Introduction

Acute myelogenous leukemia (AML) is uncommon in children, representing about 15% of all leukemia in the pediatric population [1]. Granulocytic sarcoma (GS), previously called chloroma or myeloblastoma, is an unusual manifestation of AML, and presents as a focal soft-tissue mass. Although in children the orbit is a favored site for GS, other types of cancer are much more common in this location. Thus, a GS arising in the orbit may present a significant diagnostic dilemma when it precedes the development of systemic disease. We present CT and MRI findings of an extracranial mass proven to be granulocytic sarcoma in a 6-year-old otherwise healthy boy with several months’ history of worsening unilateral proptosis. This case is unique in providing exquisite CT and MRI correlation and in demonstrating rapid response to therapy. Further, as cytogenetics were positive for the t(8;21) translocation, this case provides opportunity for discussion of the associated incidence of this translocation and concomitant better prognosis.

Case report

A 6-year-old boy presented with a 4-month history of progressive proptosis of the right eye after failing lengthy treatment for a presumed allergic reaction. He had no constitutional signs or symptoms. Results of the physical examination were normal without evidence of lymphadenopathy or organomegaly. Laboratory studies including a complete blood count and white blood cell differential showed normal values. CT of the orbit was performed, demonstrating an extracranial mass in the right orbit centered on the lateral orbital wall. The mass showed minimal bony destruction (Fig. 1). MRI (Fig. 2) defined the mass lateral to the lateral rectus muscle with inferior and superior extension. Mild destruction of the greater wing of the sphenoid was seen, and the mass wrapped around the lateral orbital wall. Isointense to gray matter on T1- and isointense to white matter on T2-weighted images, it enhanced uniformly. Differential considerations included rhabdomyosarcoma, metastatic neuroblastoma, eosinophilic granuloma, and GS.

Biopsy of the mass demonstrated small round cells that were intensely myeloperoxidase positive, which established the diagnosis of GS. A bone marrow biopsy and aspirate demonstrated 5–8% myeloblasts with Auer rods. Cytogenetic analysis of the bone marrow aspirate revealed a t(8;21) chromosomal translocation. After completion of 1 month of induction chemotherapy the proptosis resolved, and MRI demonstrated resolution of the orbital mass (Fig. 3).
Discussion

GS is an unusual manifestation of leukemia, seen in only about 5% of Caucasian cases of AML [2]. Originally termed ‘chloroma’ because of the greenish hue resulting from the large amount of myeloperoxidase present, these tumors can also be gray, white, or brown, depending on the oxidative state of this enzyme. Therefore the more general term ‘granulocytic sarcoma’ (or occasionally ‘myeloid sarcoma’) is now employed.

The incidence of GS in Western literature appears to be decreasing, but there is significant geographic and racial variation [3]. The orbit appears to be a favored site for GS in the pediatric population. In a study of Turkish patients, OGS occurred in 20 (36%) of 56 children with AML [4]. Similarly, in a review of pediatric leukemia in Africa, almost half of the patients with AML had OGS [3]. Importantly, there appears to be a very strong association of OGS with those AML cases demonstrating a t(8;21) translocation, and the presence of the t(8;21) in AML is associated with a good prognosis [5].

GS is thought to occur in bone marrow and then spread via Haversian canals to penetrate periosteum and form a soft-tissue mass. This would account for the typical location near bony structures. These tumors occur most frequently in the skull, sinuses, orbits, spine, ribs, sacrum, and sternum [6].

Fig. 1 Contrast-enhanced CT at presentation shows right proptosis and an extraconal uniformly-enhancing soft-tissue mass in the lateral orbit, with preseptal extension

Fig. 2 a Coronal FSE T2-weighted MRI shows an extraconal mass isointense to white matter extending inferior and lateral to the orbital wall. b Coronal T1-weighted MRI shows mass isointense to gray matter. c Axial gadolinium-enhanced T1-weighted fat-suppressed image shows uniform enhancement of extraconal mass