Radiologic manifestations of chloroma in pediatrics leukemia patients

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Aims and objectives

Focal aggregation of extramedullary myeloid precursor cells associated with leukemia has been referred to as granulocytic sarcoma (GS), myeloid sarcoma or chloroma. This study was designed to summarize the radiologic presentations including pertinent imaging features of chlroma in different organs.
Methods and materials

Patients

Records of all 40 patients who received a diagnosis of chloroma from 2007 to 2012 were retrieved from the division of pediatrics hematology database at our institution, which is a tertiary referral children cancer center. Diagnosis of granulocytic sarcoma was based on histologic examination of biopsy specimens from affected sites. Complete hematologic investigations, including bone marrow aspiration and trephine biopsy, were performed in all patients. The diagnosis of myeloid leukemia was based on bone marrow and peripheral blood examination. Clinical parameters, including primary hematologic diagnosis, treatment of granulocytic sarcoma, and treatment outcome, were retrieved.

The patients ranged in age from 1-12 years, 15 of the patients were females and 25 were males.

Imaging

Retrospective review of the patients’ radiological studies was done using PACS system. Most imaging was performed at the radiology department in our institution. We retrieved the few CT scans that had been obtained at private institutions. Over time, all patients had undergone multiple imaging studies. Almost all patients had CT studies fifteen patients had MR imaging which was performed with a 1.5-T magnet. Only five patients had US studies.

Two experienced radiologists reviewed all imaging studies, paying particular attention to the site of relapse and imaging features.
Results

Clinical Characteristics

The patients ranged in age from 1-12 years, 15 of the patients were females and 25 were males.

Of the 40 patients studied, ten cases had chloroma as a primary presentation of the leukemia who had no systemic disease. The other 30 patients had preexisting acute myeloid leukemia, which was either in remission or in systemic bone marrow relapse at the time of presentation.

Seven patients had unifocal granulocytic sarcoma, whereas the rest had multifocal disease. Treatment included either chemotherapy, radiotherapy, or a combination of chemotherapy and radiotherapy, with bone marrow transplantation. Complete remission was achieved in five patients, partial remission in three, and no response in three. Five patients experienced sequential extramedullary relapses (>1-year intervals), arising at different sites.

Sites of Granulocytic Sarcoma

A total of 70 granulocytic sarcoma tumors were found in 40 patients over the 6-year period. The orbit, intracranial and paranasal sinuses were the most common sites of granulocytic sarcoma.

Sites of orbital involvement (n = 17): intraorbital extraconal masses (n = 12), retroocular mass (n = 2), perineural mass (n = 1), intramuscular mass (n = 2).

Para nasal sinus involvement (n = 12). maxillary sinus affection is the commonest (n=7)

The intracranial involvement (n = 14): dural masses (n = 11), leptomeningeal (n = 1) and intra-axial mass (n = 2)

The intraspinal involvement (n = 5), pure intraspinal mass (n = 3), extension of paraspinal masses (n = 2), The subcutaneous choloroma (n = 3), The liver (n = 3), focal lesion (n = 1), periportal infiltration (n = 2), The parotid region (n = 2), skeletal muscles (n = 2), the petrous region (n = 4), abdominal cavity (n = 2), breast (n = 1), kidneys (n = 2) pancreas (n = 1), testis (n = 1) and adnexa (n = 1).

Imaging Characteristics

Myeloid sarcoma generally presents as discrete soft tissue solid nodules or masses with variable enhancement. Generally, they were isodense to muscle on CT scans,
and isointense and hyperintense (mild to moderate) on T1- and T2wi, respectively. On US, these lesions are usually iso to hypoechoic. None of these lesions show internal calcification.

Orbital chloroma can affect any of its compartments: the extra conal mass with or without bony affection is the commonest location, the extra ocular muscles, lacrimal gland as well as the optic nerve can also affected, the pattern of affection being bilateral and usually yet not always symmetrical can help in differentiation from other pediatrics orbital lesions (Fig 1,2).

On MR imaging, most of the intracranial chloroma are extra axial and they are usually iso to hypointse on T2WI with vivid uniform enhancement on post contrast series, these lesions are hypercellular so it usually shows restricted diffusion with reduced ADC value (Fig. 3). This finding may help to differentiate such lesions from other meningeal lesions in the children.

The affection of facial bone forming a mass involves the paranasal sinuses was one of the commonest encountered presentation of chloroma. It may present with aggressive behavior destructing the bony boundaries extending intra orbitally and intracranially (Fig. 4, 5).

A spinal granulocytic sarcomas are either extension from para spinal mass (Fig. 6) or intraspinal extramedullary nodules (Fig. 7) both lesions enhanced markedly.

A huge abdominal mass with engulfment of the kidney are encountered in two cases of our patients, and in both cases these lesions were the first presentation of the leukemia. The lack of internal calcifications helps in differentiation between such lesions and the neuroblastoma, and the encasement of the vessels help in differentiation of them from the masses of the renal origin (Fig.8).

The hepatic affection was noted in two pattern either periportal hypodense infiltration or focal lesions.

Breast and skin lesions were isodense to muscle on CT (Figs.9). In the skin lesions, moderate enhancement with and without rim enhancement was also noted.

In two patients, muscle granulocytic sarcoma was multifocal and showed heterogeneous enhancement (Fig. 10).
Granulocytic sarcoma sited in the pancreas (Fig. 11), lacrimal gland, testes, and kidneys tend to cause diffuse involvement rather than discrete masses.

Female genital tract is a rare location of the granulocytic sarcoma, heterogeneous hypoechoic adenxal mass lesion with low vascularity may raise the suspicious of hematological malignancy (Fig. 12).
Fig. 1

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16-yo female presented with proptosis and weight loss.

- Right parietal subglial soft tissue mass.
- Enlarged extraocular muscles bilaterally.
- Enlarged right optic nerve.

Pathological dx: granulocytic sarcoma

Fig. 2

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Fig. 3

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Fig. 4

An ill defined solid mass lesion is seen centered upon then sphenoid sinus destructing its bony boundaries shows vivid enhancement with notable restricted diffusion.

Pathological dx: granulocytic sarcoma

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7 years old female with AML presented with chronic headache

- An ill defined solid mass lesion is seen centered upon the skull base involving the nasopharynx, petrogopalatine fossa, the skull base with intracranial extra-axial mass shows vivid enhancement.

- Pathologic dx: granulocytic sarcoma

Fig. 5

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3 years old male patient with AML developed urine retention with over flow

A well defined intrapinal dural based mass seen at the sacral region extending along the neural exit foramina. It shows hypointese signal on T1WI and isointense signal on T1wi with evident enhancement on post Gd series.

Fig. 6

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1 years old male patient presented with delayed milestones

Multiple enhanced nodules seen at the intraspinally around the nerves at the sacral region.

N.B: left iliac region hematoma post bone marrow aspirat is also noted. AML is diagnosed after bone marrow analysis.

Fig. 7

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Fig. 8

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1 yo male patient with multiple scalp pumps

- Multiple enhanced nodules seen at frontal and parietal region of scalp as well as the gluteal region

Pathological dx: choloroma

Fig. 9

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Fig. 10

1 yo male patient with facial swelling

Bilateral enlarged temporalis muscles with diffuse infiltration and enhancement.

Multiple enhanced nodules seen at the muscle at the anterior compartments of both thigh

Pathological dx: choloroma
1 years old male patient with AML

- Bulky pancreatic head forming mass like lesion displacing the superior mesenteric vessels (a), totally disappeared after chemotherapy (c)
- Well defined left parotid mass lesion (b), totally disappeared after chemotherapy (d)

Fig. 11

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Fig. 12

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Conclusion

Granulocytic sarcoma (GS), myeloid sarcoma or chloroma is defined as a focal aggregation of extramedullary myeloid precursor cells associated with leukemia. GS is a mass of blast cells outside the bone marrow; the term "chloroma" was first used in 1853 to refer to the green color of the tumor caused by high content of myeloperoxidase. Yet, because up to 30% of these tumors can be white, gray, or brown, Rappaport renamed them granulocytic sarcoma in 1966 [1].

Generally, all these forms are uncommon, usually associated with medullary leukemia. Yet, it also can be found in association with other myeloproliferative disorders including myeloid metaplasia, myelofibrosis, polycythemia vera, and chronic eosinophilic leukemia [2].

Granulocytic sarcoma can affect any part of the body but orbital, intracranial, facial and abdominal affection is frequently reported. Bone affection is also common. The pathogenesis of bone involvement has been postulated to be via trans-haversian canal migration of leukemia cells from the bone marrow to the periosteum and dura [3]. Similarly, central nervous system involvement has been theorized to occur via perivascular or perineural routes from direct dural extension or through capillary migration [4]. Granulocytic sarcoma in the gastrointestinal or genitourinary system may arise de novo from nests of hematopoietic cells.

Granulocytic sarcoma usually presented in patients had a history of AML. But it could be the first presentation of the disease, even in these cases; the patients developed AML within 2 years.

Granulocytic sarcoma presented clinically with variable symptoms and signs related to either the mass itself or disturbance of the organ which harbored the mass [5].

Management of granulocytic sarcoma consists mainly of systemic chemotherapy for the underlying leukemia, and usually has good therapeutic results. Surgical debridement or radiation therapy is needed in when urgent decompression is needed, or if the lesion is refractory to systemic chemotherapy.

The radiological knowledge about the diagnostic criteria of the granulocytic sarcoma is so important and has a great impaction in the proper management. As the prompt diagnosis will facilitate appropriate treatment and disease control, and the misdiagnosis of this potentially curable disease would have lead to unnecessary surgery and increased morbidity to the patient. [6].
In conclusion, granulocytic sarcoma in pediatrics usually occurs as extramedullary manifestation of acute myeloid leukemia. The clinical presentations are variable and may include compressive or obstructive symptoms. Despite the variable nature of the disease, there are a few features, particularly of central nervous system granulocytic sarcoma, that may help to distinguish these lesions from other common complications of leukemia. Signs strongly suggestive of granulocytic sarcoma are multiple, enhancing, solid masses occurring at different sites and time points during the course of disease in a patient with either acute myeloid leukemia or myeloproliferativ or myelodysplastic disorders. So the radiologist should be aware of chloromas when evaluating a local mass of unknown etiology, especially in patients with a known history of AML. Knowledge of this diagnostic possibility can expedite treatment for the patient and eliminate unnecessary procedures.
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References


