icked Melkersson-Rosenthal syndrome and was the only initial indicator of widespread systemic disease.

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A 34-year-old woman had a progressively protruding and painful mass of the right orbit for 1 month. On examination, the soft-tissue mass was about 5 cm in diameter and firmly fixed to the right orbit with resultant protrusion of the lower eyelid and proptosis (Figure 1A and B). Visual acuity was no light perception, coupled with limited eyeball motility. There was neither hepatosplenomegaly nor lymphadenopathy. Her family history was noncontributory. Computed tomography of the orbits revealed a large mass measuring 7.1 × 4.9 × 4.8 cm and completely occupying the expanded right orbit without bony destruction (Figure 1C). Magnetic resonance imaging of the orbits showed a large, heterogeneously enhancing soft-tissue mass compressing and pushing the eyeball anteriorly and extending posteriorly to the optic canal (Figure 1D). A small incision was made through the superotemporal conjunctiva and Tenon capsule, where tumor cell invasion with a cicatrizing nature was identified and incised. Histopathological analysis of orbital biopsy specimens demonstrated atypical lymphoid cell infiltration with positive immunoreactivities for CD3 and CD2 (Figure 2). The neoplastic cells were negative for CD56, CD15, CD30, and latent membrane protein 1 (Figure 3). Evaluation of T-cell receptor gene rearrangement using polymerase chain reaction showed monoclonality over the T-Cell re-

Figure 1. Clinical photographs, computed tomographic scan, and magnetic resonance image. A, The right eye, showing obvious protrusion of the eyelid and proptosis. B, Lateral view of the right eye. C, Contrast-enhanced computed tomography of the brain revealed a huge mass occupying the right orbit with no bony destruction. D, Magnetic resonance imaging of the brain showed a heterogeneously enhancing soft-tissue mass extending from the eyeball to the optic canal.
ceptor γ chain gene, consistent with peripheral T-cell lymphoma (Figure 4A). Staging fluorine 18 (18F)-labeled fluorodeoxyglucose positron emission tomography showed lesions with striking 18F-fluorodeoxyglucose uptake over the right orbital region without evidence of hepatic, splenic, or lymph node involvement, supporting the diagnosis of primary peripheral T-cell lymphoma of the orbit (Figure 4B and C). At the 3-month follow-up, the patient refused to receive chemotherapy but completed radiotherapy. Proptosis and the protruding mass in the right eye failed to achieve satisfactory resolution.

Lymphoid neoplasms have been described as the most common malignant orbital tumor, accounting for 6% to 8% of all orbital tumors and up to 15% of all ocular adnexal tumors.1,2 A rapidly progressive protruding mass in the orbit is an unusual clinical feature that is typical of peripheral T-cell lymphoma. The study by Coupland et al3 shows the heterogeneous nature of T-cell lymphoma involving the ocular and ocular adnexal tissues and a varied associated prognosis. Our case highlights that physicians must maintain a heightened awareness of the distinct manifestation of peripheral T-cell lymphoma in developing a differential diagnosis for rapid-onset proptosis.

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Orbital Chondromyxoid Fibroma

Primary tumors of orbital bone are rare, constituting up to 2% of all orbital masses.1 Chondromyxoid fibroma (CMF) is one of the least common tumors of bone, composing less than 1% of bone tumors and less than 2% of benign bone tumors.2-3 Apart from brief case reports,4-6 orbital CMF has not been clearly documented in the ophthalmic literature. To our knowledge, we report for the first time the clinicopathological features and management options of an orbital CMF arising from the frontal bone.

Report of a Case. A 37-year-old woman had slowly progressive swelling of the left upper eyelid temporally associated with occasional headache and shooting pain for 3 years. On examination, visual acuity was 20/20 OU. The left eye showed 4 mm of proptosis with downward displacement, mild blepharoptosis, and choroidal folds at the posterior pole (Figure 1A). Computed tomography disclosed a superotemporal, noninfiltrative orbital mass with erosion of the adjacent frontal bone (Figure 1B and C). Differential diagnosis included lacrimal gland tumors, atypical dermoid cysts, and benign fibro-osseous lesions such as osteoma, fibrous dysplasia, or ossifying fibromyxoid tumor of soft parts.

The patient underwent transcutaneous extraperiosteal orbitotomy. Intraoperatively, the periosteum was separated by blunt dissection from the bone in the peripheral portions of the mass both superiorly and inferiorly. The mass proved to be located mainly in the extraperiosteal space, pushing the inferior and medial periosteal border into the orbital soft tissue. The tumor centrally showed a close connection to the bony wall. En bloc resection including the tumor, surrounding periosteum, and adjacent bony wall was performed.

Macroscopically, the mass measured 20 × 15 × 7 mm. Histopathological examination revealed a CMF (Figure 2A-C). The soft-tissue mass was surrounded inferomedially by periosteum. The periosteum showed calcification superiorly and laterally corresponding to the radio-dense margins on the computed tomographic scan. The lateral borders of the mass consisted of bone. The cellular elements displaying a low proliferation rate (MIB-1 < 1%) were positive for S-100B protein in the central chondroid area and positive for vimentin and smooth muscle actin in the peripheral fibroblastic area but negative for desmin, CD68, CD34, and CD31. Ultrastructural findings (Figure 2D and E) supported the diagnosis of CMF.

Figure 1. Clinical features of an orbital chondromyxoid fibroma arising from the frontal bone in a 37-year-old woman. A, Slowly progressive proptosis, downward displacement, and mild blepharoptosis of the patient’s left eye, of 3 years’ duration. B, Axial computed tomographic scan showing a circumscribed, round to ovoid, superotemporal orbital mass with radio-dense margins and a lucent central component isodense to muscle tissue indenting the globe inferiorly. C, Axial computed tomographic scan with bone window settings clearly delineating the incompletely formed sclerotic rim surrounding the round to ovoid mass with focal erosion (arrow) and thinning of the adjacent frontal bone. D, Marked improvement of proptosis, downward displacement, and blepharoptosis without evidence of recurrence or metastasis 2 years after transcutaneous extraperiosteal orbitotomy with en bloc resection including the tumor, surrounding periosteum, and adjacent bony wall.