

Case Study

Double malignancy manifesting as unilateral proptosis

Abstract:

Introduction: Unilateral proptosis is usually a diagnostic challenge to an ophthalmologist. . Mass lesions constitute a common cause of unilateral proptosis in elderly individuals in which lymphoproliferative lesions account for 11% of cases and secondaries make up to 2- 9% of cases. Thorough review of literature showed various tumours metastasizing to the orbit but simultaneous presence of two different tumours are extremely uncommon and we did not get any reference in literature search.. The authors wish to report the same to highlight the importance of systemic examination in unilateral proptosis in elderly.

Case Presentation: A 52 year old male presented with complaints of painless, gradually progressing prominence of right eye for 8 months. There was 15 degrees exotropia of the right eye.. Right eye examination revealed an axial proptosis of 6mm by Hertel's exophthalmometry. It was a non tender and a non pulsatile mass. CT scan of the orbit revealed right eye proptosis with enhancing soft tissue attenuation lesion involving extraconal and intraconal compartments in superolateral aspect of the right orbit abutting proximal portion of the right optic nerve. Biopsy from the right lacrimal gland revealed stratified squamous epithelium with subepithelium infiltrated by monomorphic population of small lymphoid cells and few immunoblasts suggesting a low grade lymphoproliferative lesion. The lymphoid cells were negative for Cyclin D1 and CD3 while the rest of the markers were positive confirming the diagnosis of small lymphocytic lymphoma. Ultrasound abdomen performed for assessment of intraabdominal lymph nodes or visceral enlargement showed diffuse thickening of gastric wall with perigastric lymphadenopathy. An endoscopic antral biopsy revealed intestinal type of gastric adenocarcinoma on histopathological examination

Conclusion: Orbital metastases is a significant and under-recognized clinical problem for practicing ophthalmologists and highlights the importance of high index of clinical suspicion and through systemic examination in the evaluation of a case of unilateral proptosis in the elderly.

Introduction:

Unilateral proptosis is usually a diagnostic challenge to an ophthalmologist. The causes vary to a large extent with Graves' ophthalmopathy contributing to 47% and mass lesions making up 53% of the total cases. Mass lesions constitute a common cause of unilateral proptosis in elderly individuals in which lymphoproliferative lesions account for 11% of cases and secondaries make up to 2- 9% of cases [1,2,3]. Most tumors metastasizing to the eye are carcinomas, although sarcomas, lymphoproliferative malignancies, and melanomas have also been reported. The reported carcinomas metastasizing to the orbit have primaries from breast , lung, gastrointestinal, genitourinary tract, thyroid, adrenal glands, and cutaneous malignant melanoma. [4,5]The presenting signs in patients with metastasis to the orbit include proptosis or a painful, palpable mass and ophthalmoplegia.

Less common signs of metastatic orbital tumors include diminution of vision, choroidal folds or an afferent pupillary defect.[6]Thorough review of literature showed various tumours metastasizing to the orbit but simultaneous presence of two different tumours are extremely uncommon and we did not get any reference in literature search.. The authors wish to report the same to highlight the importance of systemic examination in unilateral proptosis in elderly.

CASE REPORT:

A 52 year old male presented with complaints of painless, gradually progressing prominence of right eye for 8 months [Fig.1]. There was history of squinting of right eye since childhood. There was history of significant weight loss over the past 6 months. On examination his best corrected visual acuity in the right eye was 20/120 and in the left eye was 20/40. There was 15 degrees exotropia of the right eye.. Right eye examination revealed an axial proptosis of 6mm by Hertel's exophthalmometry. It was a non tender and a non pulsatile mass. Auscultation did not reveal any bruit over the swelling. Anterior segment examination was within normal limits except for a cataract in lens. Fundus was within normal limits. Left eye examination was within normal limits with a posterior chamber intraocular lens.. Systemic examination was within normal limits.. CT scan of the orbit revealed right eye proptosis with enhancing soft tissue attenuation lesion involving extraconal and intraconal compartments in superolateral aspect of the right orbit abutting proximal portion of the right optic nerve. All extraocular muscles were involved by the lesion and lacrimal gland could not be distinguished separately from the lesion [Fig2]. Biopsy from the right lacrimal gland revealed stratified squamous epithelium with subepithelium infiltrated by monomorphic population of small lymphoid cells and few immunoblasts suggesting a low

grade lymphoproliferative lesion. Immunohistochemistry was performed for panel of small cell Non Hodgkin's lymphoma and the panel comprised of CD3, CD20, CD5, CD 23, and Cyclin D1. The lymphoid cells were negative for Cyclin D1 and CD3 while the rest of the markers were positive confirming the diagnosis of small lymphocytic lymphoma [Fig3]

Meanwhile, the patient was examined thoroughly for other systemic problems. His thyroid function tests revealed a normal thyroid profile. Ultrasound abdomen performed for assessment of intraabdominal lymph nodes or visceral enlargement showed diffuse thickening of gastric wall with perigastric lymphadenopathy suggestive of gastric involvement of Non Hodgkin's lymphoma. An endoscopic antral biopsy revealed intestinal type of gastric adenocarcinoma on histopathological examination [Fig 4]. The patient was advised gastric surgery and chemotherapy for further management..

DISCUSSION:

Mass lesions constitute one of the common causes of unilateral proptosis in the elderly, out of which lymphoproliferative lesions account for 11% and secondaries account for 2-9% of cases. Orbital lymphoma and lymphoma of the orbital adnexae are relatively rare, representing 0.1% of all lymphomas. [7]

Lymphomas of B-cell lineage are more likely to be associated with ocular symptoms and orbital extension than lymphomas of T-cell or NK-cell lineage.[8] Orbital metastases are generally associated with a bad prognosis and most patients succumb to widespread systemic disease within months [9,10,11].

Orbital lymphomas and secondaries to the orbit typically present with proptosis or a palpable mass causing eyelid swelling [12,13]. Other symptoms include restricted extra-ocular movement, diplopia, and pain. The patient in this report had proptosis and lid edema. Decrease in visual acuity is uncommon unless there is optic nerve infiltration or compression caused by the mass [14]. Our patient had decreased vision due to cataract in the eye.

One fourth of patients who present with an orbital metastases do not have prior history of cancer [1,4]. So in this patient provisionally a diagnosis of secondaries to the orbit was considered.

Hence a systemic examination was done to rule out any secondaries infiltrating the orbit with specific attention to lung and gastrointestinal malignancies as these malignancies are common causes of orbital metastasis in males [15]. Systemic examination performed for staging of the lymphoma revealed a gastric mass. Biopsy from both sites confirmed that patient had two unrelated primary neoplasms. Careful review of literature shows that synchronous manifestations of two malignancies are uncommon. We did not find any report of unrelated second malignancy in a patient with unilateral proptosis.

Conclusion

Orbital metastases is a significant and under-recognized clinical problem for practicing ophthalmologists and highlights the importance of high index of clinical suspicion and through systemic examination in the evaluation of a case of unilateral proptosis in the elderly.

Competing interest None to declare

Author's contribution :

Dr Renuka Srinivasan Critical evaluation of the manuscript

Dr. Rajesh Nachiappa Ganesh Pathology workup and its photography.

Dr Praveen Kumar K : Clinical workup

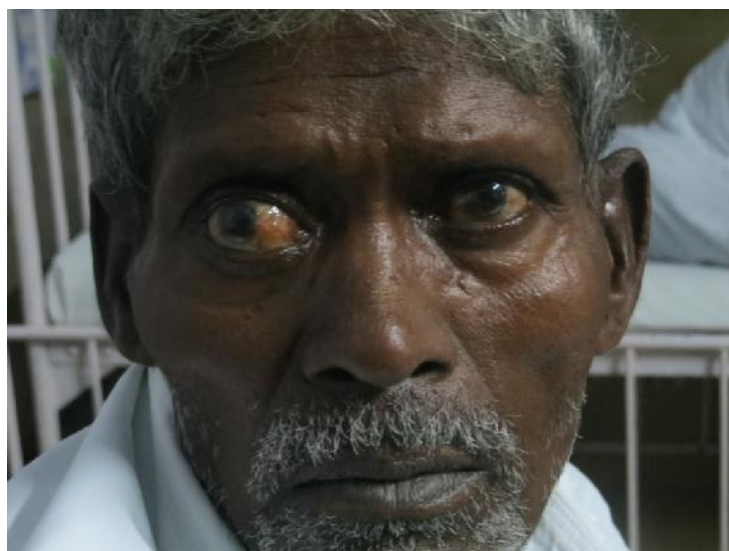
Dr Praveen Kumar S: Laboratory investigation including the imaging studies

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Figure 1



Clinical photograph showing right eye proptosis

Figure 2



CT scan of the orbit revealed right eye proptosis with enhancing soft tissue attenuation lesion involving extraconal and intraconal compartments in superolateral aspect of the right orbit abutting proximal portion of the right optic nerve. All extraocular muscles were involved by the lesion and lacrimal gland could not be distinguished separately from the lesion

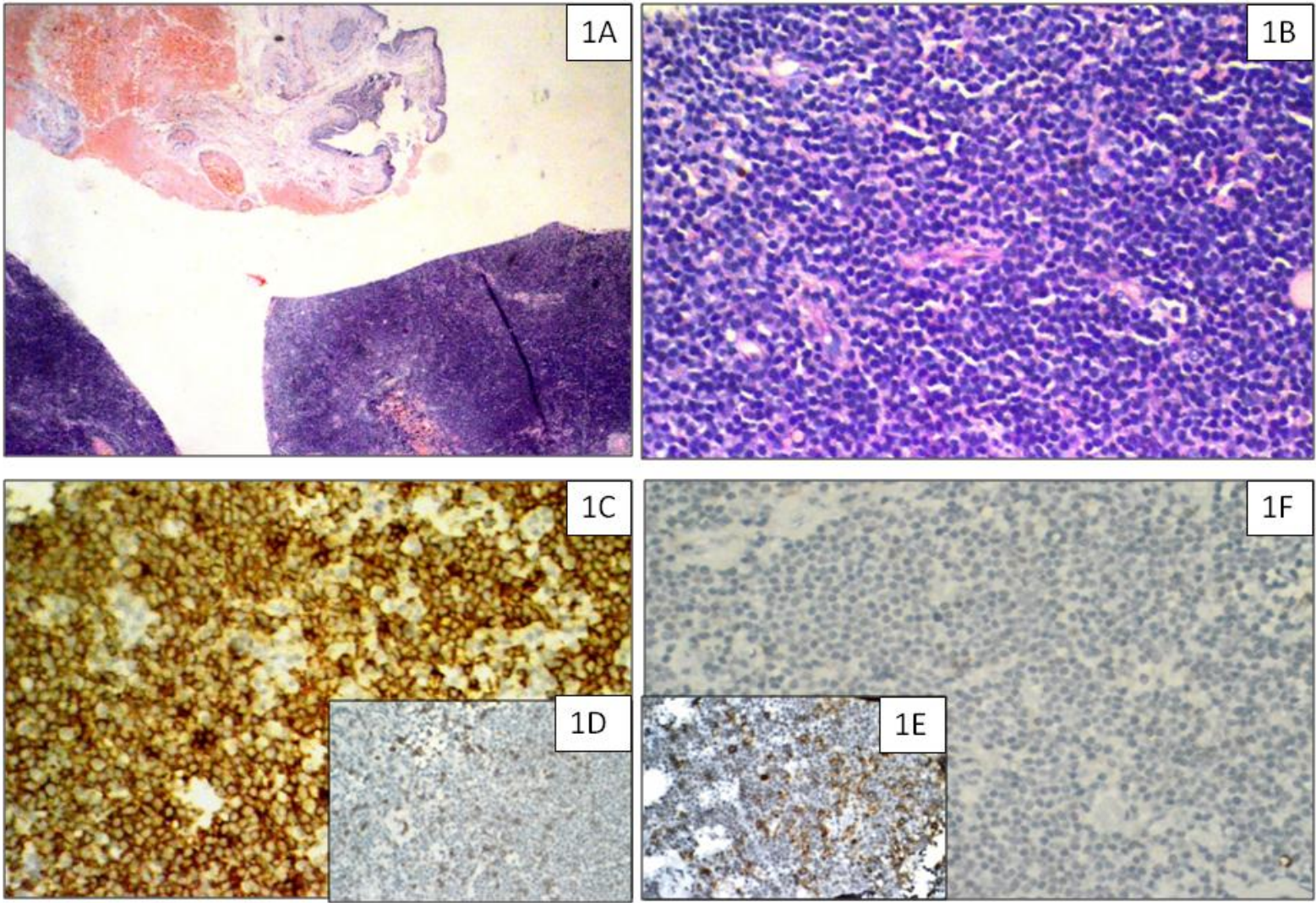
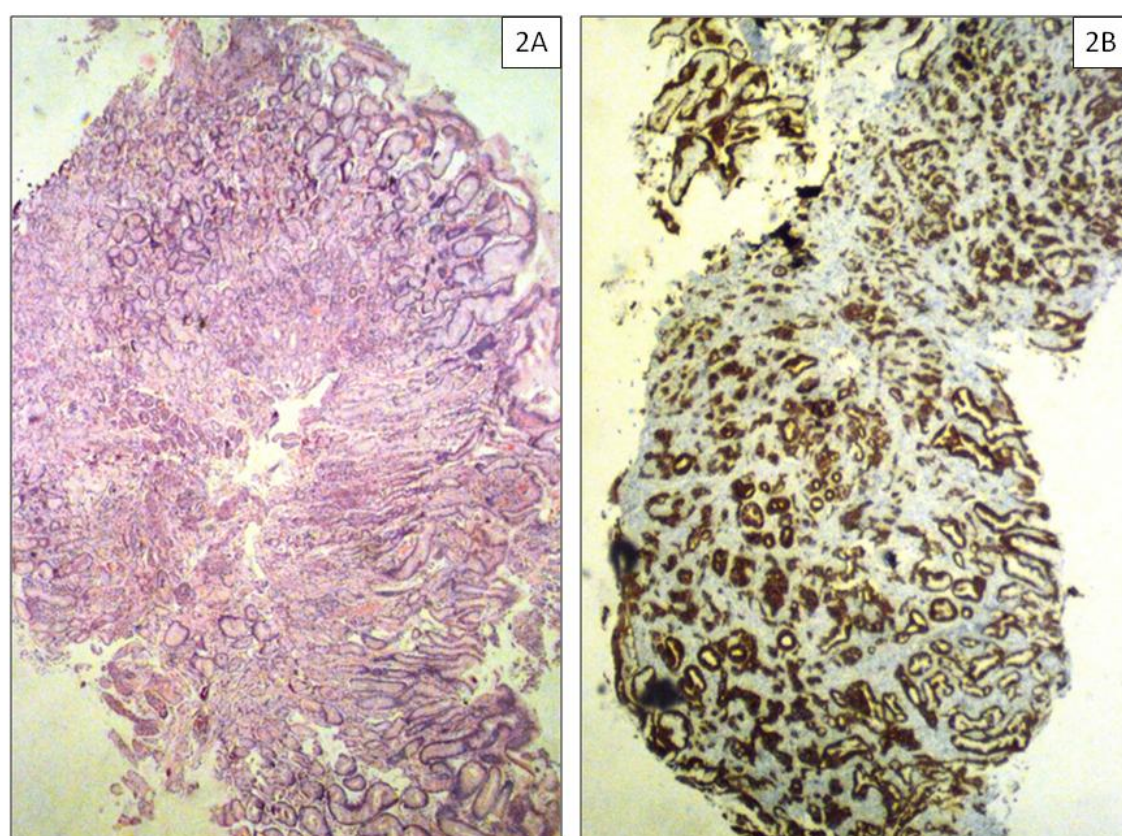


Figure3

Legend

Fig 1A – Section shows fragments of conjunctival mucosa with dense infiltration by sheets of monomorphic small round lymphoid cells, x40, Hematoxylin and Eosin.
Fig 1B – Section shows sheets of monomorphic small round lymphoid cells with inconspicuous cytoplasm and coarse nuclear chromatin, x400, Hematoxylin and Eosin
Fig1C – Section shows strong membranous positivity for CD 20 in lymphoid cells, x400, DAB stain (Immunohistochemistry with DAKO antibody, USA)
Fig 1D – Section shows occasional lymphoid cells expressing CD5, x400, DAB stain (Immunohistochemistry with DAKO antibody, USA)
Fig 1E – Section shows occasional lymphoid cells expressing CD23, x400, DAB stain (Immunohistochemistry with DAKO antibody, USA)
Fig 1F – Section shows negativity for CD3 in lymphoid cells, x400, DAB stain (Immunohistochemistry with DAKO antibody, USA)

Figure 4



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163 Fig 2A – Section from gastric antral mucosa shows infiltration by tumor cells arranged in glandular pattern, x40,
 164 Hematoxylin and Eosin

165 Fig 2B - Section from gastric antral mucosa shows infiltration by tumor cells arranged in glandular pattern, x40, DAB
 166 stain, (Immunohistochemistry with DAKO antibody, USA)

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