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Granulocytic sarcoma (chloroma) causing spinal cord compression

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Abstract. Granulocytic sarcoma (chloroma) is a rare solid tumor of myelogenous stem cells, usually appearing in patients with acute myelogenous leukemia and less commonly in patients with chronic myelogenous leukemia or myeloproliferative disorders. We present a spinal epidural granulocytic sarcoma causing thoracic spinal cord compression in a patient with chronic anemia secondary to myelofibrosis.

Key words: Chloroma – Myelofibrosis – Spine MRI

Case report

A 77-year-old man presented in October 1989 with chronic anemia, at which time bone marrow biopsy revealed myelofibrosis. He was treated with monthly blood transfusions, progressing to weekly transfusions in February 1991. In March 1991, he developed low back pain; there was no associated trauma, and the pain was worse when he was supine as opposed to standing. He had unsteadiness of gait, but motor and sensory examination was normal, as were bladder and bowel function. He was admitted for pain management in late April 1991. An EMG at that time showed peripheral neuropathy but no radiculopathy.

Plain radiographs of the spine were normal. MRI demonstrated a contrast enhancing, posterior epidural soft tissue mass extending from T5 to L2, displacing and compressing the spinal cord anteriorly (Fig. 1). Differential diagnosis included extramedullary hematopoiesis (EMH), lymphoma, and leukemic mass. No biopsy was performed because of the patient's profound thrombocytopenia (platelet counts ranged between 15,000–30,000/mm^3 despite transfusions). Radiation therapy was not offered because of potential marrow toxicity in the setting of profound pancytopenia. The patient became paraplegic nine days after the MRI, and expired 4 days later.

A limited autopsy demonstrated a tan, shiny, cylindrical, epidural mass, 0.9 cm in diameter posteriorly in the spinal canal. Microscopic examination revealed primarily immature cells of the granulocytic series. Bone marrow biopsy revealed granulocytic hyperplasia and a "left shift", consistent with blast transformation of an underlying myeloproliferative disorder. A final diagnosis of granulocytic sarcoma (chloroma) was made.

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Discussion

Epidural granulocytic sarcoma (chloroma) was first described by Burns in 1811. He described a greenish-yellow tumor of the dura mater of uncertain etiology. King first coined the term "chloroma" in 1853 (from Greek *chloros,* green), and Dock described the association between chloroma and leukemia in 1893. The characteristic color is due to elevated levels of the enzyme myeloperoxidase in tumor cells of myeloid origin; this enzyme is present in lesser amounts in normal myeloid cells [1]. However, the tumors are usually gray, white or tan-brown, as in the present case, owing to the oxidation of the enzyme upon exposure to air. The green color can be reproduced by treating the tumor with hydrogen peroxide or sodium metabisulfite. For this reason, the pathologic term granulocytic sarcoma is preferred; myeloblastoma has also been used [2]. The tumors are soft or hard [3].

Granulocytic sarcomas have been reported in 3.1–9.1% of patients with acute myelogenous leukemia (AML) [1, 4, 5]. Less commonly, they have been reported in chronic myelogenous leukemia, and more rarely, myeloproliferative disorders, including polycythemia vera, hypereosinophilia, and myeloid metaplasia. In the latter case, their occurrence is prognostically ominous, often associated with conversion to AML, or blast crisis [1]. They usually occur concurrently with clinical evidence of leukemia, although they may arise while the patient is in clinical remission, and may antedate the diagnosis of leukemia by up to two years. They are more frequent in children, with 60% of patients under 15 years of age [6]. Their incidence is rising, secondary to prolonged survival times in patients with myelogenous leukemias.

Bone (including cranium, sacrum, sternum, ribs, and spine) is the most common site, followed by skin, soft tissues, lymph nodes, and dura mater [7–9] although

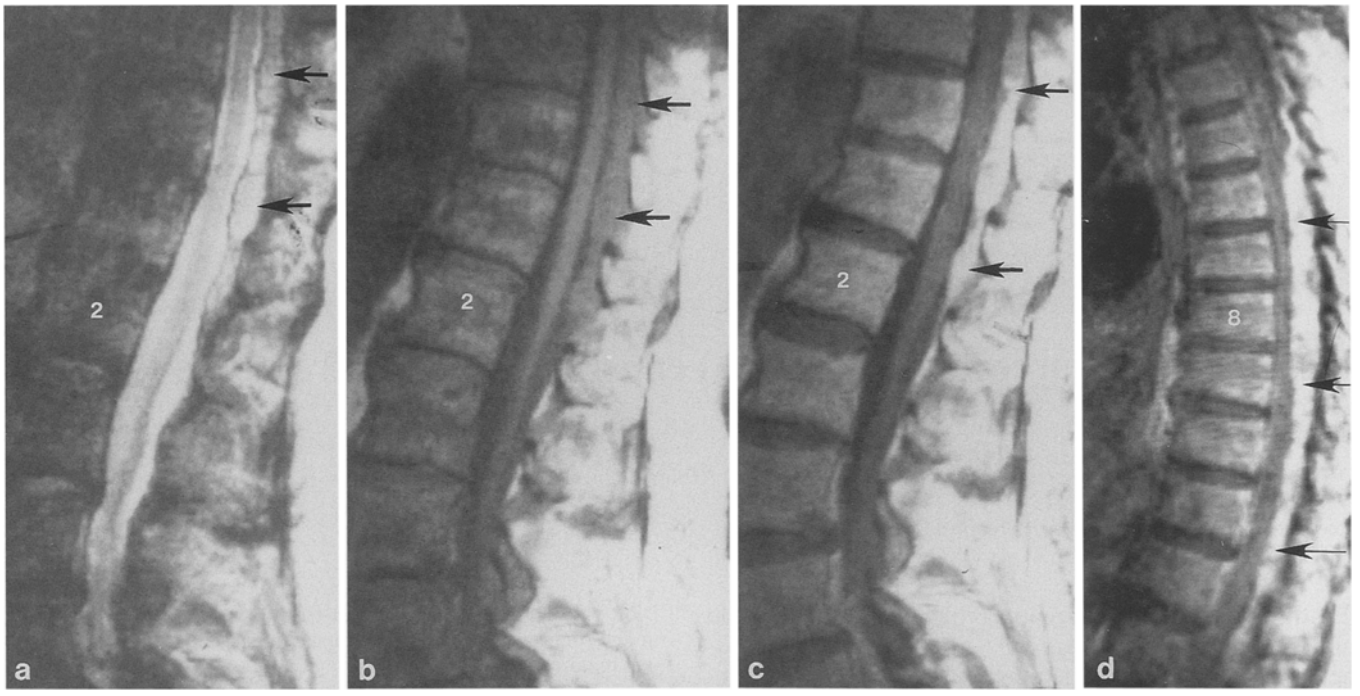

Fig. 1 a–d. Sagittal MRI demonstrates a posterior extradural mass extending from T5 to L2 *(arrows)*, hyperintense relative to the spinal cord on a T2-weighted image (**a**), isointense with cord on a T1-weighted image (**b**) and demonstrates homogeneous contrast enhancement (**c, d**). The lesion appears to compress the lower thoracic spinal cord. Autopsy revealed granulocytic sarcoma (chloroma). *2,* L2 vertebral body *8,* T8 vertebral body

involvement of every organ system has been reported [3, 10]. Involvement of bone is typically subperiosteal. It has been postulated that leukemic cells enter the periosteum by migrating from the marrow through Haversian canals, and thence invade the dura mater [11]. Direct involvement of the CNS is rare, but has been reported [11]. It has been suggested that leukemic cells infiltrate the CNS via perivenous adventitial tissue connecting the dura mater and subarachnoid space. The brain is infiltrated by direct spread from the arachnoid mater [12].

Patients with chloromas involving any tissue present with local pain (78%), tumor nodules (65%), and motor or sensory disturbances (52%), in addition to the usual signs and symptoms of leukemia [3]. Spinal canal involvement by granulocytic sarcoma has been reported to cause back and leg pain, urinary incontinence, and acute paraplegia, often with fulminant progression [4, 5]. Adequate control of the tumor can be obtained with radiation and/or chemotherapy [9]; surgical intervention is rarely necessary, except in cases of acute cord compression [2].

The CT and MRI appearances of spinal epidural chloromas have not been previously described other than in a patient with spinal cord compression secondary to epidural chloroma who became symptomatic after a lumbar puncture [13], for whom only unenhanced T1-weighted spinal images were illustrated.

Intracranial granulocytic sarcomas have been described on CT as of intermediate to high attenuation (60–80 HU) and typically demonstrate uniform contrast enhancement [6, 14, 15]; this CT pattern is, of course, nonspecific. On MRI intracranial lesions have been described as isointense with white matter on both T1- and T2-weighted images [7], or isointense with gray matter on T1-weighted images and isointense with white matter on T2-weighted images [6]. Angiographically, these tumors demonstrate mild to moderate hypervascularity, usually arising from a meningeal artery.

The differential diagnosis of a predominantly posterior, extradural spinal canal mass extending for several levels in the thoracic region includes hematoma, abscess, meningioma, granulocytic sarcoma, metastatic disease, lipomatosis, lymphoma, and EMH. Many of these lesions can be differentiated on the history or the results of imaging. However, EMH, a rare entity, may radiographically and clinically mimic chloroma, and has been called pseudochloroma [16]. Furthermore, EMH occurs in many of the groups of patient prone to chloroma, including those with leukemia and myeloproliferative disorders, and may mimic intraspinal chloroma on CT and MRI [16]. Spinal EMH is most common in the thoracic region, and has been reported to cause spinal cord compression. EMH masses can be treated with radiation therapy and/or surgery [16–21].

It is important to differentiate EMH (pseudochloroma) from granulocytic sarcoma (chloroma) for two reasons. First, chloroma carries an ominous prognostic significance, usually indicating recurrence of leukemia, or conversion of a non-leukemic condition to leukemia. Second, EMH is treated with radiation therapy, with doses of 9–35 Gy in 10–15 fractions [17, 20, 21], while chloromas have been treated with radiation doses to 30 Gy initially, followed by systemic chemotherapy [4–6, 9, 11, 15]. Since these lesions can appear identical on CT and MRI, biopsy

may be the only way to separate them. Despite the risks of bleeding in patients with leukemia and myeloproliferative disorders due to thrombocytopenia, biopsy should be strongly considered when the diagnosis is in doubt.

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