Metastasis to orbit: A case series from tertiary eye center in Western India

Viral M Bhanvadia¹, Ravija Kathiara², Ami M Shah³, Swati Devanhalli⁴

From ¹Assistant Professor, Department of Pathology, ²Assistant Professor, Department of Ophthalmology, ³Associate Professor, Department of Pathology, ⁴Professor and Director, Department of Ophthalmology, M and J. Regional Institute of Ophthalmology, Ahmedabad, Gujarat, India

ABSTRACT

This study aims to understand the pattern of the metastatic tumors to orbit in a tertiary eye center. Metastasis to orbit is extremely rare and challenging to diagnose and often mimics with primary orbital tumor. Due to the tremendous recent advances in cancer therapy, surveillance, ancillary histopathology, and imaging techniques for primary tumor diagnosis, the longevity of patients increases, leading to more chances of detection of orbital metastasis. We retrospectively studied and reviewed a case series of five patients with orbital metastasis from various systemic malignancies from 2018 to 2023. Out of five patients, three patients were from metastasis from breast carcinoma, one from neuroblastoma, and monophasic synovial sarcoma each, found a median age of 49 years (range=3–70 years). The most common symptoms and sign were painful proptosis and restriction of ocular motility, respectively.

Key words: Metastasis to orbit, Breast carcinoma, Neuroblastoma, Monophasic synovial sarcoma.

Orbital metastasis is an uncommon and varied clinical presentation that depends on the underlying tumor. Most of the time, diagnosis is unexpected and delayed affecting subsequent definitive management [1]. The incidence of orbital metastasis ranges from 1% to 13% among reported series [1,2]. The clinical symptoms of orbital metastasis, such as restriction in ocular mobility, proptosis, diplopia, pain, and periorbital swelling, are similar to those of primary orbital tumors, such as idiopathic pseudotumor and IgG4-related orbital disease or any other soft-tissue tumor of orbit.

The purpose of this case series is to review and report metastasis to the orbit, demographic and clinical features, diagnostic challenges, and management of orbital metastasis in relation to the primary tumors at the tertiary eye center of Western India.

CASE SERIES

Five patients diagnosed with orbital metastasis at the M and J Western Regional Institute of Ophthalmology, Civil Hospital, Ahmedabad, Gujarat, tertiary eye center in Western India in a 5-year period (2018–2022) were included in this case series. Approval was taken from the Institutional Ethics Committee. General data included the patient’s age, sex, history of primary cancer, clinical presentation, and history of prior treatment of primary tumor, if taken were recorded.

| Tumor data included location in the orbit, laterality, histopathology, immunohistochemistry (IHC) diagnosis, relevant radiological investigation, treatment modality, and outcome, whenever available were evaluated and mentioned in Table 1.

DISCUSSION

It is presumed that 2–5% of patients with systemic malignancies develop orbital metastases [3]. While 85% of patients with orbital involvement carried a known cancer diagnosis, in the remaining 15% metastasis was the initial manifestation of malignancy [1]. Magliozzi et al. documented 93 patients over a 38-year period and found a median age of 51 years (range=1–88 years) [3]. We found a median age of 49 years (range=3–70 years) in our case series.

Breast cancer metastasizes to the orbit more frequently, representing 29–53% of cases [4]. The time from diagnosis of breast cancer to the onset of orbital metastases is usually prolonged, with an average of 2–8.5 years [5]. In our case series, a 51-year-old female who had undergone modified radical mastectomy (MRM) post-neoadjuvant chemotherapy developed metastasis after eight years of treatment. In contrast to other primaries, bilateral metastases can be seen in 15–20% of breast carcinoma cases. Overall, orbit remains a rare site even for breast cancer metastasis. Orbital metastasis may present with symptoms such as proptosis, double vision, decreased visual acuity, pain,
chemosis, ptosis, or orbital bony involvement [6]. Three out of five cases in our case series were breast cancer patients having a median age was 51 years and painful swelling along with proptosis was their common presentation. Out of three cases of metastatic breast cancer to orbit, one case was bilateral orbital involvement associated with many metastatic skin nodules corresponding to invasive lobular carcinoma of the breast, which has the propensity of hematogeneous metastasis to the body [7].
The other two cases were of invasive duct carcinoma of the breast (Fig. 1a). The treatment can be adjusted depending on the clinical presentation, the extent of the metastatic disease, and the immunohistochemical status of the primary tumor. The prognosis of breast cancer with an orbital involvement has been reported to be very poor with survival ranges from 1 to 116 months with a mean of 31 months [7]. All three cases were lost to follow-up in our series.

Neuroblastomas are the most frequently identified extracranial tumors that affect children under the age of 4 years old, with an incidence of approximately 1–3 in 100,000 cases [8]. Orbital neuroblastoma metastasis is relatively rare and is associated with poor prognosis. Ahmed et al., studied a series of neuroblastoma cases, of which, the most common presentation was proptosis, restricted eye movement, and 12.5% were having an orbital metastasis, giving a male-to-female ratio of 2:1. The average age of presentation was 29.8 months (15–69 months) [9]. Orbital involvement falls in the high-risk category and therefore aggressive multi-agent chemotherapy consisting of very high doses is employed [9]. Neuroblastoma cases with initial symptoms of orbital involvement are rare, accounting for approximately 8% of all neuroblastomas. Our case of a 3-year-old male child presented with proptosis and subsequently detected adrenal neuroblastoma (Fig. 1b). The role of IHC and systemic workup is necessary to diagnose and rule out other small round cell neoplasms in the pediatric age group in this region. On the last follow-up, the child had taken the first dose of chemotherapy.

Synovial sarcoma (SS) is a rare malignant soft-tissue neoplasm that accounts for approximately 5%–10% of all soft-tissue tumors, often presents in children and young adults, and primarily occurs in the para-articular regions of the extremities [10]. The orbit is an extremely rare location for metastatic SS. Our study has a case of monophasic SS with metastasis to orbit, presenting as proptosis (Fig. 1c). To the best of our knowledge, we have not found any metastatic monophasic SS to orbit in the English literature. IHC is required for diagnosis and to rule out other primary spindle cell neoplasms arising from the orbit. The patient did not come for a follow-up after the orbital biopsy.

Although our study has a limited period and a small number of cases due to the rarity of orbital metastasis, a larger study from India is required for a better understanding of the epidemiology, the interval between primary to metastasis, the mode of spread of tumor, and response to treatment to the primary tumor. In literature, the majority of orbital metastases are carcinomas; the frequency of different sites of primary disease varies between different reports mainly due to study period, number of cases, and geographical variation in certain tumors. The most common is breast carcinoma (28–58.5%), followed by lung, melanoma, and liver cancer [4].

CONCLUSION

In concordance with the literature, our study also shows breast cancer commonly metastasizes to the orbit. While neuroblastoma should be considered as differentials in a small round cell tumor of a child presenting as orbital mass, a necessary systemic workup should be done to detect primary. Appropriate IHC and molecular study will be necessary to detect this extremely rare orbital metastasis of monophasic SS.
The prognosis of orbital metastasis remains poor due to the higher stage of disease and systemic metastasis. Metastasis to orbit is rare, so, ophthalmologists must remain vigilant. A higher level of suspicion, systemic workup, and IHC for precise diagnosis and appropriate timely management are important to salvage vision and life.

REFERENCES


Funding: Nil; Conflicts of interest: Nil.

How to cite this article: Bhanvadia VM, Kathiara R, Shah AM, Devanhalli S. Metastasis to orbit: A case series from tertiary eye center in Western India. Indian J Case Reports. 2023; December 01 [Epub ahead of print].