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Review Article

Eye and orbital malignancies: An overview

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ABSTRACT

Orbital tumors are classified based on where they occur within the orbit as intraconal, extraconal, and intracanalicular lesions. Patients with larger orbital lesions may present with proptosis and diplopia due to extraocular muscle involvement or mass effect, whereas dry eye symptoms are associated with lacrimal gland lesions. Rhabdomyosarcoma (RMS) is the most common primary orbital malignancy in children that invades the base of the skull from the orbit. The most common primary malignant orbital tumors in adults are the lymphoproliferative lesions of the orbit and adnexa, which accounts for up to 20% of all orbital masses and malignant lymphoma is common in above 60 years. Orbital lymphomas are mostly anterior and extraconal lesions. Hemangiopericytomas are rare encapsulated tumors with a propensity towards the superior orbit. They are often more aggressive and can spread through the orbit and intracranially. Optic pathway gliomas (OPG) are the most frequent tumors of the optic nerve. In most individuals, proptosis is the primary symptom unless it is localised posterior to the optic chiasma. Computed tomography (CT), magnetic resonance imaging (MRI), or both is regards as the gold standard for imaging of ocular and orbital malignancy. CT is useful for visualization of bony structures and identifying tumor erosion or hyperostosis of bone, while MRI is advised for imaging soft tissue structures and apical tumors. Conservative management with clinical and radiographic studies is often chosen unless patients have progressive optic nerve dysfunction, visual loss, proptosis, progressive visual field changes, or evidence of hypothalamic invasion. Management of progressive disease typically consists of chemotherapy, in select cases, radiotherapy, as supplementary or alternative therapy to surgery. In this paper, we reviewed several articles on orbital and ocular cancers and summarized.

1. Introduction

Eyes are the most crucial sensory organ in a living being. Eye malignancies are not only develop in eye ball, but also develop in orbit, adenexa, optic nerve etc. Although benign orbital tumors account for two-thirds of cases, but they can lead to a significant reduction of the quality of life. [1,2]

Primary intraocular lymphoma and ocular adnexal lymphoma are the two categories under which eye lymphomas fall. Primary intraocular lymphoma that starts inside the eyes and Ocular adnexal lymphoma, which is incredibly rare and originates in the tissue around the eyes. The most frequent malignancy of the orbit in adults is orbital lymphoma. Most of them are extraconal and anterior lesions. Mucosa-associated lymphoid tissue lymphoma (MALT), follicular lymphoma, large B-cell and mantle cell lymphomas, T-cell lymphomas, mycosis fungoides, Hodgkin lymphomas, etc. are few common types of orbital lymphomas. Lacrimal gland lymphoma typically manifests as lesions in the upper outer corner of the eye. Adenoid cystic carcinoma is an uncommon type of malignant tumor that can spread and endanger life. Squamous carcinoma or mucoepidermoid carcinoma, or adenocarcinoma is other

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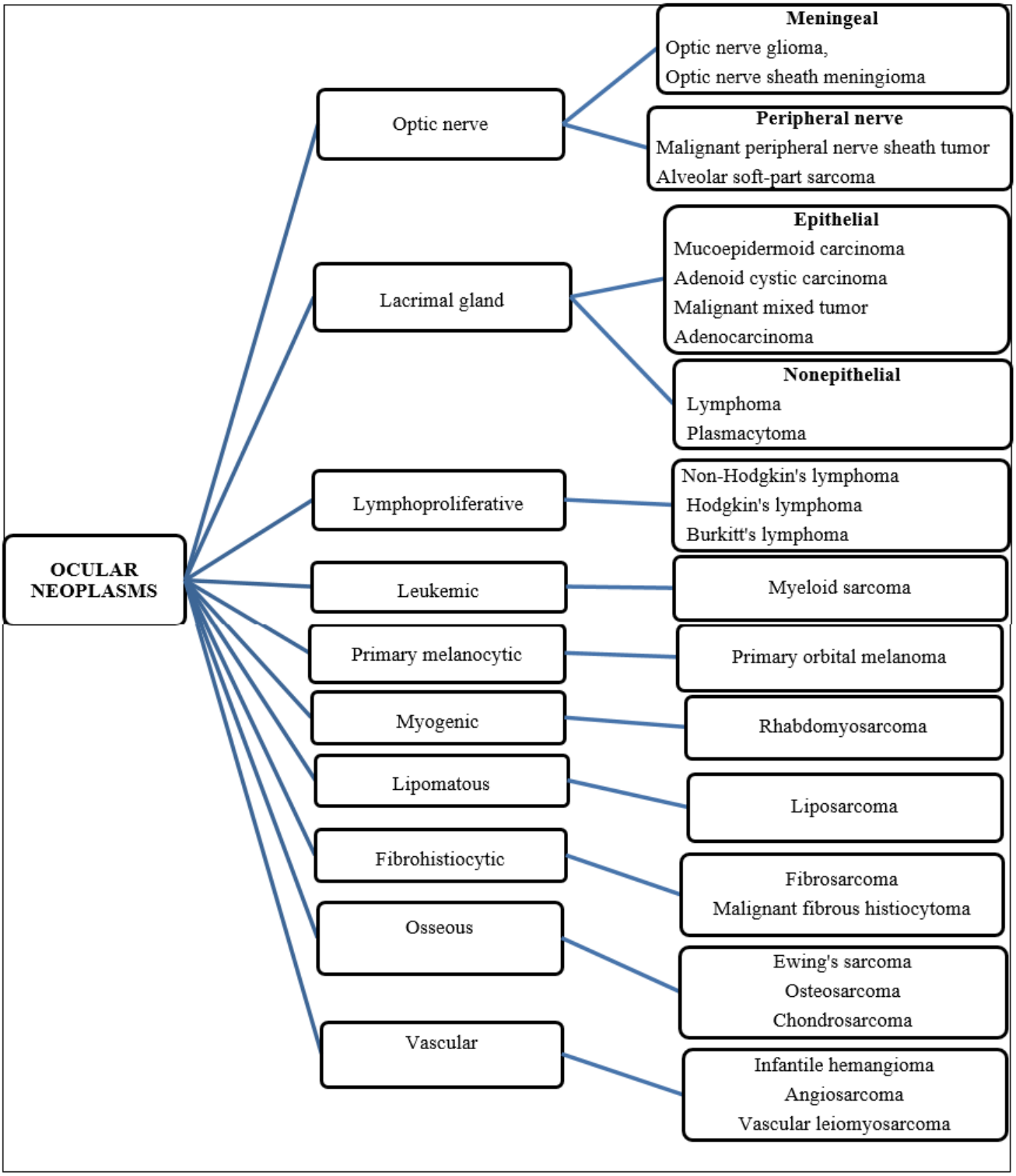

Fig. 1: Ocular neoplasms

forms of lacrimal gland carcinomas. Squamous cell cancer frequently affects the inner conjunctiva of the eyelids or the conjunctiva surrounding the cornea. There are two types of lacrimal gland tumors: epithelial and nonepithelial tumours.

The orbital sarcoma known as rhabdomyosarcoma (RMS) develops from fatty or muscular tissue. RMS is most typical primary malignant orbital tumor with well-defined, atypical, homogenous lesion inside the orbital soft tissues, including muscle, tendon, and nerves. Extraocular motility impairment, optic neuropathy, choroid folds, and optic disc edema are frequent findings.[3] It frequently manifests in the first or second decade of life in youngsters, where it is highly common.[4] It is divided into subtypes that are alveolar, embryonal, pleomorphic, and botryoid. The most prevalent subtype is embryonal rhabdomyosarcoma.[3] On larger and more aggressive lesions, bone erosion and destruction are seen.[5] The most typical quadrant for RMS is the superonasal orbit, which is also frequently extraconal and in the anterior or middle orbit. The most prevalent subtype is embryonic RMS.

Melanoma develops in cells called melanocytes of eye lid skin, eyeball and conjunctival tissue. Conjunctival melanomas can develops in palpebral and bulbar conjunctiva. Squamous cell carcinoma and melanoma can spread to the lymph nodes and other body parts. Uveal melanoma is the most frequent type of in adults.[6] Sebaceous cell carcinoma or meibomian gland carcinoma is a less common type of eyelid cancer than other types. A rare, rapidly growing flesh-colored or purplish growth called a Merkel cell carcinoma begins in the touch receptors of the eyelid.

Retinoblastoma starts in the retina and most common intraocular malignancy in children [Approximately 4% in all Pediatric tumors] under the age of 5 years. The incidence of retinoblastoma is about one case per 15000–20000 live births. $2/3^{rd}$ of Retinoblastoma develops unilaterally and remaining $1/3^{rd}$ is bilateral. On examination white reflection, strabismus, painless impaired visual acuity. Retinal blastoma is the most frequent intraocular malignancy in children under the age of five, and accounts for about 4% of all pediatric cancers. One case of retinoblastoma occurs every 15000–20000 live births, on average. Retinoblastoma develops bilaterally in 1/3 of cases and unilaterally in 2/3 of cases. On examination, white reflection, strabismus, and history of painless vision impairment are noticed.

Neuroblastoma is the most frequent extracranial solid tumor that develops in children from neural crest cells. Clinical features include unilateral or bilateral proptosis, periorbital or eyelid ecchymosis, periorbital edema, hemorrhage, strabismus, reduced ocular motility, ptosis, atrophy of the optic head, and ocular mobility disruption. Clinical feature include periorbital swelling, hemorrhage, strabismus, restricted ocular motility, ptosis, atrophy of the optic head, and ocular mobility disturbance and most common is unilateral or bilateral proptosis and periorbital or eyelid ecchymosis.[7] Malignant optic nerve gliomas are uncommon and often develop in middle age.

Malignant optic nerve gliomas are classified as anaplastic astrocytoma or glioblastoma multiforme.[8] Malignant Optic Nerve Glioma is typically affects the optic chiasm, which can cause sudden loss of vision. Afferent pupillary dysfunction, edema, or optic nerve atrophy are other characteristics. Unfortunately, the prognosis for malignant optic nerve glioma is dismal and the tumor typically kills the patient within months, even with multimodal therapy.[9]

Optic pathway gliomas are the most prevalent intrinsic optic nerve tumors of the optic nerve and which can develop anywhere in the optic pathway. Loss of vision, proptosis, relative afferent pupillary defect, diminished color vision, pupil dysfunction, and progressive field defect are the symptoms that are the presenting symptom. During an ophthalmoscopic examination, the optic disc looks like pallor, atrophied, and edematous. Chiasmal lesions may show deficiencies in the visual field.[8,10] If the tumor arises within the hypothalamus, endocrinopathies like accelerated growth and precocious puberty may manifest.[11] Plexiform neurofibroma (PNF) usually involves sensory nerves in the orbit or eyelid. Eyelid Plexiform neurofibromas are present in S shape due to fat accumulation and thickening.[12,13] Clinically, it can causes to amblyopia, anisometropia, and strabismus.[14] Leukemia is the most common unilateral malignancy of childhood manifest in between 8 to 9 years old. Pain, eyelid swelling, ecchymosis, diplopia, and proptosis can be brought on by a rapidly growing orbital mass caused by the implantation of leukemic cells in the orbit.[12,15] Intraocular leukemia presents with iris masses, Anterior chamber hyphemas, pseudohypopyons, central scotomas. Leukemic retinopathy presents with flame shaped hemorrhages in retina, perivascular infiltrations, microinfarctions, serous retinal detachments at the macula. Dermoids and epidermoid cysts are orbital cystic tumors that are non-tender, slow-growing, and slightly fluctuant in nature.

2. Diagnosis

Clinical signs and symptoms are the first step in the tumor diagnosis process and Surgical biopsies, orbital and ocular imaging such as CT, MRI, ACC, other PEML, tumor debulking, etc. are other objective measures for malignant lesions. Advanced MR techniques have been used to further evaluate, such as Magnetic Resonance Diffusion Tensor Imaging (MRDTI), Diffusion-Weighted Imaging (DWI), and Dynamic Contrast Enhancement (DCE). Immuno - histochemistry, flow cytometry, cytokine, and molecular analysis are other diagnostic methods. Sentinel lymph node biopsy is performed to look for microscopic signs of the larger sebaceous carcinomas spread.

3. Treatment

Treatment may involve radiotherapy, chemotherapy or a combination of these treatments. Local disease can be successfully treated with radiotherapy, whereas systemic disease is often treated with monoclonal antibody immunotherapy alone (i.e., rituximab) or in combination with chemotherapy.[16] Wide excision, complete surgical excision, orbital exenteration with removal of afflicted walls followed by adjuvant radiation, globe-sparing surgery with adjuvant chemotherapy or radiation, complete resection, or maximal debulking are a few of the approaches that have been used. Some cancers are surgically managed by orbital exenteration, which involves removing the afflicted walls and is followed by adjuvant radiation. Advances in endoscopy, microsurgical techniques, and intraoperative image guidance are further helps. Additionally, the advent of multimodal therapy has significantly increased survival rates for many malignancies.[4,17] In leukemia, systemic chemotherapy and bone marrow transplantation are also used for better results.

4. Conclusion

In this article, different types of orbital, ocular and adenexa malignancies are presented. Less harmful diagnostic tools, surgical approaches have to be developed. Early detection of malignancy with proper clinical evaluation, selection of specific treatment modality will be helpful to prevent the further damage.

5. Ethical Approval

I certify say that this kind of manuscript does not require ethical approval by the Ethical Committee of our institution.

6. Conflict of Interest

Author has no conflict of interest to declare.

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