

Orbital Involvement in Primary Paranasal Sinus Space Occupying Lesions

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Commentary

Orbit is an innocent bystander that is afflicted by lesions of the Paranasal Sinuses (PNS). Orbits share 3 walls with the sinuses, frontal sinuses on the supero-medial aspects of the orbits, ethmoid sinuses medially and maxillary sinuses inferiorly. Between the ethmoids and the orbits, there is lamina papyraea which is a very thin bone. Between the maxillary sinus and the floor of the orbit, the bone is weakened by the passage of the infraorbital nerve. The periorbita or the periosteum of the orbit is a tough structure that resists invasion by tumors. Once the tumor transgresses this robust layer, it spreads to the orbital spaces easily due to lack of barriers that check tumor spread. There is also rich vascular communication between the sinuses and the orbit, which acts as a conduit for spread of infections and tumors. The last but not the least is the communication between the nasal cavity and the orbit through the Nasolacrimal Duct (NLD).

The most common involvement of the orbits is in acute and chronic infections of the sinuses. This commentary is restricted only to tumors and other expansile lesions of the sinuses.

Rootman et., al described the orbital involvement in PNS space occupying lesions (SOL) as 5% of all orbital tumors [1]. More than 50% of the patients with sinus and nasal tumors have the signs and symptoms related to the eye or orbit [2]. The most common presenting symptoms of PNS SOL with orbital invasion are non-axial proptosis, diplopia, visual loss, nasal obstruction and/or discharge. Sometimes, the ophthalmic signs and symptoms may be the first presenting features of PNS SOL [3]. Visual disturbance can be the first symptom in malignant lesions of the sinuses and it happens when the orbital apex is infiltrated, and this may indicate an advanced stage of the tumor causing high morbidity and early mortality.

Apart from the clinical cues, the mainstay in diagnosis of PNS SOL is imaging. Advanced imaging techniques and the expanded use of CT scanning and MR imaging allow clinicians to view with increasing detail the normal and pathologic structures of the orbit. Detailed radiographic images delineate the extent of the tumor invasion of the orbit and show the nature of the pathologic process, thereby enabling the clinician to narrow down the differential diagnoses and build an appropriate treatment plan [4]. Obtaining scans with image guidance protocol helps the surgeons during surgical removal ensuring safety and minimizing injury to vital structures such as optic nerve, brain etc.

CT scans show bony remodelling, bony destruction, and invasion of intracranial structures in addition to soft tissue involvement of the orbit. MRI shows soft tissue involvement clearly, indispensable in the planning of treatment and post therapy evaluation of sinus tumors [5].

In addition to imaging, endoscopic visualization of the lesion, where possible, enables for a quick biopsy leading to early diagnosis.

Among the common benign expansile lesions of the sinuses affecting the orbit are the mucoceles of the sinuses. The symptoms and signs are basically due to pressure and encroachment of the orbit. Mucoceles are cystic lesions, the contents of which are usually sterile and mucoid. They can affect any age group and are usually caused by obstruction to the sinus ostium by trauma, tumor or inflammation with expansion of the sinuses and remodeling. Patients present with facial deformity, palpable mass, proptosis usually non-axial in addition to nasal symptoms. The vision is rarely affected unless the lesion is very large. Treatment is by endoscopic sinus surgery, the mucocele is drained and the obstruction removed by enlarging the sinus ostium. Occasionally there is recurrence after surgery.

Fibro-osseous lesions comprise fibrous dysplasia, ossifying fibroma, and osteoma. They usually affect cranio-facial bones, start in childhood, progress through puberty, and stabilize in adulthood. Mono-ostotic lesions are seen in 30% and poly-ostotic in 70%. They cause facial deformity and non-axial proptosis. Fibrous dysplasia shows ground glass appearance in CT scan. Ossifying fibroma shows sharply circumscribed lesion with an egg-shell rim on the CT scan, and is usually mono-ostotic. Histologically has islands of osteoid tissue rimmed by osteoblasts. Osteoma is of 3 histological types: Ivory, spongiform and mixed type. All the above lesions can co-exist with sinus mucocele if they obstruct sinus openings.

The other benign tumor that is worth mentioning is inverted papilloma. The inverted papilloma is locally invasive. Recurrence is common especially if incompletely removed. Maxillary sinus and lateral nasal wall are the most commonly involved sites. It usually presents as NLD obstruction to the ophthalmologist. It is often clinically missed and seen quite unexpectedly in the lacrimal sac during dacryocystorhinostomy. Complete sac excision with medial maxillectomy along with NLD is necessary. Malignant transformation to squamous cell carcinoma occurs occasionally.

Malignant epithelial tumors cause nasal symptoms such as epistaxis, but in 2/3rd of the cases the first presentation is to the ophthalmologist. The common ophthalmic symptoms include diplopia, decrease in vision, pain and paresthesia due to perineural spread. Proptosis may or may not be a predominant symptom. Spread to orbital apex is usually responsible for loss of vision.

Squamous cell carcinoma has the worst prognosis among the malignant epithelial tumors. Radical surgery involving removal of sinuses, skull base and exenteration is necessary to save the life of the patient. Palliative radiation and chemotherapy are used in advanced cases. The other malignant tumors include adenocarcinoma, and adenoidcystic carcinoma.

Neuroendocrine tumors such as esthesioblastoma are rare, but can affect the orbit and the intracranial space in advanced stages. The management is surgical excision and radiation. The eyeball and orbital

contents may be preserved. Chemotherapy is considered if there is metastasis.

Other rare malignant tumors include malignant solitary fibrous tumor, rhabdomyosarcoma and malignant nerve sheath tumor.

Lymphoma is another primary malignant tumor that affects the sinuses and extends to the orbit. It can be either a B-cell or a T-cell lymphoma. Natural Killer cell lymphoma occurring in mid-line can be aggressive. After staging, these tumors are treated with radio/chemotherapy. CNS involvement would necessitate intra-thecal chemotherapy [2].

Whenever the patient presents with diplopia or ophthalmoplegia or pain in a PNS SOL lesion, the odds ratio for this lesion being malignant is 3.67, 3 and 2 respectively.

In conclusion, a PNS mass lesion should always be considered in the differential diagnosis when a patient presents with diplopia and ophthalmoplegia in order to prevent delays in diagnosis and management. Any non-axial proptosis, especially in the down and out direction, likewise, should promptly be investigated for a PNS SOL.

These mass lesions should be effectively managed by multidisciplinary teams involving ENT/head and neck surgeons along with ophthalmologists, neurosurgeons and medical/radiation oncologists as necessary.

References

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