Orbital liposarcoma masquerading as a hemangioma

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Liposarcomas are the most common soft tissue sarcomas in adulthood, comprising approximately 20% of all sarcomas; most present in the extremities and retroperitoneum (1). Despite the large amount of adipose tissue in the orbit, orbital liposarcomas are rare. Diagnosis can be challenging due to the rarity of the entity and the pathological similarity to benign adipose tumors (2, 3). Fortunately, the advent of immunohistochemical staining has aided in diagnosis. Management of orbital liposarcoma also can be challenging, as illustrated by the case described herein (4).

CASE REPORT

A 67-year-old man reported proptosis, pain, diplopia, and tearing in his right eye for 1 year. Brain imaging with computed tomography (CT) and magnetic resonance imaging (MRI) revealed a well-defined, somewhat oval-shaped mass in the intraconal space of the right orbit measuring 2.7 cm in anterior-posterior, 2.7 cm in transverse, and 2.1 cm in craniocaudal dimension (*Figure 1*). The patient had 20/20 visual acuity bilaterally, with motility limited in abduction and infraduction

Figure 1. Axial computed tomography demonstrating a large intraconal mass in the right orbit with significant proptosis.

and 4 mm of proptosis. Anterior segment examination disclosed superficial punctate keratopathy and 2+ conjunctival injection; fundus examination demonstrated a sharp disk with normal vasculature and normal macula.

The mass was removed using Stryker navigation. A Krönlein lateral orbitotomy with bone flap was performed with subsequent reconstruction of the lateral orbital wall with osteoplasty. The tumor appeared to be encapsulated but adherent to the inferior rectus muscle at surgery. The tumor and pseudocapsule were removed entirely, but because of the adherence to the inferior rectus muscle, it was not believed that complete excision was possible. The specimen was excised in two parts: one measured $2.9 \times 2.4 \times 2.3$ cm and one $1.1 \times 0.6 \times 0.3$ cm. Histologically, the cells were loosely arranged without a prominent plexiform vascular pattern. The nuclei were mostly bland. The tumor infiltrated the extraocular muscle fibers. Staining and immunohistochemistry revealed a Ki-67 index of $<3\%$. The specimen was desmin positive, smooth muscle actin negative, MDM2 positive (*Figure 2*), and S-100 positive. Interphase fluorescence in situ hybridization was performed with probes for MDM2 and intrachromosomal comparison for the chromosome 12 centromere regions. Gains of MDM2, compared to the centromere region, were seen in 58% of cells; 16% of cells contained a high-level increase in MDM2. One year after excision no residual orbital mass was evident.

DISCUSSION

Orbital liposarcomas are typically classified along cytogenetic, morphologic, and clinical lines as either well differentiated (including adipocytic, sclerosing, inflammatory, spindle cell, and dedifferentiated subtypes [5]), myxoid/round cell, and pleomorphic types (6). The well-differentiated category is also referred to as the atypical lipomatous tumor/well-differentiated liposarcoma/dedifferentiated liposarcoma

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Figure 2. Spindle cells in a myxoid background with MDM2 staining, consistent with well-differentiated liposarcoma (250×).

category (ALT-WDLPS/DDLPS). In the orbit, the myxoid type is most common, representing 60% of cases; well-differentiated types account for 30%, and pleomorphic lesions make up the remaining 10% (7).

The classification scheme reflects distinct chromosomal aberrations in each category. Well-differentiated (ALT-WDLPS) lesions have ring or giant marker chromosomes composed of material from chromosome 12q13-15 with amplification of MDM2 and often CDK4 (3). Myxoid/round cell liposarcomas contain a reciprocal translocation involving chromosomes 12 and 16 resulting in production of the TLS/FUS-CHOP fusion transcript. The pleomorphic type has a complex karyotype, as one might expect, with numerous genetic imbalances described; however, 12q13-15 amplification is rarely present (3). The lesion

in question had high levels of expression of MDM2, placing it in the well-differentiated category of orbital liposarcomas.

Once the diagnosis has been made, local control and full excision represent the best approach to enable a cure. Unfortunately, this tumor often invades surrounding structures, as it did in our case. The mass invaded the extraocular muscles, and complete excision would have led to irreparable deficits. Neoadjuvant and adjuvant radiation therapy has led to decreased recurrence rates in liposarcomas in general; however, data for its use in orbital liposarcoma are scarce (8, 9).

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