Granulocytic Sarcoma with Orbit, Cauda Equina, Muscle and Peripheral Nerve Extension but without Bone Marrow Involvement

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Abstract

We report a very rare case of granulocytic sarcoma (GS) with muscle and peripheral nerve extension but without bone marrow involvement. A 53-year-old woman presented with sciatic pain and diplopia. Magnetic resonance imaging revealed bilateral orbital and cauda equina region tumors. The blood cell count, and bone marrow histology and cytology were normal. The characteristic cerebrospinal fluid (CSF) cytologic picture of CD14+, CD33+, CD4+, CD56+ and positive nonspecific erastase staining suggested the diagnosis of GS. The patient underwent intrathecal and systemic chemotherapy, as if she had acute myeloid leukemia (AML). This case emphasizes the value of CSF cytological examination and the use of an immunocytochemical marker.

Key words: granulocytic sarcoma, meningeal extension, cerebrospinal fluid cytology

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Introduction

Granulocytic sarcoma (GS) is a rare localized tumor composed of immature granulocytic precursors (1). Cases have been reported under the term chloroma, which refers to the green coloration of some of these tumors on gross examination (1, 2). GS occurs most often with acute myeloid leukemia (AML) or chronic myeloid leukemia (CML); however, it is seen rarely prior to the onset of bone marrow disease (3-5). The diagnosis is difficult, especially if the tumor does not occur in the context of AML. Here we report a very rare case of GS with cerebrospinal fluid (CSF) extension but no bone marrow involvement of leukemia at presentation.

Case Report

A 53-year-old woman began experiencing pain in her left sural region. Her medical history included angina pectoris and atrial fibrillation. After one month, she experienced diplopia and bilateral leg weakness. The range of her left sural pain increased and she suffered left sciatic pain. She presented with left eyelid ptosis two months after onset. Brain magnetic resonance imaging (MRI) revealed bilateral orbital tumors (Fig. 1A, B). The orbital tumor decreased spontaneously (Fig. 1C). Her left eyelid ptosis and diplopia were resolved; however, she developed a progressive two-month history of right arm and bilateral leg weakness. Sagittal T1-weighted gadolinium (Gd)-enhanced image revealed high signal spots from the conus to cauda equina (Fig. 1D). The blood cell count was normal. CSF cell count was 56/μm³ (95% mononuclear cells). CSF angiotensin-converting enzyme and lysozyme were slightly elevated at 0.3 IU/l (normal, 0) and 0.4 μg/ml (normal, 0). CSF flow cytometry was not done. Nerve conduction study showed demyelination and axonal degeneration. Biopsy of the left anterior tibialis muscle showed mild myopathic change and aggregated mononuclear cells (Fig. 1F). Biopsy of the left sural nerve showed collapse of the myelin sheath (arrows, Fig. 1G), but no other vasculitis was found. Teased fiber preparation showed ovoid myelin. The tentative pathological diagnosis was multiple motorsensory axonal neuropathy. She began to...
undergo steroid therapy (1,000 mg/day for three days followed by daily steroid supplementation for two months) for the possible involvement of an autoimmune disease such as sarcoidosis, and her muscle weakness improved with the disappearance of high signal spots from the conus to cauda equina two months after steroid therapy (Fig. 1E). She was admitted to our hospital five months after onset. Her general physical examination was unremarkable. Cranial nerve testing was normal. Motor examination revealed 4/5 strength in the upper and lower extremities with slight atrophy in the proximal muscles. Her deep tendon reflexes were diminished in the lower extremities without pathological reflexes. There was decreased pinprick sensation in the left leg and decreased vibration sensation in the bilateral feet. Neck stiffness and Kernig’s sign were absent. The blood cell count, and bone marrow histology and cytology were normal. Her antinuclear antibody levels, including anti SS-A, anti-SS-B, and anti-ds-DNA were all negative. She had normal or nega-
tive studies for serum angiotensin-converting enzyme, sphi-
lis, serology, p- and c-ANCA, rheumatoid factor and chest
X-ray. Motor conduction velocity of both tibial nerves and
sensory conduction velocity of both sural nerves were de-
creased. The amplitude of compound muscle action potential
of both tibial nerves and sensory nerve action potential of
both sural nerves were decreased. Electromyography (EMG)
of the triceps, rectus femoris and tibialis anterior muscles
showed chronic denervation of long duration and high am-
plitude motor unit potentials with a reduced interference pat-
tern on voluntary contraction of the muscle. At lumbar
puncture, CSF pressure was 140 mmH₂O. Cell count was 51/
mm³. Cell morphology of CSF showed atypical mononuclear
cells (Fig. 1H). CSF protein concentration of 161 mg/dl was
elevated and the glucose level of 70 mg/dl (blood glucose
82 mg/dl) was normal. CSF flow cytometry gave the follow-
ing results:keratin-, CD3-, CD19-, CD20- and CD56+.
She was first suspected of having NK cell lymphoma six months
after onset. She was treated with chemotherapy consisting of
intrathecal methotrexate 15 mg, arabinoside cytosine 40 mg
and prednisolone 10 mg twice a week. CSF cytology re-
turned to normal. She had improvement of both lower mus-
cle weaknesses; however, she experienced severe headache
thirteen months after onset. The present case emphasized that CSF cytology
may be the key to the diagnosis of GS.

GS is difficult to diagnose and is often initially misdiag-
nosed, particularly if the tumor does not occur in the context
of AML. As in the present case, most of these cases are
confused with non-Hodgkin’s lymphoma (NHL) or, less
often, with undifferentiated carcinoma. The characteristic cy-
tological picture of CSF tumor cells in our case emphasizes
the value of CSF cytology in the diagnosis of GS, especially
when there is no hematological involvement. The prognostic
significance of GS occurrence even without initial myeloge-
nous disorders is considerable because it suggests that these
patients will develop AML. The interval is relatively short
and leukemia will occur within a period of months after dis-
covery of the tumor. In localized tumors which resemble
NHL or undifferentiated malignant neoplasm, GS should be
suspected. The present case emphasized that CSF cytology
may be the key to the diagnosis of GS.

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