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CT of Orbital Multiple Myeloma

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Plasma cell myeloma is a rare cause of exophthalmos. In 222 intraorbital tumors, Forrest [1] found only one case; Offret [2] reported two cases in 676 orbital tumors. In a review of the literature on orbital myeloma, Rodman and Font [3] found only 30 proven cases. Since then, four additional cases have been reported [4–7]. We report two additional proven cases of orbital myeloma, both of whom underwent orbital computed tomography (CT). Both had proptosis, the presenting feature in one, an extension of known disease in the other. To our knowledge, only one previous case of orbital myeloma has been investigated with CT [4].

Materials and Methods

Both patients were scanned with a Mark 1 EMI head scanner using 8 mm collimator and the images were viewed on a 160 × 160 matrix. Precontrast scans were followed by contrast-enhanced scans using 50 ml of sodium iothalamate (Conray 420).

Case Reports

Case 1

A 29-year-old man had progressive proptosis of his left eye. One year earlier, he was diagnosed as having multiple myeloma, at which time he complained of weight loss, malaise, and backache, and had a significant hepatosplenomegaly. Hematologic investigation revealed a severe anemia (hemoglobin 7.9 g/dl). Serum urea and electrolytes were within normal limits. Bence-Jones proteinuria was present. He had elevated total serum proteins (10.2 g/dl) with a reversal of the normal albumin-globulin ratio. Further investigation revealed a monoclonal gammopathy of the IgG type (7896 mg/dl; 904 IU/ml) with an associated immunoparesis. A bone marrow examination confirmed a diagnosis of multiple myeloma with depressed erythropoiesis and granulopoiesis.

Chemotherapy, consisting of melphalan (Wellcome Pty. Ltd., Alkeran) and prednisone, was instituted. One month later, he developed lower limb paresis and loss of bladder sensation secondary to involvement of the fifth dorsal vertebral body with extradural compression of the cord.

His condition remained unchanged until his present admission with left-sided proptosis. CT (fig. 1) displayed a retroorbital mass on the left side. This was thought to be due to myeloma, but because of his poor general condition, a biopsy was not performed. Radiotherapy to the left orbit caused a rapid regression of the proptosis; however, 1 month later, a more severe proptosis occurred on the same side. This responded poorly to radiotherapy. He developed a terminal septicemia which failed to respond to therapy. An autopsy was refused on religious grounds.

Case 2

A 52-year-old woman had a 2 month history of progressively increasing left-sided proptosis and associated blindness. She had no other symptoms. Examination revealed marked proptosis of the left eye with edema of both the upper and lower eye lids, and chemosis. The globe was fixed and nontender, and appeared to be destroyed by the tumor. The left cheek was swollen and infiltrated by the tumor. Plain skull and oribital radiographs showed a large, soft-tissue mass over the left orbit but no definite bone destruction. Hematologic examination revealed a severe anemia with a hemoglobin level of 5.5 g/dl. Serum urea and electrolytes were within normal limits.

CT demonstrated a large mass occupying the left orbit with infiltration laterally into the temporal fossa (fig. 2A). Normal orbital contents could not be identified, and there was thinning of the posterolateral orbital wall. After the intravenous introduction of iodinated contrast medium, considerable enhancement was noted (fig. 2B).

Surgical biopsy of the soft-tissue mass in the region of the orbit showed the presence of homogeneous sheets of well differentiated plasma cells infiltrating the temporalis muscle. After biopsy, protein electrophoresis showed a monoclonal gammopathy of the IgA type (2300 mg/dl; 263 IU/ml). A bone marrow aspirate revealed increased numbers of immature plasma cells, with binucleate forms being prominent. No flame cells were seen. A diagnosis of IgA myeloma was thus made. Radiologic survey of the skeleton revealed no lytic lesions.

The patient was treated with systemic chemotherapy and local radiotherapy to the left eye. After her initial therapy, she refused further treatment and was lost to follow-up.

Discussion

The diagnosis of orbital myeloma in the presence of generalized multiple myeloma does not usually create a

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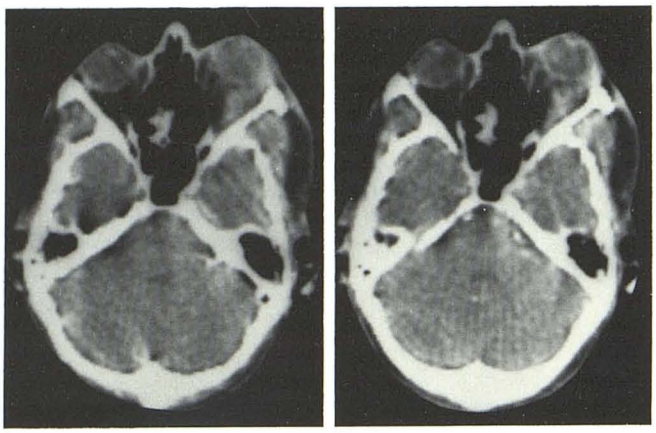

Fig. 1.—Case 1. Pre- (**A**) and post- (**B**) contrast scans of the orbits. Mass in posterolateral aspect of left orbit, causing proptosis. Minimal enhancement of mass after contrast.

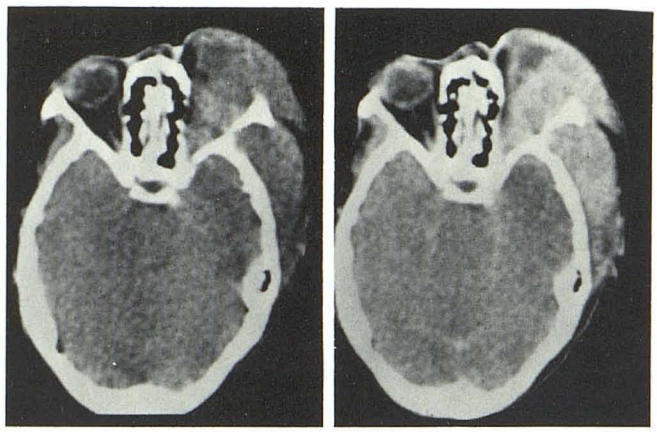

Fig. 2.—Case 2. **A**, Nonenhanced orbital scan. Large retroorbital mass on left with tumor extension into temporal fossa. **B**, Moderate enhancement after intravenous iodinated contrast medium. Probable thinning of posterolateral wall of left orbit.

problem. The characteristic lytic bone lesions on skeletal survey, the replacement of bone marrow by tumor tissue, and the production of myeloma proteins and their constituent polypeptide chains allow the diagnosis to be made in most cases. Myeloma presenting only with proptosis is usually only diagnosed on biopsy. This was so in case 2, where a diagnosis of myeloma was not considered prior to biopsy.

Plasma cell myeloma can affect the eye in many ways. Involvement of the conjunctiva [5], ciliary body [8], cornea [9], sclera, choroid, and iris [10] have all been described. Retinal hemorrhages and papilledema may occur. Intracranial extension may cause ocular nerve palsies, especially involving the sixth nerve [4]. Clinically, orbital myeloma presents most commonly with proptosis. Unlike other malignant neoplasms, pain is rarely experienced [11]. Visual impairment is the second most common feature, varying from total blindness to only slight decrease in vision [4].

Clarke [11] divided orbital myeloma into two categories: (A) primary orbital myeloma—the patient has ocular features suggestive of an orbital tumor, and the lesion arises from the walls or contents of of the orbit; and (B) secondary orbital myeloma—the patient has paraorbital myeloma, and eventually develops symptoms and signs of an orbital tumor due to orbital invasion. The paraorbital structures that are primarily involved include the cranial bones, paranasal air sinuses, nose and nasopharynx.

Either lesion may be the presenting features of what later turns out to be multiple myeloma [12]. In both of our patients, the orbital tumor seemed to arise from within the orbit rather than from adjacent structures. This would tend to fit the description of Clarke type A category. In case 2, the tumor had spread outside the orbit at the time of presentation (fig. 2).

In the radiologic investigation of a patient with suspected orbital myeloma, plain radiographs of the skull and orbit and orbital tomography are very helpful. Bony destruction should strongly suggest orbital myeloma. A radiologic skeletal survey may reveal generalized disease.

Before the advent of CT, angiography had been used by some as a means of diagnosing orbital myeloma preoperatively [13]. None of the angiographic features is specific for myeloma, and the use of intravenous iodinated contrast medium in patients with myeloma is not without danger. Renal complications have been well described in myeloma, especially in the poorly hydrated patient [14].

CT has had a large impact in the diagnosis of orbital disease [15]. Although a definitive histologic diagnosis is not possible, the malignant nature and site of origin of the retroorbital tumor can be clearly visualized. It is a rapid, accurate, safe means of demonstrating retroorbital mass lesions including myeloma.

The CT findings in both of our cases were of a retroorbital mass causing proptosis. In case 1, the globe could be seen separate from the mass lesion, but the optic nerve could not be separately visualized. In this patient, there was minimal enhancement of the lesion after intravenous iodinated contrast medium (fig. 1) (The patient was well hydrated both before and after the scan.)

In case 2, the globe was infiltrated and destroyed by tumor. The left globe was markedly proptosed, and there was tumor extension into the soft tissues of the temporal fossa, with probable thinning of the posterolateral wall of the orbit (fig. 2). Significant enhancement of the lesion occurred after intravenous contrast medium (fig. 2B).

Both of our patients are of particular interest. Case 1 was only 29 years old when first diagnosed as having multiple myeloma. Myeloma generally occurs in the 40–70 year age group and is relatively uncommon in the young, although recently reports on myeloma occurring in young patients have been published [4, 16–18]. In case 2, the presenting feature was proptosis, and only after surgical biopsy was the correct diagnosis made.

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