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Orbital metastatic disease is rare. But with increasing longevity, there has been an increase in the incidence of metastatic disease involving the ocular adnexa in recent years. Carcinomas are more common than melanomas and sarcomas; and adults, aged 40 years and above, are more commonly affected than children. Bilateral orbital metastasis is rare, except in breast carcinoma in adults, and neuroblastoma in children. Common locations of origin include the breast, lung, prostate, gastrointestinal system, and, in Caucasians, cutaneous melanomas. Melanoma and breast cancer have a strong tendency to localize in the orbital fat and muscles, while prostate cancer is well known to metastasize to the bone (hyperostotic lesions).

The metastatic lesions (such as those from breast cancer) may appear months or years after the diagnosis and management of systemic disease. They appear well before the discovery of the primary lesion in a quarter of cases, especially with lung, gastrointestinal, thyroid, and renal carcinomas. When metastasis occurs before the discovery of the primary lesion, exhaustive medical investigations would be necessary to find the primary site including a thorough history taking, a high index of suspicion, and a multidisciplinary approach.

In recent years, there have been advances that make it easier to find the primary tumor. An increase in carcinoembryonic antigen (CEA) is non-specific but, nevertheless, can alert one to the presence of metastasis. Similarly, an increase in prostate-specific antigen in a patient with prostate cancer can suggest metastasis. Some of the other tests include human chorionic gonadotropin (HCG) in seminoma and 5-hydroxyindoleacetic acid (5-HIAA) in urine in carcinoid tumors.

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Advanced imaging techniques, fine-needle aspiration biopsies (FNAB), as well as advanced serological and molecular studies may further aid in the identification of the primary tumor. Despite these advances, the primary site remains unknown in about 2% of oncological referrals. Identification of primary site does not alter the management or the prognosis in a majority of cases except lymphomas.

Patients with orbital metastases usually present with rapid onset of diplopia, mild proptosis, displacement of the globe, ptosis of the eyelid, eyelid swelling, pain and redness of the eye with chemosis, and more importantly a decrease in vision. Rarely, the patients develop enophthalmos rather than proptosis. Misdiagnosis and lengthy delay in diagnosis are common in metastatic cancer to the orbit. The signs are often confused with orbital cellulitis, myositis, and idiopathic orbital inflammatory syndrome (pseudotumor). The other differential diagnoses that need to be considered are acute active thyroid orbitopathy, carotico-cavernous fistula, orbital apex (Tolosa-Hunt) syndrome, primary lacrimal gland tumors, and lymphoproliferative disorders. The ophthalmologist, the general physician, and the oncologist should therefore maintain a high level of suspicion in patients with a known history of malignancy, presenting with an acute or subacute ocular, orbital, or adnexal "inflammatory mass" lesion.

Radiological features include diffuse intraconal infiltration, enlargement of a single extraocular muscle, extraconal infiltrative mass, or bony destruction with adjacent soft tissue involvement. Orbital metastasis from breast cancer tends to be diffuse and irregular, often growing along the rectus muscles and fascial planes. On the other hand, orbital metastasis from carcinoid tumor, renal cell carcinoma, and melanoma tends to be more circumscribed in the early stages, before they spread inside the orbit.

Majority of cases (90%) can be diagnosed with a needle aspiration biopsy. Some cases of scirrhous carcinomas (from the lung or breast) or small apical lesions may not yield sufficient tissue for diagnosis. Where a metastasis is sus-



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pected, an open biopsy with debulking of the tumor may be carried out especially if there is optic neuropathy. FNAB can be performed with the tumor exposed by open surgery. The advantage of doing FNAB is less damaging to normal structures especially when there is no clear demarcation between abnormal and normal tissues. The other advantage of needle biopsy is that it not only helps in cytological diagnosis, but also provides a "cell block" for a reasonable histological diagnosis.

Patients with widespread metastasis are treated with chemotherapy. In carcinoma of the breast and prostate, hormone therapy may be considered. Radioiodine in thyroid carcinoma and immunotherapy in melanoma may be helpful.

Isolated orbital metastasis or progression of orbital disease despite chemotherapy can be treated with radiotherapy or proton beam therapy in cases of melanoma. The mean survival of patients with orbital metastasis is about 1.3–2 years, and the survival is not different in patients with and without a known primary. There is slightly a better prognosis in patients with orbital metastasis from breast carcinoma. In some metastatic tumors, such as renal carcinoma, long-term survival after removal of a solitary orbital metastasis has been reported.

In conclusion, although the prognosis is poor, orbital metastases can be treated to give patients a better quality of life.

## Further Reading

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