Alveolar Rhabdomyosarcoma That Metastasized to the Orbit

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A 22-year-old woman with a history of hyperthyroidism and a paravaginal alveolar rhabdomyosarcoma developed diplopia 3 months after successful autologous bone marrow transplantation. Results of computed tomographic scan showed apparent enlargement of the right inferior rectus muscle, and thyroid eye disease was suspected. However, results of further evaluation revealed an alveolar rhabdomyosarcoma that metastasized to the orbit.

Rhabdomyosarcoma is the most common primary malignant neoplasm of the orbit in children.1,2 When it occurs in the orbit, it is almost always a primary lesion.3,5 Only two cases of rhabdomyosarcoma that metastasized to the orbit have been mentioned in the literature, but neither has been described in detail.6,7

REPORT OF A CASE

In August 1990, a 20-year-old woman developed a right-sided nontender paravaginal mass that was found to be an alveolar rhabdomyosarcoma (Figure 1). Immunostaining for muscle-specific actin yielded positive results. Results of staging workup revealed vertebral and bone marrow metastases. She received an 11-month course of chemotherapy with cyclophosphamide, doxorubicin hydrochloride, vincristine sulfate, ifosfamide, and etoposide phosphate that resulted in a complete remission by April 1991.

In February 1992, 10 months after completing chemotherapy, the patient was found to be hyperthyroid; she was treated with radioablative therapy of the thyroid gland in March 1992. At the same time, she developed discomfort in the right thigh and a right footdrop. Results of workup revealed recurrent tumor in both pelvic sidewalls and the pancreas. She received local radiotherapy (50.4 Gy) to the pelvic masses and four cycles of chemotherapy consisting of etoposide and ifosfamide. The patient was then treated with carboplatin, cyclophosphamide, and etoposide, followed by which she received an autologous untreated bone marrow transplant. Two months later, computed tomographic scans of the chest, abdomen, and pelvis showed no tumor, and the patient was thought to be in a complete remission. She had no visual complaints at that time.

Three months after the bone marrow transplantation, the patient noted a black shadow in the superior portion of the visual field of the right eye. She saw an ophthalmologist several days later, at which time results of an examination revealed visual acuity of 20/20 with normal color vision OU. Results of visual field testing revealed a superior temporal temporal defect in the field of vision of the right eye, and results of ophthalmoscopy showed a mild epiretinal membrane in the eye. One week later, the patient noted vertical binocular diplopia and was referred to the orbital unit of The Wilmer Ophthalmological Institute (Baltimore, Md).

Visual acuity with correction was 20/20 OD and 20/15 OS. Near vision was J1 OU. The patient identified 10 of 10 Hardy-
Rand-Rittler color plates with each eye. Kinetic perimetry yielded normal results in both eyes, whereas results of static perimetry revealed a slight reduction in sensitivity in the right eye without a clear-cut visual field defect. There was no relative afferent pupillary defect. There was 3 mm of proptosis on the right eye measured with a Hertel exophthalmometer. There was limitation of elevation of the right eye with a right hypertropia of 7 prism dipters at distance and near in the primary position, which increased to 14 PD on attempted downward gaze and changed to a right hypotropia of 9 PD on attempted upward gaze. The left eye did not demonstrate an abnormality of ocular motility. Bilateral corneal and facial sensations were equal and normal. Results of slit-lamp examination revealed normal anterior segments, lenses, and vitreous in both eyes. Results of applanation tonometry revealed normal intraocular pressures. There was no eyelid retraction or lid lag, nor was there inferior scleral show on either side. Results of ophthalmoscopy revealed a normal right optic disc with horizontal linear, choroidal striae in the posterior pole that were consistent with a retrolubular mass. The retinal veins were slightly dilated. No hemorrhages or exudates were observed. The left disc, macula, vessels, and periphery were normal. Results of computed tomo-

graphic scan revealed a mass in the right inferior orbit that was just superior to the inferior rectus muscle in the anterior orbit (Figure 2, left) and that involved the inferior rectus muscle more posteriorly (Figure 2, right).

The patient underwent biopsy of the right inferior orbital mass via a Caldwell-Luc procedure without complication. Results of histopathologic examination of the biopsy specimen revealed numerous small, round cells, distributed among fibrovascular septa with densely staining nuclei surrounded by clear or vacuolated cytoplasm, that were consistent with an alveolar rhabdomyosarcoma (Figure 3). Immunostaining was not performed owing to the histopathologic similarity of this biopsy specimen on hematoxylin-eosin staining to the primary lesion (Figure 1). A systemic workup yielded negative results for additional metastatic disease. The patient is currently receiving local radiotherapy.

**COMMENT**

Hematogenous metastases to the orbit by any malignant process are uncommon. Of 1114 orbital tumors described in three separate series, only 70 (6%) metastasized to the orbit, and none of the metastatic tumors was a rhabdomyosarcoma.

Only two, and perhaps three, cases of rhabdomyosarcoma that metastasized to the orbit have been reported in the literature. Horn and Enterline described a 12-year-old girl who developed a primary alveolar rhabdomyosarcoma in the occipital region that was treated with surgical excision and radiation. The tumor recurred with metastases to the abdomen, spine, calvarium, and orbits. This case was mentioned in a list and no further description of the or-
alveolar lesions was provided by the authors. In a case reported by Henderson, a primary rhabdomyosarcoma arose in the soft tissues of the cheek near the parotid gland. Three years later, the patient developed an orbital mass associated with clinical evidence of metastases to the liver and lungs. There was no histologic confirmation of the orbital mass. Finally, Kirk and Zimmerman described a 9-year-old child who developed a renal tumor that exhibited areas of rhabdomyosarcomatous differentiation at 14 months of age. Almost 8 years later, the patient developed an orbital rhabdomyosarcoma that was thought by the authors to be a second primary lesion rather than a metastasis.

Our patient had an extraorbital alveolar rhabdomyosarcoma that metastasized to the orbit 26 months after the primary paravaginal lesion was discovered and 3 months after an autologous untreated bone marrow was transplanted. Although orbital metastasis of a rhabdomyosarcoma is extremely rare, it should be considered when any patient with a history of an extraorbital rhabdomyosarcoma develops signs and symptoms of an orbital process.

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REFERENCES