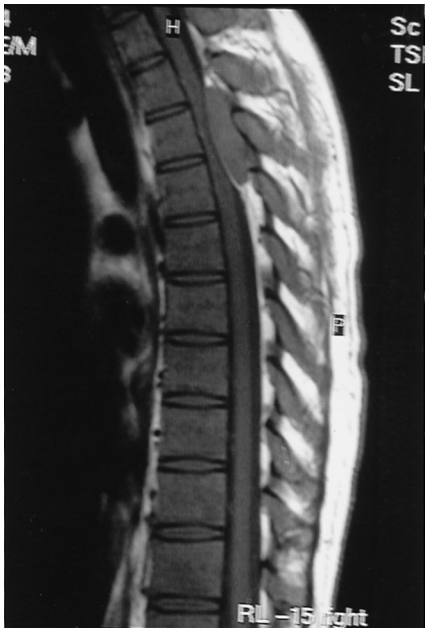

Fig. 1 Initial examination. Sagittal T1W SE thoracic MR revealed a solid intraspinal lesion extradurally located between the 2nd and 5th thoracic vertebral levels and about 5×2×2 cm in size.

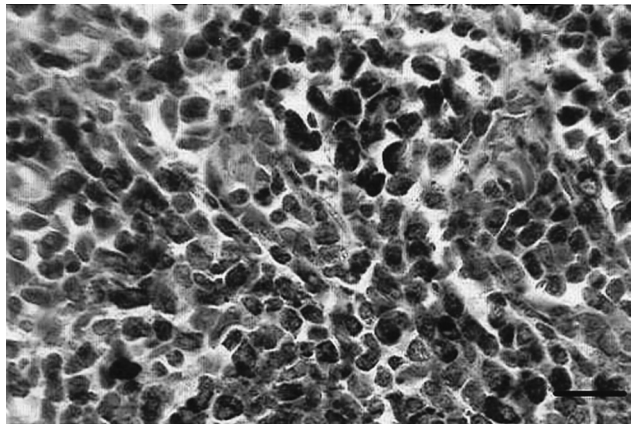

Fig. 2 Histologic section of epidural mass of granulocytic sarcoma. The immunostained section shows an expression of myeloperoxidase in approximately 90% of the cells. Bar indicates 50 μm.

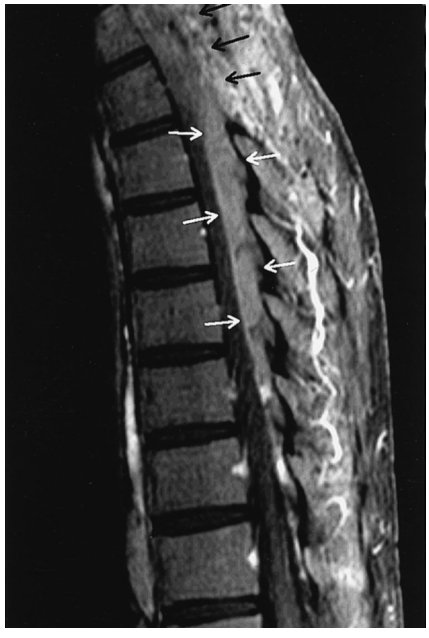

Fig. 3 Sagittal T1W SE post-contrast MR at the time of relapse, which proved that thoracic levels 2–5 were laminectomized (black arrows), showed a solid lesion at an extradural location at thoracic levels 6–9 (white arrows).

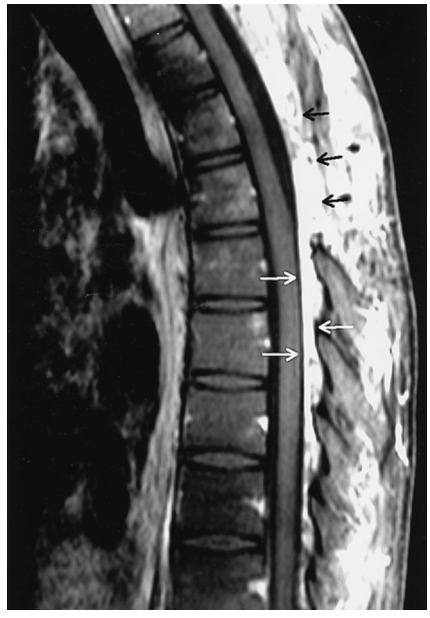

Fig. 4 After chemotherapy, the sagittal T1W postcontrast MR image clearly documented the disappearance of the spinal solid mass (white arrows) under the laminectomy defect (black arrows).

air bronchograms within a consolidated area of the left lung. Flow cytometry resulted in positive myeloperoxidase staining of CD13, CD15, CD33, CD34, and anti-HLA DR. The result suggested a diagnosis of acute promyelocytic leukemia. Chromosomal analysis yielded t(15;17) and q(22;21) translocation. Because of the nonexistence of disseminated intravascular coagulopathy, the regimen of cytosine arabinoside at the dose of 100 mg/m^2/day for seven days and daunorubucine at 45 mg/m^2/day for 3 days was implemented. Repeated spinal MR clearly documented the disappearance of the spinal solid mass (Fig. 4). On follow-up, the paraplegia had resolved completely. After proving the onset of remission, the patient was referred to another medical center to be evaluated for bone marrow transplantation.

Discussion

Granulocytic sarcomas are rare tumors made up of immature granulocytic cells. They are generally seen on the clinical pathway of acute myeloblastic leukemias. They rarely originate in the subperiostal regions of the bones several months before the clinical appearance of leukemia. When the cut surfaces of the specimens show a green colour because of myeloperoxidase enzyme within the cells, they are called chloroma. Chloromas are generally seen on the ribs, sternum and orbital bones as well as in the soft tissues, lymph nodes and skin [1, 5]. Some rare cases of chloroma with the onset from spine and meninges have been encountered in the literature [2, 3, 6, 7, 8]. As they are solid tumors, chloromas originating in the spinal column might acutely cause parestesia and plegias. Those originating in the thoracic regions may lead to paraparasia and paraplegia [9, 10]. Our patient was admitted with complaints of back pain, numbness and weakness of the legs, and upon physical examination we found paraparasia. If the granulocytic sarcoma cases are initially associated with general signs of leukemia, they are difficult to diagnose when evaluating the spinal masses. The cases with spinal onset were generally aleukemic cases, and the diagnosis was made after surgical intervention [3, 4, 7, 11].

In our case, diagnosis of the disease was made after surgical intervention, and leukemic transformation developed 10 months later. If the granulocytic sarcoma case is nonleukemic at the beginning of the disease, it could be confused with a lymphoma under a histochemical diagnosis. The differential diagnosis can be carried out by

antilysozyme immunoperoxidase staining [6, 8, 12]. In our case, a pathological diagnosis of large cell lymphoma was made at first, and then it was changed to chloroma by myeloperoxidase staining. However, in some of the granulocytic lymphoma cases without a myeloproliferative or acute leukemic component there is no development of any leukemic transformation. However, it has been reported that majority of the cases show acute leukemic transformation in about 10 months [8].

The patient was admitted to our hospital with acute abdominal symptoms 10 months after the initial diagnosis. We found an acute myeloblastic transformation state with a solid mass at the thoracic epidural region and pneumonia of the left lower lobe. The therapeutic strategy for granulocytic sarcoma cases that has been recommended to use either chemotherapy or radiotherapy or a combination of both [6, 7]. In our case, the diagnosis was established after surgical intervention, and radiotherapy was indicated. Since the second solid mass diagnosed as chloroma was near the previous lesion, and radiotherapy was performed previously, a second course of radiotherapy could not be used at the time of recurrence. Since special treatment protocols are not available for the state of acute leukemic transformation of granulocytic sarcomas, the treatment strategies do not differ. As radiotherapy and surgical interventions had already been performed, we chose one of the protocols used to treat acute myeloblastic leukemia. When the remission was assured, the patient was referred to another medical center for bone marrow transplantation.

We aimed to introduce a case with spinal compression syndrome due to recurrent granulocytic sarcoma at the thoracic epidural region. Granulocytic sarcoma should always be kept in mind when making the differential diagnosis in patients with signs of spinal compression. After the diagnosis of granulocytic sarcoma is made, those cases should be followed strictly for acute leukemic transformation.

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