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Granulocytic Sarcoma (Chloroma): Sphenoidal Sinus and Paraspinal Involvement as Evaluated by CT and MR

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Granulocytic sarcoma (chloroma) is an unusual tumor composed of primitive cells of the myeloid series. It occurs in younger patients with acute myelogenous leukemia. The lesions present grossly as solid, greenish tumors composed of primitive precursor cells of the granulocytic series. Chloroma refers to the greenish color of the tumor, which can be attributed to the myeloperoxidase in the tumor cells [1]. The tumor may occur before, during, or after the onset of systemic myelogenous leukemia. No sex predilection is noted. Common sites of the tumor include the orbits, skin, bone, and sinuses, with CNS involvement rarely seen. Systemic chemotherapy and local irradiation techniques are being developed for this entity [2], and the frequency with which it is recognized will certainly increase as further reports are made.

Radiologic evaluation by CT of the intra- and extracranial leukemic infiltrates is well documented in the current literature [1, 3–10]. Limited reports describe the MR findings of intracranial [6, 11, 12] and paraspinal [13] granulocytic sarcomas. We present two cases of chloroma, one with sphenoidal sinus involvement and the other with a paraspinal lesion, that were accurately depicted by MR with CT correlation.

Case Reports

Case 1

A 50-year-old woman with a 4-year history of chronic myelogenous leukemia presented with worsening anemia, fatigue, and a 1-week history of frontal headaches and diplopia. Admission laboratory studies demonstrated a white blood cell count of 49,000 cells/mm³, with 33% myeloblasts, hematocrit = 30.5%, and platelets = 771,000. Physical examination was significant for a right cranial nerve VI palsy, without papilledema, hemorrhage, or exudate; the remainder of the examination was unremarkable. Unenhanced cranial CT demonstrated an isodense mass lesion in the posterior aspect of the right sphenoidal sinus extending through the clivus into the region of the prepontine cistern (Fig. 1A, arrow). Subsequent cranial MR imaging with parameters of T1 (600/16/2 [TR/TE/excitations]), spin-density (2600/17/1), and T2 (2600/90/1) accurately demonstrated the full tumor burden in the sphenoidal sinus (Figs. 1B, 1C, and 1D, respectively), and clearly showed the lesion's margins, particularly its pos-

terior extension, where it invaded and destroyed the clivus and extended into the anterior aspect of the prepontine cistern. While the CT examination showed direct evidence of bone destruction in the clivus, it is clear on the sagittal MR image that the clivus must have been destroyed, as its normally bright T1 signal was replaced by an isointense lesion. The signal intensities of the lesion in this case were isointense relative to gray matter on T1-, spin-density, and T2-weighted sequences. On the basis of clinical, laboratory, and radiologic findings, we determined that this cranial mass lesion was a granulocytic sarcoma (chloroma), and treatment was instituted subsequently. The patient received 20 Gy of cranial irradiation to the area over a period of 2 weeks, with significant improvement in symptomatology over the subsequent 2 months. She showed a satisfactory white blood cell count response to reinstitution of oral Hydreia therapy for her myeloblast crisis.

Case 2

A 36-year-old man with a 1-year history of acute myelogenous leukemia presented with a 6-week history of progressively worsening low back pain radiating to his left flank without motor or sensory deficits. Laboratory studies demonstrated normal white and red blood cell counts, and a physical examination showed tenderness to palpation over the lower thoracic spine and left flank regions. MR imaging of the lower thoracic spine region was performed using T1- (500/17/2) (Figs. 2A and 2B) and T2- (1600/120/2) weighted sequences. These studies showed an isointense paraspinal mass on the left at T11, extending through the neural foramen into the spinal canal with impingement on the T11 nerve root, thecal sac, and spinal cord, with invasion of the vertebral body. CT of this area prior to needle biopsy (Fig. 2C) revealed an isodense paraspinal lesion but did not delineate the component of the tumor, which lay within the spinal canal. This area was shown clearly to be involved by tumor on the MR examination. Additionally, the tumor within the neural foramen was delineated far better on the MR images than it was on the CT examination, on which there was a barely perceptible loss of fat density in the neural foramen. CT-guided needle biopsy showed granulocytic sarcoma histologically. Irradiation to this area of the thoracic spine was instituted, consisting of 21 Gy over 2 weeks, with pain control administered by IV morphine. The patient reported some improvement in symptoms, and a follow-up MR study 3 weeks after completion of irradiation treatment showed significant reduction in the size of the paraspinal mass (Fig. 2D).

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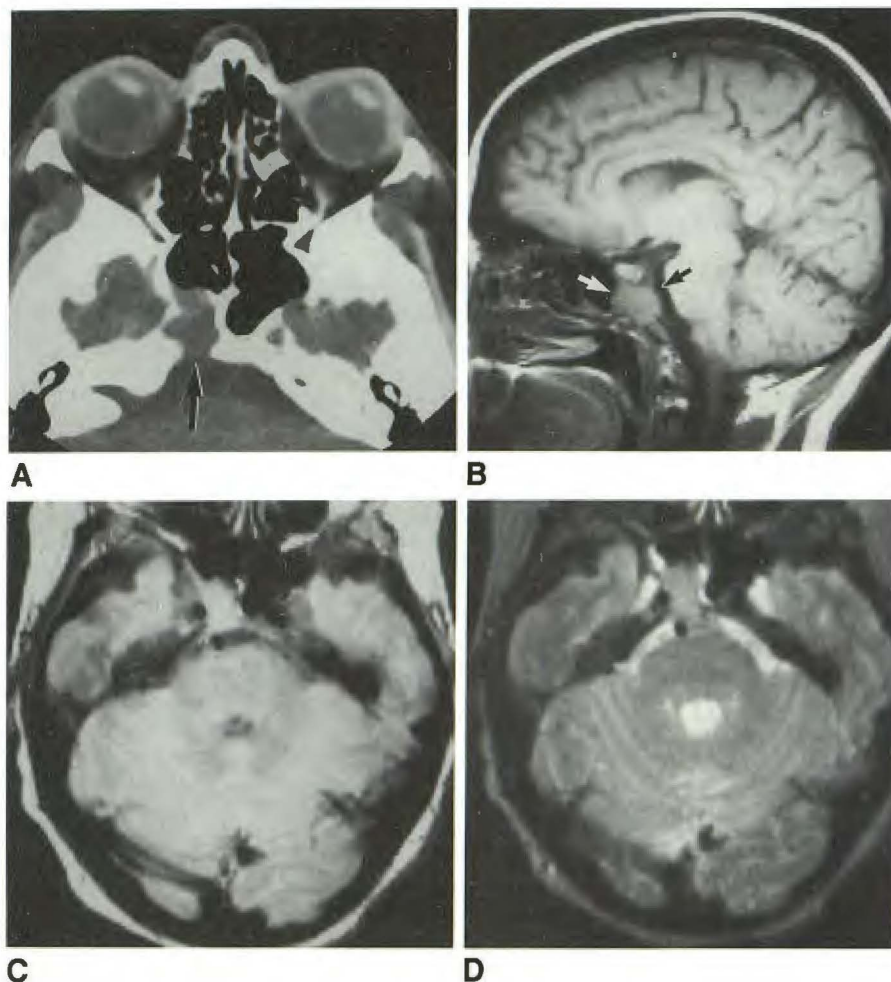


Fig. 1.—A, Unenhanced cranial CT scan shows mass in posterior aspect of right sphenoid sinus, with clear destruction of clivus (arrow).

B, T1-weighted sagittal MR image (600/16/2) shows hypointense mass (arrows) extending posteriorly through clivus (black arrow). Tumor involves the posterior sphenoid sinus, clivus, and pontine cistern.

C, Spin-density axial MR image (2600/17/1) shows isointense lesion in posterior right sphenoid sinus.

D, T2-weighted axial MR image (2600/90/1) shows isointense mass in posterior right sphenoid air cells.

Discussion

Granulocytic sarcoma (chloroma) is a solid tumor of myelogenous cells seen in patients with acute or chronic myelogenous leukemia [1, 3–13]. The rate of occurrence is approximately 3–9% of patients with acute or chronic myelogenous leukemia [12, 13]. The tumor occurs most often in patients under age 15, and there is no sex preference [12]. The lesion may occur before or after a laboratory diagnosis of leukemia has been made or chemotherapy begun [14–16]. The myeloperoxidase within the tumor cells gives it a greenish appearance on gross inspection, thus leading to the original designation of chloroma [8]. Nevertheless, the gross appearance of the tumor is somewhat variable, and may mimic the appearance of lymphoma, rhabdomyosarcoma, or other neoplasms [17].

Chloromas occur in both intra- and extracranial locations. Intracranial chloromas are thought to arise from dural and subarachnoid infiltration by leukemic cells via superficial arachnoidal veins and surrounding adventitia [4, 5, 12, 18]. An intracerebral leukemic mass may result if the pia-glial membrane is disrupted [4]. Extraaxial chloromas arise by the same pathophysiologic mechanism, with the two favorite sites of involvement being the orbits and paranasal sinuses [3, 5, 8,

12]. Extracranial chloromas may occur virtually anywhere in the body. Previous reports have included paraspinal/intraspinal masses [3–5, 13, 16, 17, 19], chest [3, 5, 17], abdominal/pelvic viscera [5, 14, 17], bone—particularly skull, sinuses, spine, sternum, ribs, and pelvis [3, 17]—and other tissues, including lymph nodes, skin, and salivary glands [17]. The tumors tend to present in these locations as a mass lesion with local invasion and symptomatic pain referable to the structures involved, and rarely may be multiple [1]. As with intracranial chloromas, the paraspinal/intraspinal lesions are thought to arise from perivenular arachnoid spread of leukemic cells [4, 12]. Both the thecal sac and spinal nerve roots may be involved, although not as commonly as the cranial nerves [4].

Radiologic characteristics of chloromas have been defined by a number of different imaging techniques. On CT, intracranial lesions have been demonstrated as iso- or slightly hyperdense relative to normal brain parenchyma, often with surrounding edema, and as having a tendency for uniform contrast enhancement [9–12]. The differential diagnosis of chloroma (intracranial) on CT includes lymphoma, meningioma, and metastatic lesions, among other possibilities. Spinal involvement has been previously well documented by myelographic and CT-myelographic techniques, with nonspecific

Fig. 2.—A, T1-weighted axial MR image (500/17/2) shows isointense paraspinal mass with extension through neural foramen into spinal canal (arrows), with T11-vertebral body involvement (arrowhead).

B, T1-weighted coronal MR image (500/17/2) shows extradural component of paraspinal lesion extending through foramen at T11, with impingement on spinal cord.

C, Axial unenhanced CT scan shows left paraspinal mass at T11 level. Intraspinal component of tumor cannot be recognized as separate from thecal sac, except for portion in neural canal, where it obliterates the fat density.

D, T1-weighted coronal MR image (500/17/1) 3 weeks after irradiation therapy shows interval reduction in size of paraspinal tumor mass.



findings of an isodense intra- or extradural mass impinging on the thecal sac and/or nerve roots, occasionally causing complete spinal block [2, 13, 19]. Considerations in a differential diagnosis of a paraspinal lesion, based on the above two techniques, would include para/intraspinal abscess, hematoma, metastatic lesions, or primary tumor such as a neurofibroma [4, 13]. Additionally, gallium-67 scans have shown increased uptake in chloromas [1].

With the advent of MR cranial and body imaging, chloromas are beginning to be defined by this radiologic method as well. Although limited by the total number of reports on MR evaluation of chloromas, certain tissue characteristics are emerging on T1- and T2-weighted images. We would suggest that intracranial chloromas (and those arising in contiguous structures such as the orbits or paranasal sinuses) show a propensity for slight hypointensity on T1-weighted images and isointensity on T2-weighted images [11, 12] relative to white matter (Figs. 1B–1D). This pattern of signal intensity may also be seen with meningiomas [20]. The extracranial appearance of chloromas on MR is less well characterized, but paraspinal chloromas have been described as having intermediate signal on T1- and T2-weighted images. The signal characteristics in our case are in agreement with the previously reported findings [13]. This pattern is in contrast to that seen with tumors

of neural origin, which typically manifest hyperintensity on T2-weighted images. Our second case also demonstrates that MR imaging is a useful radiologic method for monitoring the therapeutic response of paraspinal chloromas to irradiation and/or chemotherapy. No use of gadopentetate dimeglumine or other paramagnetic enhancing agent has been reported to date.

Therapy for both intra- and extracranial lesions has shown favorable results, with prompt recognition and early initiation of treatment protocol. Intracranial lesions have shown high sensitivity to irradiation with or without intrathecal chemotherapy [2, 4, 9, 11, 12]. Similarly, with intra- and paraspinal involvement, localized irradiation of the tumor mass and systemic chemotherapy have shown promising results [2, 3, 5, 10, 13]. Radiologic evaluation concurrent with and following therapy has provided useful information about therapeutic efficacy.

An intra- or extracranial mass lesion in a patient with proved or suspected myelogenous leukemia presents a diagnostic dilemma for the clinician and radiologist. While rare in terms of the actual number of recognized cases relative to patients with myelogenous leukemia, chloroma (which is responsive to irradiation and chemotherapy) is an important diagnostic consideration.

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The reader's attention is directed to the commentary on this article, which appears on the following pages.