Case Corner

Management of patients with thyroid ophthalmopathy
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INTRODUCTION

Patients with thyroid-associated ophthalmopathy (TAO or thyroid eye disease TED) may present diagnostic and therapeutic challenges. As the disease evolves, a myriad of ophthalmic complications may occur that requires the involvement of multiple ophthalmic disciplines. Orthoptists are valuable members of the eye care team who are often called upon to manage the diplopic complaints of these patients, and to document changes in the motility during the protracted follow-up period before surgical therapy is appropriate. In the following discussion, our participants offer their insights into the care and management of patients with thyroid eye disease.

DISCUSSION

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The treatment of ocular problems related to thyroid-associated ophthalmopathy (TAO) is aimed at preventing visual loss and relieving symptoms of exposure keratopathy and restrictive myopathy. Since patients with TAO may develop eye problems at any stage, long-term follow-up is favored. A neuro-ophthalmologist and orthoptist are an effective team in caring for these patients.

At initial evaluation, confirmation of the diagnosis is important along with gathering baseline information. It is important to inquire about the patient's complaints. Usually keratopathy, swelling, and diplopia are more pronounced in the morning; presumably due to increased orbital edema during recumbency overnight. Patients who smoke are counseled to quit since cigarette smoking worsens TAO.

Once the diagnosis of TAO is established, attention is directed toward four important areas: external symptoms, exposure keratopathy, restrictive myopathy, and compressive optic neuropathy. We feel that it is most important to rule out the latter since immediate intervention is necessary. All parameters of sensory afferent visual function are tested, and, if abnormal, retested to ensure reliability. We particularly emphasize the determination of best corrected visual acuity. A good refraction is mandatory and, if vision is still reduced, a thorough search for any other cause (such as cataracts, macular degeneration, or exposure keratopathy) must be ruled out before attributing visual loss to compressive optic neuropathy. The presence of compressive optic neuropathy warrants either orbital decompression surgery, external radiation, or both. Oral corticosteroids are only used temporarily in the perioperative/radiation phase.

The neuro-ophthalmologist takes a thorough history and examines all pa-
tients with suspected TAO. Several important features on examination are emphasized including the following:

1. sensory afferent visual function (best corrected visual acuity, visual field examination with Goldmann or Humphrey perimetry, color comparison testing, and pupillary examination for relative afferent defect)
2. external and orbital examination (exophthalmometry and orbital sonography)
3. eyelid function (lid retraction, temporal flaring, lid lag, and lagophthalmos)
4. slit-lamp examination (exposure keratopathy)
5. intraocular pressure in primary position and upgaze (to determine if a rise in pressure exists—although forced duction testing is thought to be more reliable in cases of restrictive myopathy)
6. funduscopy (with optic disc photographs)

In some cases, imaging with thin cut axial and coronal CT scan of the orbits is ordered, especially if the diagnosis is uncertain or if orbital decompression surgery is being planned.

All patients are also evaluated by an orthoptist. The role of the orthoptist in our clinic is to aid in the diagnosis, quantify and document the ocular motility, restore binocular single vision by use of prisms if possible, and assist the ophthalmologist in the follow-up of these patients. Since motility problems may arise several years after the diagnosis of thyroid dysfunction, a baseline orthoptic examination is helpful. A Lees screen is included to monitor ocular motility patterns and enhance inter-examiner reliability when dealing with such incomitant and sometimes progressive motility problems. Comparing a Lees screen from one visit to the next can easily establish progression or stability of ocular movements. A synoptophore is particularly useful for measurement of torsion. We feel that both the Lees screen and the synoptophore minimize variability because the chin is in a fixed and reproducible position, whereas prism measurements may vary depending upon head position. A Maddox rod is utilized if the patient cannot fixate in primary position. Frequent monitoring of the ocular deviation is important in our clinic since the patient may not be evaluated by a strabismologist until stability is reached.

Most of our patients with TAO present with or develop diplopia. All have difficulty with upgaze and some also have difficulty with horizontal eye movements. The majority of patients with diplopia are given Fresnel® prisms to restore reasonably comfortable binocular single vision, at least in primary position. These patients are monitored frequently by the orthoptist and the prisms are easily changed as the strabismus evolves. Occasionally, patients are unable to manage with prisms due to various reasons—incomitant deviations, large deviations, and torsion. In this group of patients, temporary occlusion with 3M Blenderm™ tape on one lens of their glasses is recommended. They are still followed at regular intervals to document any change in their motility. Patients are not considered for strabismus surgery until there is stability in both their disease and ocular deviation for a minimum of six months.

In our experience, women with TAO outnumber men by 4.5 to 1, which is in keeping with other practices. On average, thyroid dysfunction is present for 3 years before diplopia is manifest; however, this can range from one year to as long as 20 years. The onset of diplopia occurs on average at age 55.

In our practice, 50% of patients have been diagnosed with thyroid dysfunction
prior to referral. These patients are referred for various reasons—proptosis, diplopia, lid retraction, decreased visual acuity, or foreign body sensation. The other 50% present with similar symptoms, but have not been previously diagnosed with thyroid dysfunction. Of this later group of patients, it is interesting that about half of them actually have symptoms of diplopia at initial presentation.

Approximately 25% of patients in the neuro-ophthalmology clinic require treatment for compressive optic neuropathy. We prefer bilateral simultaneous orbital decompression surgery, but occasionally patients receive 2000 Rads of external beam radiation if their systemic health precludes surgery. Occasionally, we have recommended both decompression and radiation in patients with severe disease.

In the past three years, only 2% of our TAO patients have been proven to have myasthenia as well. In these patients, eyelid ptosis prompted this consideration and Tensilon testing confirmed our suspicions.

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In the work-up of patients with thyroid myopathy, the usual tests done include: measurements in all fields of gaze (both at distance and at near), fields of single binocular vision plotted with a perimeter, and torsion in primary and downgaze measured with the double Maddox rod test. Muscle involvement is normally determined by measurements in all fields of gaze. Ultrasound is done primarily to confirm the diagnosis.

At the time that patients present with their motility problem, the acute stage of thyroid myopathy has usually passed. If the acute stage is present, steroids and occasionally radiation may be of benefit to alleviate the acute congested stage. The coexistence of myasthenia gravis should be considered in all patients with thyroid myopathy. Symptoms include generalized muscular weaknesses, upper eyelid ptosis, and strabismus with diplopia that show variability with fatigue. People that present with thyroid myopathy are evaluated by our neuro-ophthalmology service to rule on the presence of an associated myasthenia gravis. If myasthenia coexists, then this should be brought under control and stable measurements should be obtained for at least three months prior to any surgical treatment of the thyroid myopathy.

All measurements should be taken into account when planning treatment for a patient with thyroid myopathy. The most usual presentation is either an esotropia or a vertical misalignment (i.e., usually consisting of a hypotropia). Versions usually show limitations of abduction and elevation. Therefore, the amount of esotropia increases in abduction and the amount of vertical deviation increases in elevation.

In those patients with small comitant vertical deviations, prisms can be used—either in a temporary fashion with Fresnel™ press on prisms or permanently with ground in prisms—to control the deviation in primary and reading position. As long as this works, extra ocular muscle surgery is not necessary. If the deviation is extremely incomitant or large, prisms have limited benefit.

The optimal time for surgical treatment of strabismus is when the acute phase is over, the thyroid function is normal, and stable measurements have occurred for at least three months. Indications for surgery are diplopia in primary position, reading position, or both, and significant abnormal head position to produce fusion.

Special attention should be given to what happens to the preoperative mea-
measurements in downgaze. For example, if a hypotropia disappears in downgaze, you should be suspicious that the ipsilateral superior rectus is involved. The amount of inferior rectus recession should be carefully adjusted to treat only the deviation in primary position and not allow for full elevation. If the entire restriction is released, then the superior rectus postoperatively can take over and cause a limitation of depression in downgaze and an overcorrection will occur. The same is true with the amount of esotropia. If the amount of esotropia in primary either decreases or is eliminated in downgaze, then any medial rectus surgery for the deviation in primary may cause an overcorrection postoperatively (i.e., exotropia in downgaze). In those patients that present with esotropia and vertical deviation, caution should be taken not to weaken all adductors, that is, weaken the medial recti and the inferior rectus simultaneously, because an exotropia may occur postoperatively in downgaze.

In all cases of thyroid myopathy adjustable sutures are invaluable. It is very difficult to determine the exact amount of recession necessary to relieve all restriction and obtain all the goals of fusion in primary and reading positions. Adjustable sutures allow the surgeon to accomplish this with a much higher degree of predictability.

The number of patients with thyroid myopathy seems to remain fairly constant over the years. There has been speculation that the numbers have increased or decreased, but in our clinical experience the numbers seem to be fairly stable.

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The majority of the patients I see with thyroid ophthalmopathy are referred after diagnosis for the management of the related strabismus. Nevertheless, I occasionally see patients with new diplopia in whom the cause has not been determined and who ultimately turn out to have thyroid-related myopathy.

With new onset diplopia, I may suspect thyroid myopathy based on the typical motility patterns (discussed below) and/or because of associated signs or symptoms such as eyelid retraction, proptosis, conjunctival injection or chemosis, exposure keratopathy, or even visual loss related to compressive optic neuropathy. These patients require a comprehensive ophthalmologic exam to address these issues. Visual fields should be performed and any visual compromise addressed immediately. If the diagnosis is in question, I generally get an orbital CT with coronal reconstruction to assess extraocular muscle (EOM) size, or alternatively perform diagnostic A-scan in the office to assess EOM size. Additionally, I will order a complete thyroid panel and have the patient evaluated by an endocrinologist. Once the diagnosis is confirmed, I have the patient followed by neuro-ophthalmology if there are any visual issues and oculoplastics for significant proptosis and/or lid abnormalities.

In managing the strabismus related to thyroid myopathy, one must first determine where the patient is in the course of the disease. Strabismus surgery is rarely if ever indicated in patients whose disease is active or unstable, patients who are undergoing treatment such as orbital radiation or systemic steroids, or patients who are likely to undergo orbital decompression in the near future. These patients should be managed expectantly with observation or prisms where helpful. I reserve strabismus surgery for patients with significant deviations, which are not amenable to prism correction, and in whom the deviation has been stable for at least six months.
My strabismus evaluation is as follows. History should be directed at whether or not diplopia is present. When present, I determine under what conditions (activities) diplopia is present. I ask what gaze positions are affected, and whether the diplopia is horizontal, vertical, or both. My motility examination begins with testing of versions and ductions, looking for any limitation (restriction). I will then measure the ocular deviation by prism cover test in as many positions as possible, although with severe restriction it may not be possible to measure extreme gaze positions (especially up-gaze). When performing prism-cover testing I always try to place the prism over the most restricted eye to avoid exaggerated secondary deviations. I try to subjectively neutralize diplopia in each measurable gaze position and plan surgical correction appropriately. Deviations should match the restrictions noted on duction testing. If there are inconsistencies, other causes of diplopia should be considered.

I have found my patients to generally parallel the expected findings in this disease, at least clinically, with inferior rectus (IR) involvement being most common followed by medial rectus (MR) involvement. MR involvement is almost always associated with IR involvement, while IR involvement is more frequently isolated. Superior rectus involvement is a distant third and usually associated with IR involvement as well. I have only seen lateral rectus (LR) involvement in a small number of patients. While MR involvement with esotropia is generally quite obvious, SR involvement may be masked by IR involvement. Comparisons of side gaze deviations as well as up and down gaze are critical in determining what muscles are involved.

For example, with IR involvement a hypotropia is present (unless the contra-lateral IR is equally or more involved). The hypotropia should increase in attempted up-gaze and decrease or disappear in down-gaze, if the latter does not occur one should suspect ipsilateral SR involvement. Additionally, the hypotropia should increase with abduction and decrease with adduction as the involved eye moves into and then out of the field of action of the involved IR. If the hypotropia fails to increase on abduction one should again suspect coexistent SR involvement. The findings should always be corroborated with what is seen on duction testing, and finally forced-ductions (generally done at the time of surgery) can further delineate questionable cases.

Management is directed at eliminating diplopia and relieving restrictions. I generally reserve prism for unstable cases (temporizing), and small deviations either in non-operated or postoperative patients in whom further surgery is unlikely to improve alignment. I have generally found press-on prisms to be poorly tolerated over the long term, and I vastly prefer ground-in prism for stable patients with relatively small deviations.

When approaching these patients surgically, one must consider both the deviation and the motility. For example, while unilateral IR recession will generally eliminate a primary position hypotropia, bilateral asymmetric surgery is often needed to maximize improvement in up and down gaze as well as minimizing restriction. One must always remember, however, the importance of down-gaze, and not be overly aggressive with IR weakening. If SR involvement is suspected, I will try to confirm that with forced ductions at the time of surgery and adjust the surgical plan as necessary. It is generally preferable to have residual up-gaze restriction and diplopia than to create limited down-gaze and/or down-gaze hypertropia or exotropia. Esotropia with limited abduction is easily addressed sur-
gically with appropriate MR recession(s). I am always careful to point out to pa-
tients preoperatively that while surgery can improve and in some cases eliminate
diplopia, it is often not reasonable to ex-
pect complete normalization of motility (especially in severe cases) and that post-
operative prism may be necessary.

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A Graves disease patient often has mul-
tiple ocular findings that must be ad-
dressed by several ophthalmic specialty
areas, including neuro-ophthalmology,
cornea, oculoplastics, and strabismus. The clinician should always be suspicious
that an adult patient with acquired diplo-
pia (in some or all gaze positions) may
have thyroid eye disease.

In our practice, the motility work-up
emphasizes examination for limitation of
eye movement. Ductions in each eye are
compared with the fellow eye or with ex-
pected norms and any restriction of range
is noted. Prism and cover measurements
in diagnostic gazes are taken when possi-
ble. Krimskey estimates are made where
moderate to severe limitation of motility
exists, remembering that these angles
will vary depending on the patient’s head
position, effort, and degree of discomfort.
Symptomatic motility restrictions are al-
ways confirmed with forced duction test-
ing. This may be performed in the office
or immediately prior to EOM surgery. In
the case of a restriction of elevation, it is
also important to check passive downgaze,
which may have a similar though less pro-
nounced limitation.

Some patients with thyroid eye disease
adopt a head position for comfort or diplo-
pia resolution. Head tilt test and double
Maddox rod assessment are usually unre-
markable. To complete the exam, notation
is made of the exophthalmometry reading,
lid fissure height, lid function, and slit
lamp inspection of the cornea for signs of
exposure.

To establish the diagnosis of thyroid eye
disease, a CT scan or MRI is obtained. Or-
bital ultrasonography is not consistently
available at our institution but we have
found radiographic evidence of enlarged
rectus muscles on orbital sections in the
majority of patients studied. Even so, the
patient’s clinical picture is most persuas-
ive when making treatment decisions.

The possibility exists for a double diag-
nosis of myasthenia gravis along with thy-
rroid eye disease. In our experience, this
has been a very rare occurrence. The pa-
tient in whom there is variability of duc-
tion limitation and any amount of ptosis
rather than lid retraction should receive
further testing for myasthenia gravis. In
a chart review of 36 randomly selected pa-
tients with the diagnosis of TED seen in
the past five years in our practice, none
had myasthenia gravis. One patient with
acquired restrictive strabismus and a
slight ptosis was investigated for both dis-
eases. This gentleman actually had a neg-
avative orbital CT scan but a positive ACH
receptor binding antibody test. As in this
case, the situation may call for a resolu-
tion of a differential diagnosis rather than
the verification of both diagnoses.

We estimate that more than half of our
Graves ophthalmopathy patients are smok-
ers. Possible adverse effects of smoking on
the progression and treatment of thyroid
eye disease are discussed. At the same
time, we recognize that smoking is a very
difficult habit for many people to break,
with no guarantee that their eye problems
will resolve if they do stop.

The characteristic infiltration and swell-
ing of extraocular muscles in Graves dis-
ease can cause pressures within the orbit
resulting in optic nerve compression. Opt-
ic nerve damage with permanent visual
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loss will occur unless immediately treated. Oral steroids, low dose orbital radiation, or orbital decompression are advised for those who exhibit clinical signs of optic neuropathy. This approach is not recommended for simple EOM restrictions due to myopathy. Radiation or orbital decompression does not preclude the possibility of strabismus surgery at a later date.

Non-surgical management of diplopia in TED patients is employed when possible. Since these patients must demonstrate a stable clinical picture for 3 to 4 months or longer before initial muscle surgery, we offer a trial of Fresnel prisms to restore an area of single binocular vision in primary and downgaze. Some are unable to tolerate this due to incomitance, but the prism can be easily removed and a monocular patch used if desired. The occasional patients with small deviations or who wish to avoid surgery may be helped with differing amounts of prism on the distance and near segments cut along the bifocal line. Press-on prism is also used postoperatively for patients who have a small residual deviation that is “not quite” fused. Once a selected amount of prism is worn and found to work well, it is incorporated into the glasses.

The important factors in deciding when to operate for thyroid-related strabismus are: the patient is symptomatic, the deviation is poorly corrected with prisms, and the deviation has proved to be stable. These qualifications apply to re-operations as well as to the first procedure.

It is well known that surgery to correct thyroid myopathy must be aimed at releasing tight or tethered muscles. For this reason, most procedures involve recession of one or more affected rectus muscles. Adjustable sutures are used extensively on our patients, sometimes on more than one muscle per eye.

To minimize the downgaze limitation and lid retraction that can accompany inferior rectus recession, the amount of recession is kept to the absolute minimum necessary to release the restriction. For large vertical deviations, inferior rectus weakening is combined with recession of the contralateral superior rectus. A small undercorrection in upgaze is more forgiving than an imbalance in downgaze. Because of our approach, we have encountered few problems with downgaze limitation in our patients. Where it persisted, this has been successfully treated with small amounts of vertical prism in the glasses. Some patients have benefited from a Faden procedure on the more mobile eye. One patient who had bilateral inferior rectus recessions had mild symmetric limitation of downgaze, which she managed by lowering her chin when going down stairs.

We have not noted any trend toward increase or decrease in number of thyroid eye disease patients referred to this practice over the past ten to fifteen years.