Alacrima presenting in the third decade

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Aqueous tear deficiency can occur as a congenital or acquired process. Alacrima (lacrimal gland hypoplasia) refers to a wide spectrum of lacrimal secretory disorders that are usually congenital. Symptoms can range from complete absence of tears to hyposecretion of tears; rare presentations include those where there is a selective absence of tearing in response to emotional stimulation but a normal secretory response to mechanical stimulation.

Alacrima has been reported as an isolated congenital finding in a healthy child with no family history of lacrimal secretory disorders.[1] It may be inherited, with numerous Mendelian genetic presentations,[2–4] or may be part of ocular or systemic syndromes.[5]

The developmental mechanism of alacrima is not clear. Possible theories are lacrimal (superior salivary) nuclear aplasia, failure of central or peripheral nervous system innervation,[6] persistence of physiologic alacrima of the newborn[1] or primary lacrimal gland aplasia/hypoplasia.[2]

Acquired causes of aqueous tear deficiency may result from autoimmune conditions, radiation, metastatic and nonmetastatic infiltrative processes of the lacrimal gland, infections, injury and medical therapy.

We describe a young adult who presented with unilateral lacrimal gland hypoplasia.

Case report

A healthy 20-year-old woman presented with a 2-month history of irritation, redness and photophobia in her left eye. She had worn soft contact lenses in both eyes for 5 years. She reported that her left eye was no longer tearing and that emotional stimuli would cause tearing from the right eye but not from the left. A review of systems, including neurologic and rheumatologic systems, was completely normal. She stated that she had had normal tearing with emotion from both eyes until 2 months previously. She denied any history of previous injury, chemical burn, radiation or surgery. She had no history of any joint problems, sexually transmitted disease or malignant disease. Her family history was unremarkable. She was not taking any medication.

The patient's best corrected vision was 20/20 in the right eye and 20/50 in the left eye. She had normal pupils, normal ocular motility and normal facial nerve function. Slit-lamp examination showed the lids, lid margins and puncta to be normal, as were the lid movements. The conjunctiva in her left eye was injected, and diffuse punctate corneal erosions and filamentary keratitis were present. Corneal sensation was normal, and the anterior chamber was quiet. Examination of the right eye was completely normal.

On Schirmer's test with anesthesia, at 5 minutes there was more than 20 mm of wetting in the right eye and less than 1 mm of wetting in the left eye. The lacrimal gland was palpable on the right side only.

Laboratory investigations gave normal results for levels of complement and angiotensin-converting enzyme, and titres of antinuclear antibody, rheumatoid factor and antineutrophil cytoplasmic antibody. Serum protein electrophoresis gave normal results, as did chest radiography.

Orbital computed tomography showed the lacrimal gland on the right side to be normal, whereas that on the left side was small and atrophic or hypoplastic (Fig. 1).

The patient responded well to treatment with a left inferior punctum plug and frequent application of artificial teardrops.

Comments

Most cases of alacrima occur in children who are

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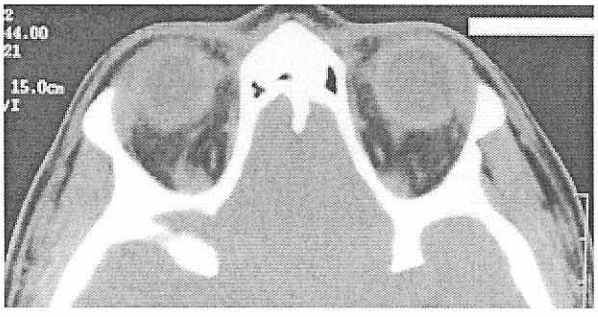

Fig. 1—Axial computed tomographic image of orbits, showing normal lacrimal gland on right and small atrophic or hypoplastic lacrimal gland on left.

otherwise healthy and have no family history of aqueous tear deficiency. The disorder tends to be congenital and bilateral, with early presentation. No single cause has been found. It is possible that alacrima may occur as an abnormal persistence of the normal infantile state. At birth the lacrimal gland is not fully developed, and tears are not produced for some time.[1] This cause has an early presentation, which makes it inapplicable in our case.

Congenital absence of the lacrimal gland or acini may be found as a component of several syndromes, including anhidrotic ectodermal dysplasia, familial dysautonomia, congenital Sjögren's syndrome and Allgrove's syndrome.[7,8] None of the symptoms or signs of these syndromes were present in our patient.

Acquired causes of aqueous tear deficiency that result from autoimmune conditions or infiltrative processes of the lacrimal gland may be uncovered by diagnostic tests when the pretest likelihood is high enough to order them. For example, a patient with joint pain or swelling may have collagen vascular disease, and an investigation consisting of determination of complement levels and titres of antinuclear antibody, rheumatoid factor and antineutrophil cytoplasmic antibody as well as serum protein electrophoresis may reveal a specific (e.g., Wegener's syndrome) or nonspecific (e.g., rheumatoid arthritis) collagen vascular disease. A patient with weight loss or a computed tomographic scan suggestive of a malignant or infiltrative lacrimal gland mass (e.g., lymphoma, sarcoid) would benefit from lacrimal gland biopsy.

To our knowledge, unilateral lacrimal gland hypoplasia presenting in adulthood is rare. Given our patient's good health apart from contact lens wear, it is possible that she had congenital unilateral alacrima with low aqueous production that decompensated with age. However, we cannot rule out the possibility that she had acquired unilateral hypoplasia of the lacrimal gland that resulted from a viral infection or an evolving collagen vascular disease that has not yet manifested systemically.

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