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Title: The Interprofessional Management of a Patient with a Pineal Gland Cyst

Abstract: Patients with pineal gland cysts or tumors have severe visual and systemic symptoms. A proper diagnosis and an interprofessional approach to management are recommended in order to improve the patient’s quality of life.

I. Case History

- **Patient demographics:** 19 year old Caucasian female
- **Chief complaint:** Patient presents as a new patient to The Eye Center at Southern College of Optometry on 07/19/16 for a vision therapy evaluation. Patient complains of dizziness that began 1 year ago after her pineal gland was surgically removed due to a cyst. The severity of the dizziness is worsened when looking at moving targets and also while watching television, washing the dishes, sitting in a moving car, scrolling through her phone or a computer screen, and while reading. She gets tired very easily while reading and no longer reads at all. She now requires her mother to read to her. She also reports frontal headaches when doing near work or when focusing on a specific task for an extended period of time, such as doing the dishes or laundry. She denies double vision or nausea.
- **Ocular, medical history:**
  - Endoscopic surgery to remove the entire pineal gland on July 28, 2015
  - Patient states that before the pineal cyst was discovered, she suffered severe headaches, nausea, and dizziness. The nausea and dizziness were so bad that she no longer had an appetite. Imaging of the brain was subsequently performed which identified the pineal cyst as the cause of her symptoms.
  - Patient currently has the following systemic symptoms (all of which began shortly after her pineal gland was surgically removed):
    - Hormone problems
    - High cortisol levels
    - Uneven sleep patterns (difficulty with both falling asleep and staying asleep)
    - Dizziness and headaches (not as bad as before the surgery but still affecting her activities of daily living)
    - Tension in her neck
  - Refractive history: patient currently wears single vision glasses full time
- **Medications:**
  - Zinc, melatonin, vitamin C supplements, vitamin B supplements, multivitamin, garlic
- **Other salient information:**
• Patient has dropped out of school due to her inability to read for an extended period of time.

II. Pertinent findings

- **Clinical:**
  - Best corrected distance visual acuity: 20/15- OD, OS, OU
  - Best corrected near visual acuity: 20/20 OD, 20/15- OS, 20/20 OU
  - Refraction: -0.25-1.00x105 OD, -0.75-0.75x103 OS, +1.00 add OU
    - Recommended to have separate distance and near vision glasses
    - The patient states that she feels like she is moving “like a skyscraper in the wind” when looking at her phone with the updated near Rx
  - Cover Test: 4 exophoria at distance, 6 exophoria at near
  - Stereo acuity: 20 sec of arc on Randot, Positive forms, No suppression
  - Near Point of Convergence: To The Nose on 3 attempts
  - Amplitude of Accommodation: 8 diopters OD and OS
  - Prism Bar Vergences:
    - BI distance: x/4/2
    - BO distance: x/4/2
    - BI near: x/6/4
    - BO near: x/18/12
    - Patient reports dizziness and a headache during this test
  - Pursuits: some fixation losses, good control and no dizziness noted
  - Saccades: undershooting noted most of the time. Some discomfort noted
  - Developmental Eye Movement (DEM):
    - Adjusted vertical time: 42 seconds
      - Z-score: -3.03
    - Adjusted horizontal time: 48 seconds
      - Z-score: -2.85
    - Ratio: 1.14
      - Z-score: -0.83
    - Errors: zero
      - Z-score: +0.57
  - Anterior Segment Exam: unremarkable OD and OS
  - Further testing could not be completed at this exam due to patient discomfort and dizziness.

- **Physical:** N/A
- **Laboratory studies:** N/A
- **Radiology studies:** One year post-surgical MRI results (07/13/2016):
  - No complications at the occipital surgical site demonstrated
  - Pineal region is free of pineal gland and cyst. No sign of recurrence of cyst or other lesion in the pineal region
  - Normal dimensions and signal flow voids noted in the internal cerebral veins, basal veins of Rosenthal, vein of Galen, and within the straight sinus.
• No intracranial pathology present. The brain and remainder of intracranial anatomy and vascular circulation are normal
• Others: N/A

III. Differential diagnosis

• Primary/leading: Saccadic Dysfunction OU
• Others: Dizziness, Accommodative Insufficiency OU

IV. Diagnosis and discussion

• Elaborate on the condition/Expound on unique features:
  o The pineal gland is a small endocrine gland located centrally between the two hemispheres of the brain near the third cerebral ventricle. Its main functions consist of producing melatonin and helping to regulate sex hormone production by the pituitary gland.
  o Surgical removal of the pineal gland is a lengthy and challenging neurological procedure. This patient had her entire pineal gland as well as a pineal cyst endoscopically removed via the infratentorial supracerebellar approach. During this procedure, the patient is in a semi-seated position so that the pineal gland can be visualized more easily. A bur hole is made on the midline of the skull in the occipital region. The endoscope is inserted between the superior surface of the cerebellum and the inferior surface of the tentorium cerebelli. The entire pineal gland is then excised. This procedure is very challenging due to the pineal glands close proximity to the brainstem and the cerebellum and due to the fact that the endoscope must bypass several cerebral veins.
  o Prior to surgical intervention, patients with pineal gland tumors or cysts are often symptomatic.
    ■ Systemic symptoms include:
      o hydrocephalus
      o headaches
      o nausea
      o altered mental status
      o abnormal sleep pattern
      o endocrine disorders
      o uncoordinated body movements
    ■ Visual/ocular symptoms include:
      o Papilledema
        • Present in 69% of patients involved in a study by Hankinson et al.
      o double vision
      o saccadic dysfunction
      o accommodative insufficiency
      o reduced visual acuity
o Dorsal Midbrain Syndrome/Parinaud’s Syndrome
  • Bilateral mydriasis
  • Paresis of upgaze
  • Light-near dissociation
  • Convergence retraction nystagmus
  • Lid retraction (Collier’s sign)
  • Convergence dysfunction
  • Skew deviation
  • Present in 75% of patients involved in a study by Hankinson et al.³
  • These systemic and visual symptoms are typically due to compression of the brainstem, cerebellum, or cerebral aqueduct by the pineal tumor/cyst.
    o Pineal gland is in close proximity to several structures involved in visual/ocular functions:⁴,⁵
      • superior colliculi (involved in saccadic eye movements)
      • posterior commissure (involved in pupillary light reflex)
      • rostral interstitial nucleus of medial longitudinal fasciculus (riMLF; involved in vertical gaze)
      • Pretectum (involved in pupillary light reflex and optokinetic response)
    o After surgical removal of the cyst/tumor or the entire pineal gland, many patients report improvement in their symptoms, but some sequelae remain long-term, as is the case for this patient.
      o Study by Hart et al. found that 55% of patients suffered long-term visual disturbance postoperatively, typically convergence or accommodative dysfunctions.⁶

V. Treatment, management

• Treatment and response to treatment:
  o Prescribed separate distance and near vision glasses in order to help alleviate symptoms with near work.
  o Binasal occluders were applied to both the distance and near vision glasses with translucent tape. The patient reports improved visual comfort with the occlusion. It has also reduced her feelings of dizziness.
    • As described by Steven Gallop in 1998, binasal occluders help to decrease the visual stress. They also act as a visual reference point that can help to stabilize and organize visual input.⁷
  o This patient is currently undergoing weekly vision therapy sessions and doing home activities about 5 days a week. The patient has participated in 5 sessions so far.
    • improve saccadic eye movements and accommodative abilities so that the patient may return to her normal activities of daily living
Because of the patient’s multiple systemic symptoms, she is regularly monitored by a neurosurgeon (to monitor for any intracranial changes or abnormalities), an endocrinologist (to control melatonin and hormone levels), and a chiropractor (to relieve tension in her neck that began after the surgery).

VI. Conclusion

- **Clinical pearls, take away points if indicated:** Because of the pineal glands close proximity to the brainstem, cerebellum, and cerebral veins, development of a tumor or cyst on the gland can cause widespread systemic and visual symptoms. As seen by this patient, many of these symptoms still persist even after surgical intervention. A collaborative interprofessional approach should be taken to manage these symptoms in order to improve the patient’s overall quality of life. The interprofessional team should include, but need not be limited to:
  - a neurosurgeon (to continually monitor for any intracranial changes postoperatively)
  - an optometrist (to manage ocular and visual sequelae through the use of lenses, binasal occluders, prism, or vision therapy)
  - an endocrinologist (to manage melatonin and hormone levels)
  - a chiropractor (to alleviate tension in the neck and back that can occur postsurgically)

Management of patients with pineal tumors/cysts will most likely be long-term, and optometrists should take an active role in the management of these patients both pre and postoperatively. Optometrists should be aware of the multiple systemic symptoms that patients with this condition might be suffering from in order to make the necessary referrals.

Bibliography: