Primary Adenocystic Carcinoma of the Nasal Cavity with Ocular Signs of Foster Kennedy Syndrome

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Abstract

Foster Kennedy Syndrome presents with anosmia, ipsilateral optic nerve atrophy, and contralateral optic nerve edema. This unique case is secondary to a rare adenoid cystic carcinoma of the nasopharynx, skull base and sella region.

I. Case History:
   i. 49 yo CM with CC: Vision loss OS x 2 years
   ii. LEE 1980s, unremarkable
   iii. Medical history: BPH, adenoid cystic carcinoma
   iv. Medications: Topiramate, ondansetron, naproxen, folic acid, thiamine, HCTZ
   v. History of intense headaches; followed by neurology and resolves with Topiramate

II. Pertinent findings:
   i. BVA: OD: 20/25; OS: 2/500 EV, central vision HM
      Color vision: 1/13 OD, unable OS
      Pupils: PERRL 3+ LAPD
      EOMS: FROM OU
      External: 1 mm left ptosis, (-) proptosis
   
   ii. Slit Lamp Exam: Unremarkable
      Tonometry: OD, OS: 14 mm Hg
      DFE: Unremarkable, except:
      ONH: OD: 0.10 rd- elevation all quadrants, less prominent T
           OS: 0.10 rd- 1+ diffuse pallor
   
   iii. HVF 30-2 OD/ Kinetic OS
      OD: Small island S to fixation preserved
      OS: Small island SN to fixation
      Fundus Photos
   
   iv. CT Scan: Neoplastic lesion
      MRI: Large nasal cavity mass, extension to sellar, suprasellar, parasellar, cavernous sinus, right petrous apex, and bilateral Meckel’s cave. Severe mass effect on optic chiasm; severe atrophy of pre-chiasmatic left optic nerve. Complete encasement of bilateral cavernous carotids and right petrous carotid. Mass effect on gyri recti and bilateral medial temporal lobes.

III. Differential Diagnosis
   i. Foster Kennedy Syndrome
   ii. Pseudo-Foster Kennedy Syndrome
   iii. Compressive Optic Neuropathy
iv. Infiltrative Optic Neuropathy

IV. Diagnosis and Discussion
We present a unique case of Foster Kennedy Syndrome (FKS) secondary to a rare adenoid cystic carcinoma of the nasopharynx extending into the skull base and sella region. FKS may present in three ways: unilateral direct optic nerve compression with increased intracranial hypertension (ICH), bilateral direct optic nerve compression without ICH, or chronic ICH without direct compression. This particular case involves atrophy of intracranial portion of the left optic nerve secondary to direct compression and disc edema of right optic nerve without clear evidence of true ICH.

V. Treatment, management
Deferred to neurosurgery and oncology for management of ACC. Currently unresectable, patient underwent palliative radiation treatment followed by a repeat MRI to determine if it becomes resectable post radiation.

Bibliography:

VI. Conclusion
FKS is a rare ophthalmic condition consisting of optic nerve pallor followed by disc edema of the contralateral eye, both resulting from an intracranial tumor. The key clinical pearl is that it is pertinent for eye care providers to recognize this syndrome in order to obtain urgent imaging and co-management, although patient prognosis is often poor. A unique take away point is that routine ocular examinations are important for ocular health, as well as systemic health. Early detection is critical in these cases; treatment may have been more favorable and prognosis improved for this patient with timely routine eye care.