

Advances in Orbital Tumors

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Take-Home Points:

- A wide variety of benign and malignant neoplasms, inflammatory, and non-neoplastic conditions occur within the orbit
- Primary orbital neoplasms include epithelial tumors, bone and soft tissue tumors, lymphoid tumors, melanocytic tumors, vascular tumors, tumors of the central nervous system and its meningeal coverings, germ cell tumors, and others
- Secondary orbital tumors are either metastatic in origin, or they may secondarily invade the orbit from adjacent structures including the eyelid, conjunctiva, intraocular structures, paranasal sinuses, and nasopharynx
- The lacrimal gland is the only epithelial structure normally present within the orbit. Its epithelial tumors are identical to epithelial salivary gland tumors

Orbital Tumors:

- Orbital tumors and simulating lesions are not very common. However, the orbit is a location where a wide spectrum of neoplastic surgical pathology can occur¹⁻⁹. The Wills Eye Hospital Oncology Service reviewed 1,264 consecutive patients with a space-occupying orbital lesion over a 30-year period⁷:
 - 810 (64%) were benign
 - 454 (36%) were malignant
 - Rhabdomyosarcoma was the most common malignancy in children (median age 7 years)
 - Lymphoma was the most common malignancy in older adults (median age 71 years)
 - Epithelial tumors of the lacrimal gland were more common in middle-aged patients (median age 40 years)

- The general categories of orbital tumors are listed here, and they can be further subcategorized into specific diagnoses ⁸:
 - Inflammatory lesions that simulate neoplasms;
 - Cystic lesions;
 - Lacrimal gland primary epithelial tumors;
 - Metastatic cancer;
 - Secondary tumors.
 - Lymphoid tumors and leukemias;
 - Osseous, fibrous, and cartilaginous tumors;
 - Vascular and hemorrhagic lesions;
 - Primary melanocytic tumors;
 - Peripheral nerve tumors;
 - Myogenic tumors;
 - Fibrous connective tissue tumors;
 - Lipomatous and myxomatous tumors;
 - Histiocytic tumors and pseudotumors;
 - Optic nerve, meningeal, and other neural tumors.

The Orbit: Normal Anatomy and Components:

- The human adult orbit measures approximately 40 mm in height, 45 mm in depth, and has a volume of 30 mL
- The posterior and peripheral borders of the orbit are defined by seven bones of the skull, face, and nose: the frontal, zygomatic, palatine, lacrimal, sphenoid, ethmoid and maxillary
- The major intraorbital components are:
 - The globe;
 - The lacrimal gland;
 - Extraocular muscles;
 - Smooth muscle;
 - Fibroadipose tissue;
 - Blood vessels;
 - Peripheral nerves and sympathetic ganglia;
 - The optic nerve and its meningeal coverings; and,
 - The cartilaginous trochlea

The Lacrimal Gland:

- The lacrimal gland is situated anteriorly in the superotemporal quadrant of the orbit. It is mostly a serous gland, and is divided into two parts, the larger orbital lobe and the smaller palpebral lobe
- Ducts from the gland open into the superior conjunctival fornices and transmit their secretions into the tear film

- Lacrimal gland tumors account for approximately 5%-10% of orbital tumors.
- The histopathologic classification of these tumors is similar to the World Health Organization's classification of salivary gland tumors
- They can be divided into epithelial and non-epithelial, benign and malignant tumors
- Pleomorphic adenoma (benign mixed tumor) is the most common benign tumor of the lacrimal gland. They have a propensity to recur, and they can undergo malignant transformation into carcinoma ex-pleomorphic adenoma. Non-invasive carcinomas have an excellent prognosis after complete excision, while the prognosis is guarded for invasive carcinomas.
- Adenoid cystic carcinoma is the most common malignant epithelial tumor of the lacrimal gland. Mucoepidermoid carcinoma are the next most common, while acinic cell carcinoma, salivary duct carcinomas, adenocarcinomas, and epithelial-myoeplithelial carcinomas are exceedingly rare.

Lymphoproliferative Lesions:

- The orbit normally lacks lymphoid tissue but contains scattered lymphocytes, especially within the lacrimal gland tissue. These cells presumably are the progenitors of orbital inflammatory and neoplastic lymphoid proliferations
- Orbital lymphoproliferative lesions involve the spectrum of benign/reactive processes (reactive lymphoid hyperplasia), to malignant lymphomas
- The differential diagnosis may include orbital inflammatory disease
- Orbital lymphomas are similar to those elsewhere in the body, presenting as primary tumors or as a manifestation of systemic lymphoma
- The majority of orbital lymphomas are low-grade, non-Hodgkin lymphomas including B-cell chronic lymphocytic leukemia and marginal zone / mucosa-associated lymphoid type (MALT) lymphoma
- Hodgkin lymphoma is rare in the orbit

Secondary Tumors:

- Orbital metastases may involve the globe, optic nerve, intraconal and extraconal compartments
- Secondary tumors may invade the orbit from adjacent structures including the eyelid, conjunctiva, intraocular structures, paranasal sinuses, and nasopharynx
- Lung carcinoma, breast carcinoma, prostate carcinoma, and malignant melanoma, are the most common metastatic tumors to the orbit in adults
- Adrenal neuroblastoma, Wilms tumor, and Ewing sarcoma are more common in children

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