We describe a 23-year-old woman with iridocyclitis, enophthalmos, facial hemiatrophy, and transient numbness of her contralateral upper and lower extremities. The patient was found to have white matter densities in the right hemisphere in magnetic resonance T2-weighted images and vascular malformations involving right vertebral, right carotid, and right anterior cerebral arteries. Histopathologic evaluation of a biopsy specimen of anterior orbital fat and lacrimal gland revealed fibrosis and chronic inflammation. These findings were consistent with the diagnosis of progressive facial hemiatrophy (Parry-Romberg syndrome) in association with iridocyclitis and intracranial vascular malformations.

Progressive facial hemiatrophy (Parry-Romberg syndrome) is a rare entity of unknown origin usually appearing early in life as a slowly progressive atrophy of one side of the face. We report a case of Parry-Romberg syndrome with ocular and intracranial findings. The association of Parry-Romberg syndrome with intracranial vascular abnormalities is rarely reported in the literature.

REPORT OF A CASE

A 23-year-old white woman was first seen with the complaint of painless, intermittent blurred vision in her right eye. Her ocular history was remarkable for enophthalmos of her right eye attributed to a childhood softball injury. Her best-corrected visual acuity was 20/60 OD and 20/20 OS. The left eye was normal. External examination of the right eye revealed 7 mm of enophthalmos and ipsilateral ptosis (Figure 1). Motility was full. A furrow was present above the right eyebrow. Lashes of the right upper eyelid and brow were sparse medially. The eyelids and conjunctiva were not inflamed. Corneal edema and granulomatous keratic precipitates involved the inferior aspect of the cornea. A mild anterior chamber reaction was present. The pupil reacted poorly. Posterior synechiae were seen inferiorly. The iris had sectorial transillumination defects. The lens was clear. Funduscopic examination revealed diffuse nonspecific mottling of retinal pigment epithelium. A photograph of the patient taken years after her softball injury showed no enophthalmos or furrow. A computed tomographic scan evaluation showed enophthalmos with thickened extraocular muscles and no signs of orbital or facial fractures. An increase in soft tissue density was noted throughout the right orbit, especially at the apex, where diffuse atrophy of orbital fat was present (Figure 2). Shortly after coming to our office the patient developed a transient numbness of her left upper and lower extremities. Emergent magnetic resonance imaging showed areas of increased signal intensity in the right hemisphere on T2-weighted images with no visible changes on T1-weighted images (Figure 3). It also showed an area of hypodensity in the right basal ganglia area on T1-weighted images that was hyperdense on T2-weighted images, consistent with an acute white matter infarct (Figure 4). The results of magnetic resonance angiography showed a hypoplastic and narrowed right vertebral artery, a nar-
rowed right intracavernous carotid artery, and absence of an A1 segment of a right anterior cerebral artery (Figure 5). An anterior orbital biopsy was performed, and the results were pertinent for nonspecific inflammation with areas of fibrosis. The above constellation of findings was consistent with the diagnosis of Parry-Romberg syndrome in association with intracranial malformations. The patient’s ocular symptoms improved with the use of fluorometholone drops. When the patient’s condition stabilizes the surgical options for correcting the enophthalmos will be considered.

PATHOLOGIC FINDINGS

Histopathologic examination of orbital biopsy tissues revealed focal chronic nongranulomatous lymphocytic inflammation of the retrobulbar fat and the lacrimal gland. Medial thickening of arterioles with perivascular lymphocytic infiltration was noted in the retrobulbar fibrofatty tissue. The biopsy specimen from the lateral rectus muscle showed atrophy, fibrosis, and focal lymphocytic infiltration (Figure 6).

COMMENT

Progressive facial hemiatrophy was first reported by Parry1 and then described as a syndrome by Romberg.2 It consists of slowly progressive atrophy of skin, subcutaneous tissue, fat, and muscles of one side of the face with an onset during the first or second decade of life.3 Ocular symptoms are reported in 10% to 40% of cases.4 The most common orbital finding is progressive enophthalmos secondary to atrophy of orbital fatty tissue.4 Systemic manifestations occur in 7% of cases.5 They include ipsilateral atrophy of various internal organs or extremities. Central nervous system involvement in the form of ipsilateral atrophy of cortical white matter with intracortical calcifications and unidentified densities in the cortex, manifested as an increase in the signal intensity on T2-weighted magnetic resonance images, has been described.6 Pathologic specimens show a thickened dermis with signs of homogenization and sclerosis of collagen bundles and medial thickening with perivascular lymphocytic infiltration of arterioles in the subcutaneous tissue. Biopsy specimens of muscle reveal degenerative changes with lymphocytic infiltration, a decrease in the number of fibers,
and loss of striations. The cause of progressive facial hemiatrophy is unknown. There are several pathogenetic theories including a viral infection, a sympathetic nervous system dysfunction, a heredodegenerative disease, an autoimmune process, previous trauma, or a disturbance of an angiogenesis during growth and development. The association of intracranial vascular malformations described herein support the theory of an arrested angiogenic process as a cause of Parry-Romberg syndrome.

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REFERENCES


Figure 4. Axial T1-weighted magnetic resonance scan showing hypodensity in the basal ganglia (left; arrow) that increases in signal on T2-weighted images and is consistent with bleeding and an area of an acute infarct (right; arrowhead).

Figure 5. Magnetic resonance angiography showing a hypoplastic right vertebral artery (left; arrow) and a narrowed right intracavernous carotid artery (right; arrow).

Figure 6. Biopsy specimen of the lacrimal gland showing perivascular nongranulomatous lymphocytic inflammation (left; hematoxylin-eosin, original magnification ×25). Lateral rectus biopsy specimen showing fibrosis and lymphocytic infiltration of the muscle fibers (right; hematoxylin-eosin, original magnification ×25).