Orbital Metastases: Are we underestimating the problem

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Orbital metastases represent a small but increasing percentage of all orbital tumors, reported in different case studies and series to have an incidence of 1% to 13%. Breast cancer is by far the most common primary site, accounting for 28.5%–58.8% of cases of orbital metastases, followed by lung, prostate, gastrointestinal, kidney and skin (melanoma) cancers.[1-2] Unilateral disease is the usual presentation while intra-orbital anatomical distribution involves predominantly the lateral and superior quadrants.

Due to a tissue-specific preference of breast cancer to extra-ocular muscle and surrounding orbital fat, diplopia resulting from mobility deficits is a prevalent symptom. Other common symptoms and signs include proptosis, eyelid swelling or visible mass, pain, ptosis and blurred vision caused by infiltration or compression. Enophthalmos is a less common but distinctive sign of orbital infiltration by scirrhous breast adenocarcinoma.[2-3]

Definite diagnosis of an orbital lesion requires an orbital biopsy, either by fine needle aspiration method (FNA) or an open biopsy. However, in patients with known metastatic cancer, the latter may be avoided if there is a strong clinical and imaging suspicion for metastatic disease.

Metastatic lesions to the orbit usually present as irregularly shaped masses on non-contrast CT scan which are isodense to muscle while with contrast injection, they show slight enhancement.

On MRI, metastatic disease is usually hypointense to fat on T1-weighted images (T1WI) and hyperintense to fat on T2WI. This appearance may help to differentiate it from an orbital pseudotumor, which is usually isointense to fat on T2WI. When hyperintense lesions are seen on T1WI, a very vascular metastasis (e.g. thyroid, renal) or melanoma metastasis should be suspected.[4] The combined involvement of the orbit and adjacent structures, such as the paranasal sinuses, is a rare condition revealed by imaging studies.[5]

In addition to metastasis, differential diagnosis of an orbital process should include inflammatory lesions, benign tumors (such as hemangiomas) and lymphoproliferative disorders. Idiopathic orbital inflammatory syndrome (IOIS or orbital pseudotumor), sarcoidosis and Wegener granulomatosis are inflammatory conditions that may present in similar manners. Given that inflammatory signs are common in orbital metastases from breast cancer, they could be misdiagnosed as thyroid orbitopathy, cellulitis, myositis, scleritis or endophthalmitis. The distinguishing feature of orbital metastases is a rapid onset and progressive course with combined motor and sensory deficits, non-responding to antibiotics or steroids.[6]

Treatment for orbital metastases is inevitably palliative, given that hematogenous spread of cancer to the orbit is a sign of systemic disease and involvement of other sites. Surgical intervention is generally not recommended, unless it is performed for diagnostic purpose (biopsy) in patients with no previous history of cancer or as palliation (tumor resection or enucleation) in cases of unmanageable local symptoms.[7]

The main treatment option is radiotherapy with high rates (60%–80%) of clinical improvement of local symptoms and vision. External-beam irradiation is the most common and accessible modality with a total dose of 20–40 Gy delivered in fractions over 1–2 weeks.[1-2] Stereotactic Radiation Therapy (SRT) has recently evolved as an alternative modality in an effort to apply high doses of radiation to a well-defined volume with steep dose gradients outside the target volume.[7]

Prognosis of patients with metastatic orbital tumors is rather poor with a median survival ranging from 22 to 31 months for breast cancer. Nevertheless, rare cases of long-term survival after the diagnosis of breast cancer presenting as an orbital mass have been reported.[8]
References