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Orbital Wall Infarction in Sickle Cell Disease: MR Evaluation

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Summary: Orbital wall infarction in a patient with sickle cell disease can present with periorbital swelling and subperiosteal collection and thus can mimic infection on CT. However, MR not only provides excellent morphologic information but, by characterizing the nature of the collection as containing blood, and by identifying bone marrow abnormality, can lead to the diagnosis of orbital wall infarction.

Index terms: Sickle cell disease (SS, SC); Orbits, magnetic resonance; Orbits, abnormalities and anomalies

Bone infarction is a common feature of sickle cell disease (SSD). However, orbital wall infarction remains a rare complication with only a few cases reported in the literature. We present this case to illustrate the usefulness of magnetic resonance (MR) in helping to differentiate cellulitis from infarction.

Case Report

During one of multiple admissions for vasoocclusive crises, an 18-year-old woman with known SSD developed left facial swelling. Physical examination on admission had revealed a healthy-appearing woman with normal temperature. The morning after admission, the patient developed left facial swelling with pain and tenderness. The patient was begun on oral and topical antibiotics. Ophthalmologic exam revealed normal visual acuity with no limitation of ocular motility. The left upper lid was markedly edematous. Computed tomography (CT) (pre- and postintravenous contrast material) (Figs. 1A and 1B) performed on the third day of admission revealed pre- and postseptal inflammatory changes with a lateral extraconal collection that was thought to represent either an abscess or a subperiosteal hemorrhagic effusion. The walls of the orbit were unremarkable.

Over the next 2 days, even though the patient remained afebrile, she developed worsening proptosis and mild limitation of ocular motility. An MR image was obtained on a 1.5-T unit; it revealed extensive orbital inflammatory changes (Figs. 1C–1E) and a subperiosteal collection. This subperiosteal collection demonstrated heterogeneous signal on noncontrasted T1-weighted images with slightly increased intensity when compared with adjacent muscle (particularly on fat-suppressed images). It remained of high signal on both proton density- and T2-weighted images. These signal characteristics were more consistent with the presence of blood products than of pus or, less likely, simple fluid. The sphenoid wing also demonstrated abnormal increased signal on T1- and T2-weighted images within the marrow, indicating edema and hemorrhage (Fig. 1E).

On the same day (day 5), a bone marrow scan (Fig. 1F) revealed decrease in uptake of the left orbit compatible with infarction. A Tc-99m three-phase bone scan (Figs. 1G and 1H) revealed an increase in soft-tissue uptake indicating hyperemia. An associated slight increase in uptake of the left sphenoid wing was also noted on delayed static images. This finding was felt to represent osteoblastic activity with reactive bone formation (1).

Because of continued clinical concern, the patient was brought to the operating room for exploration which revealed a sterile hemorrhagic subperiosteal effusion. All cultures (including those in resin bottles sensitive to antibiotic-treated material) were subsequently negative. Postoperatively the patient did well with complete resolution of her symptoms. Repeat MR performed 2 weeks after discharge revealed resolution of the inflammatory changes with residual abnormal signal in the sphenoid wing.

Discussion

SSD has been noted to involve all ocular and orbital structures, including conjunctiva, iris, vitreous, retina, and orbital apex. Bone infarction, a predominant feature of SSD, can rarely involve the orbit and frontal bones masquerading as orbital cellulitis. To our knowledge, there have been eight cases reported in the clinical English-language literature. Orbital wall infarction was confirmed in four cases by demonstrating decreased activity on either Tc-99m bone scanning (three cases) (2–4) or bone marrow imaging exams (one

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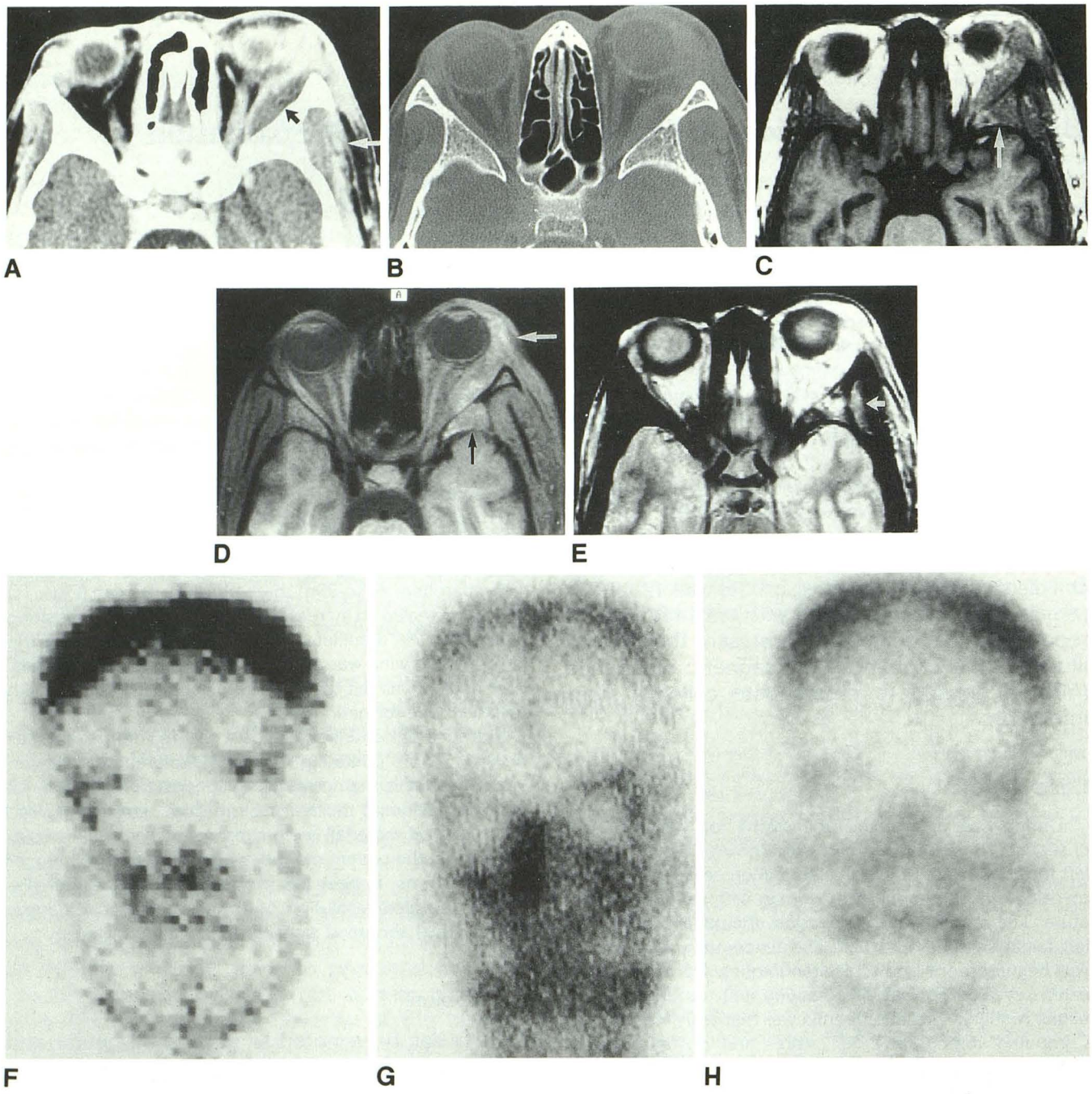

Fig. 1. *A*, Axial contrast-enhanced CT scan reveals an elliptical-shaped low density (*black arrow*) in the extraconal superolateral aspect of the left orbit that was interpreted as representing a collection. Thickened enhancing tissue is seen on periphery of this low density and anterior to it in the region of the lacrimal gland. In addition, there is swelling and increased density of tissues in the temporalis fossa (*white arrow*).

B, Bone window images fail to show any bone abnormality.

C, Axial MR scan (600/15/2) (TR/TE/excitations) performed on the same day as the CT scan reveals slightly increased intensity in the region of the left sphenoid wing (*arrow*) as well as a collection of similar intensity in the left superolateral orbit.

D, Axial nonenhanced MR scan (500/15/2) with fat saturation reveals increased intensity in the left sphenoid wing (*black arrow*) and similar intensity in the collection in the lateral aspect of the left orbit. There is also increased intensity noted in the enlarged displaced left lacrimal gland (*white arrow*).

E, Axial MR scan (3000/30/1) reveals increased intensity in the left sphenoid wing and in tissues lateral to it in the temporalis fossa (*arrow*). The increased intensity of the intraorbital collection is not differentiated from adjacent fat on this image.

F, Bone marrow scan, static images, shows an asymmetric decrease in activity in the left lateral orbital wall, compatible with infarction.

G, Tc-99m three-phase bone scan: static blood pool images show a diffuse increase in activity over the left face compatible with soft-tissue inflammatory changes.

H, Tc-99m three-phase bone scan: static delayed images show slight asymmetric increase in activity over left bony orbit representing osteoblastic activity.

case) (5). Three cases reported "normal" Tc bone scans (5). Mallouh et al, Seibert et al, and Karacostas et al (2, 6, 7) all reported cases that described epidural collections seen on CT in association with orbital inflammation. Two were taken to surgery for suspected abscess (the third patient was not a surgical candidate) where a sterile hematoma was found in each case (2, 6). Garty et al (4) describes an additional two cases of periorbital swelling with ecchymosis on physical exam which were also felt to be secondary to bone infarction, as demonstrated by decreased activity on Tc-99m scans. Surgical confirmation or further imaging was not performed in either case.

Although the presentation of orbital wall infarction as periorbital swelling is slowly becoming recognized, it is still difficult in an acute clinical setting to differentiate between infection and sterile infarction. Often the clinical history, ie, afebrile illness, is helpful. Although a collection may be seen with either inflammation or infarction, the additional feature of hemorrhage within the effusion (either intraorbital, as demonstrated in our case, or epidural, as shown by others) may also support the diagnosis of infarction. The etiology of this hemorrhagic material is unknown. There is usually a lack of trauma or evidence for bleeding diathesis in either family or patient. Karacostas et al (7) postulate that microvascular "sludging" of sickled cells may lead to local vessel wall necrosis and subsequent extravasation of blood products within the adjacent spaces.

Appropriate imaging studies provide additional information. Tc-99m bone scanning in conjunction with bone marrow imaging remains the mainstay for evaluation of possible bone infarction once that question has been raised. MR has already proved helpful in evaluation of bone marrow infarction in long bones although focal hemorrhagic collections were not reported (8). In the orbit, MR may provide useful morphologic detail of the orbital contents as well as of the adjacent bony orbital wall. While CT fails to show any bone abnormality, hemorrhage and edema in the marrow space are well demonstrated by MR and may suggest infarction as in our case. MR is helpful in characterizing collections and thus differentiating nonhemorrhagic from hemorrhagic collections. These additional insights may provide the clinician with enough information to support the diagnosis of infarction (even in light of an associated collection), thus obviating the need for invasive surgery and/or long-term antibiotic therapy.

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