Radiation-Induced Chandler's Syndrome

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Chandler's syndrome is characterized by a triad of symptoms — corneal edema, glaucoma, and iris and angle changes — that result from abnormal proliferation of the corneal endothelium. This is one of the entities of iridocorneal endothelial syndrome that also includes "essential iris atrophy and iris nevi." These entities are linked by unilaterality, a progressive course, and the absence of a family history [1]. While the underlying condition that leads to corneal endothelial changes is unknown, a viral etiology [2] and a progressive corneal endothelioma [3] have been proposed.

We report here two adults with Chandler's syndrome, both of whom

had received scalp radiation treatment for tinea capitus when they were children.

Case Description

A 50-year-old woman presented with the complaint of blurred vision in her left eye and seeing colored haloes around lights with that eye. Visual acuity was 20/20 OD and 20/40 OS, with intraocular pressures of 15 and 50 mmHg, respectively. The examination of the right eye was normal. The left eye had marked corneal edema and broad adhesions of the iris to the periphery of the cornea. The pupil was drawn in the direction of the adhesions, and the iris showed patchy stromal atrophy; the optic discs and visual fields were normal. Specular microscopy showed increased pleomorphism and polymegathism and reduction in endothelial cell density of the affected eye compared to the contralateral normal eye (1,000 cells/mm^2 and 2,500 cells/mm^2, respectively). The clinical and specular microscopy features were suggestive of Chandler's syndrome.

Four years later, the patient was admitted to the hospital with weakness of the right arm and leg that had troubled her for a year. Physical examination disclosed hypoesthesia and a motor deficit of the right arm and leg, as well as multiple basaliomas located mainly on the left side of the nose, face and scalp, some of which were excised and diagnosed as being basal cell carcinoma. Computed tomography revealed a left axial frontoparietal lesion. That tumor was completely excised and the histopathologic diagnosis was meningioma. Her medical history revealed that she had undergone radiotherapy to the scalp for tinea capitis 40 years previously.

Comment

We report a patient with Chandler's syndrome who had received scalp radiation treatment for tinea capitus when she was a child. In addition to the ophthalmological presentations, the patient also had cranial meningioma and multiple basal cell carcinoma lesions of the face and scalp.

The etiology of Chandler's syndrome is unknown. In light of the endothelial changes found in this condition, which are common in neoplastic processes as well, it was suggested that this condition may be a benign tumor of the corneal endothelium [3]. Radiation is one of the confirmed environmental causes for the development of neoplasia of the central nervous system. We and others have shown that patients irradiated for tinea capitis in their early childhood had a significantly increased rate of head and neck tumors, such as meningioma and skin cancer [4]. Chandler's syndrome has never before been associated with irradiation. Dose reconstruction indicated that the dose reaching the base of the brain would be around 15 and 6 cGy to the eye [5]. It is therefore plausible that both the head and neck tumors in our first case described here could have been radiation induced. Furthermore, insofar as her affected eye was located on the same side as the head and facial lesions, it may well be that the eye was located within the radiation beam and thus, like her other tumors, the corneal changes could have resulted from the irradiation applied to the scalp.

The radiation origin of Chandler's syndrome may be overlooked since it is possible that patients irradiated for benign conditions in their childhood generally tend either to forget or were too young to remember having undergone the radiation treatment, and this could account for the lack of corroborative evidence in other similar presentations. Since the first case with radiation-induced Chandler's syndrome, we question each new patient with Chandler's syndrome whether he or she had ever been exposed to radiation to the head. Indeed. we encountered an additional case.

This 60-year-old woman presented with the complaints of pain and blurred vision in the right eye. Examination of the eye was suggestive of Chandler's syndrome, as the high intraocular pressures of 30 mmHg was accompanied by marked corneal edema, large (7 mm) and distorted upwards pupil, patchy iris, stromal atrophy, and attachment of the iris to the upper extremity of the cornea. Specular microscopy showed high variability, increased pleomorphism, reduction in the number of hexagonal cells, and low endothelial cell density (1,500 cells/mm^2 in the affected right eye compared to 2,500 cells/mm^2 in the normal left eye). When queried, the patient reported having undergone radiotherapy to the scalp for tinea capitis 50 years previously.

We contend that our two cases of Chandler's syndrome following irradiation to the head provide evidence to support a cause-and-effect rationale. More such cases will be detected once the physician is alerted to suspect a possible radiation origin of Chandler's syndrome and explores this line of questioning when taking the patient's history.

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