Ancient Schwannoma of the Orbit

Sang In Khwarg, MD; Mark J. Lucarelli, MD; Bradley N. Lemke, MD; Morton E. Smith, MD; Daniel M. Albert, MD, MS

The ancient schwannoma is a rare variant of a neurilemoma with a course typical of a slow-growing benign neoplasm. Histologically, it can be confused with a malignant mesenchymal tumor because of increased cellularity, nuclear pleomorphism, and hyperchromatism. Despite the degree of nuclear atypia, mitotic figures are absent. We describe the clinical and histopathologic features of an ancient schwannoma of the orbit.


The ancient schwannoma is a rare histologic variant of a neurilemoma derived from the neural sheath of Schwann. In 1951, Ackerman and Taylor reported 10 neurogenic tumors of the thorax that showed microscopic features typical of schwannomas but that were distinctive because of hypercellular areas with nuclear pleomorphism and hyperchromatism suggesting fibrosarcoma, as well as hypocellular areas with considerable fibrosis. The clinical course, however, was benign in all cases during a mean follow-up period of 3 1/2 years. They interpreted these findings as degenerative changes indicative of long duration and, thus, designated the tumors ancient neurilemomas (schwannomas). Since that first report, several authors have described ancient schwannomas in a variety of locations in the head and neck. We could find only a single reference illustrating orbital ancient schwannoma. The histopathologic features, but no clinical findings, were described.

REPORT OF A CASE

A 52-year-old woman sought care because of epiphora and exophthalmos of the right eye of 4 months' duration. The patient denied diplopia and periocular pain. Her medical history included no relevant information.

Orbital computed tomography with contrast showed a well-demarcated, oval, nonenhancing mass in the intraconal space of the right inferotemporal orbit (Figure 1). Orbital ultrasonography demonstrated low to medium internal reflectivity. The clinical diagnosis was schwannoma vs cavernous hemangioma. Hemangiopericytoma and fibrous histiocytoma were considered less likely diagnostic possibilities.

A right lateral orbitotomy was performed through an extended eyelid crease incision. The mass was gray, smooth, and well encapsulated. The adjacent orbital fat was normal, and there were no fibrous attachments of the mass to the surrounding tissue. The mass was removed in its entirety without violation of its capsule (Figure 2).

At 11 months after the operation, the patient was doing well. Visual acuity was 20/25 OU, and the globe position had normalized.

PATHOLOGICAL FINDINGS

The mass measured 27 × 20 × 20 mm, and the cut surface was avascular, smooth, and yellow. Microscopically, the tumor was composed of plump spindle-shaped nu-
clei with tapering cytoplasm forming interconnecting bands (Antoni type A neurilemoma, Figure 3). In a few areas, the cells were scattered in a loose myxoid matrix (Antoni type B neurilemoma, Figure 4). Some nuclei were hyperchromatic and pleomorphic (Figures 3 and 4). An occasional multilobulated nucleus was noted (Figure 4). No mitoses were present. The diagnosis was peripheral nerve sheath tumor (schwannoma) with ancient features.

**COMMENT**

The histopathologic features of ancient schwannoma are characterized by areas of increased cellularity with nuclear pleomorphism and hyperchromatism, areas of hypocellularity, and considerable fibrosis with focal hyalinization. Microcystic degeneration, hemorrhage with hemosiderin-laden macrophages, lipid-filled foam cells, and local calcification also may be seen. Despite the degree of nuclear atypia, mitotic figures are absent. Occasionally the nuclei show vacuoles or signs of karyorrhexis that could be misinterpreted as mitoses.

These cellular changes can make accurate pathological diagnosis difficult. Increased cellularity and nuclear atypia may lead to the erroneous interpretation of malignant tumor. Dahl reported 6 of 11 cases of ancient schwannoma that were originally misdiagnosed as malignant tumors, such as neurosarcoma, neurofibrosarcoma, or malignant schwannoma.

Certain histological features can assist in the differentiation of ancient schwannomas from malignant tumors. These include the lack of mitoses, evidence of previous hemorrhage, areas of degeneration with hyalinization, and the presence of a histological capsule. Immunohistochemical testing can aid the identification of neural sheath-derived tumors; the results are positive for vimentin, neuron-specific enolase, and S-100 protein and negative for cytokeratin.

Malignant schwannomas are exceedingly rare in the orbit. Some forms of malignant schwannoma exhibiting so-called Schwann cell differentiation with densely packed, often palisading, short spindle cells may resemble ancient schwannoma. These malignant schwannomas, however, show mitoses and infiltrative growth, distinguishing them from ancient schwannomas.

Malignant changes of benign schwannomas have been reported rarely. Schwannomas are less likely than neurofibromas to undergo malignant change. The diagnosis of malignant change of a benign schwannoma requires the following criteria: (1) demonstrable areas of benign schwannoma; (2) unequivocal malignant foci manifested by increased cellularity, numerous mitoses, anaplastic cells, and invasiveness; (3) transitional areas between malignant and benign regions; and...
In the present case, some nuclei were large, hyperchromatic, pleomorphic, and, occasionally, multilobulated. These nuclear features are consistent with degenerative nuclear atypia and suggestive of ancient schwannoma. Mitoses, necrosis, and invasiveness were absent. Moreover, no transitional areas were observed between the areas typical for schwannoma and the foci of atypical cells. Although the duration of this patient’s tumor is unknown, the histological constellation is diagnostic of ancient schwannoma.

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Corresponding author: Mark J. Lucarelli, MD, Department of Ophthalmology and Visual Sciences, University of Wisconsin–Madison Medical School, F4/348 Clinical Science Center, 600 Highland Ave, Madison, WI 53792-3220.

REFERENCES