Orbital Chondromyxoid Fibroma

Primary tumors of orbital bone are rare, constituting up to 2% of all orbital masses. 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. Apart from brief case reports, 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.
Postoperatively, recovery was fast and unremarkable. Two years after surgery, the patient showed marked improvement of proptosis, downward displacement, and blepharoptosis without evidence of recurrence or metastasis (Figure 1D).

**Comment.** Chondromyxoid fibroma manifests most frequently in the second and third decades of life, more often in males than in females.²,³ The long bones are the most common site, followed by the flat bones and the bones of the hands and feet.³ Craniofacial involvement is relatively rare.³ Histopathological differential diagnosis includes chondrosarcoma, enchondroma, or chordoma.²

Chondromyxoid fibroma with orbital involvement has been reported only in brief case reports.⁴⁻⁶ Hashimoto et al⁴ and Cruz et al⁵ described a CMF of the ethmoid sinus destroying the medial orbital wall. Wolf et al⁶ reported an intracranial CMF of the frontal-sphenoid junction with secondary orbital involvement. In our patient, there was—to our knowledge for the first time—marked downward displacement and indentation of the globe with choroidal folds caused by progressive orbital tumor growth. Because the adjacent periosteum was intact, we elected to take an extraperiosteal approach, separating tumor and periosteum from bone as much as possible. In the area of close adhesion to the adjacent frontal bone, an en bloc resection including the tumor, surrounding periosteum, and adjacent bony wall was performed. Complete en bloc resection is important regarding both histopathological diagnosis and the prevention of tumor recurrence. Simple curettage may favor underdiagnosis and explain in part why the entity has been rarely documented.

In conclusion, CMF should be considered as a rare benign lesion in the differential diagnosis of primary orbital bone tumors.

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quamous cell carcinoma (SCC) in the periocular region can invade the orbit and intracranial cavity. One route of such spread, known as perineural spread (PNS), involves the contiguous spread of the tumor along the potential space between a nerve and its sheath. Perineural spread is associated with a high rate of recurrence, metastasis, and poor prognosis. The usual treatment for SCC with PNS is surgical resection followed by wide-field radiation therapy (RT). We herein report a case of recurrent SCC with PNS treated with intravenous chemotherapy as a single modality with complete and sustained resolution of clinical and radiographic signs of PNS.

**Report of a Case.** A 70-year-old man had undergone excision of an invasive SCC (<1 cm wide) of the right cheek with positive excision margins 2 years before the referral. He developed right facial paralysis and lower eyelid ectropion a few weeks after excision. He underwent surgical repair of paralytic ectropion presumed to be due to idiopathic Bell’s palsy. The ectropion recurred after 8 months. Magnetic resonance imaging revealed an infraorbital right cheek mass that extended into the masticator space. The patient was referred to the M. D. Anderson Cancer Center for further management. The histologic sections of the original lesion on the cheek were reviewed at our institution and the diagnosis of SCC was confirmed (Figure 1). No perineural invasion was noted on the representative section of the original outside specimen.

Extraocular motility examination suggested a right abduction deficit and right esotropia (Figure 2A). Ocular adnexal examination results were significant for right eyebrow ptosis and hypesthesia of the right cheek and cornea. The patient had 11 mm of lagophthalmos, right lower eyelid paralytic ectropion, and an associated corneal ulcer. The patient denied significant pain or paresthesias.

A diagnosis of recurrent SCC of the cheek with extensive PNS and resultant multiple cranial neuropathies was made. Repeated magnetic resonance imaging again revealed a right premaxillary soft tissue tumor with extensive PNS along the right infraorbital nerve with extension to the right pterygopalatine fossa, the right cavernous sinus, and the right Meckel cave (Figure 3A and C).

Given the extensive skull base spread, it was felt that the lesion was not surgically resectable. Because the patient had no significant pain, RT was deferred. Intravenous chemotherapy consisting of 736 mg of carboplatin and 380 mg of paclitaxel was administered every 3 weeks for 4 cycles. After 2 cycles of chemotherapy, the patient experienced significant resolution of clinical (Figure 2B) and radiographic (Figure 3B and D) signs of PNS. Three years after completion of chemotherapy, the patient remains without clinical or radiographic evidence of disease recurrence.

**Comment.** We report here an impressive durable response to systemic cytotoxic chemotherapy delivered as single-modality treatment for advanced recurrent SCC with PNS. Chemotherapy for head and neck SCC is usually delivered as induction neoadjuvant chemotherapy before surgery or RT or concurrently with RT. We were unable to find any other example in the literature of the use of systemic chemotherapy as single-modality treatment for SCC with PNS.

Perineural spread occurs in 2.5% to 14% of head and neck SCCs and 1% to 8% of periorbital SCCs. Of those cases, only 30% to 40% are symptomatic. Patients who develop symptoms may have facial paresthesia, facial paralysis, exposure keratopathy, facial pain, diplopia, and hearing loss. Perineural spread is often missed until advanced stages; thus, the prognosis for patients with PNS is often poor. McNab et al reported that of 21 patients with PNS of SCC in the periorbital region, 13 (62%) had died of disease by the 3-year follow-up.

**Figure 1.** Histologic section of the lesion on the cheek shows squamous cell carcinoma (hematoxylin-eosin, original magnification ×10).