Temporal Arteritis: Headaches With A Risk For Sudden Blindness

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Temporal arteritis (TA), also called Giant-Cell Arteritis (GCA), is a condition that is a true ophthalmic emergency. Even though it is a chronic inflammatory disease, involving many blood vessels of the body (thus the term “arteritis”), most notably TA/GCA has a predilection for affecting the blood vessels of the head and neck. There is a distinct risk of sudden blindness or stroke, if the inflammation involves the vasculature to the eye or brain.

TA occurs in patients usually older than 70 yrs of age, affecting women more often than men. A 50-year retrospective study out of the Mayo clinic of 173 cases had a 79% female incidence, and the mean age of diagnosis was 74.8 years.

Even though clinical suspicion is raised when an adult presents with temporal headaches, only about half of patients with biopsy-proven disease actually complain of headaches. TA/GCA is a rheumatologic condition associated more commonly with other general systemic symptoms, such as malaise, unintentional weight loss, proximal muscle aches (polymyalgia rheumatica), loss of appetite, weakness, and anemia. Symptoms of involvement in the head and neck can include jaw claudication (pain with repeated chewing), temporal headaches, temporal scalp tenderness, scalp dysesthesias (tingling or numbness, especially when touching the hair), and possibly double vision or, more worrisome, a transient loss of vision (amaurosis). The goal for doctors is to make the diagnosis and treat the disease before irreversible visual loss or stroke occurs.

The diagnosis is established by a constellation of clinical symptoms (it is considered highly suspect if three or more of the above-listed symptoms are present). Laboratory tests which may aid in making the diagnosis include elevated serum acute-phase reactants (ESR and CRP), thrombocytosis, and mild to moderate anemia. But there are many cases of TA/GCA that have a very low, normal ESR. Some studies have shown that an elevated CRP is more likely to be seen with this disease. And keep in mind that an elevated ESR may be found in many other diseases besides TA (infection, occult malignancy, etc).

The gold standard for making the diagnosis is still tissue biopsy. And the superficial temporal artery on the scalp is an easily-obtainable specimen under local anesthesia. It is important to obtain an adequately long specimen (at least 2 cm is recommended).
Temporal Arteritis, continued from page 1

preferred), since TA/GCA is well-known for having “skip areas” of involvement of vessels, meaning that there may be patchy areas of vessel engulfed with the inflammation, next to very normal areas of uninvolved vessel. Surgical pathology preparation should be done with 3 mm serial cross-sectional segments, to avoid false negative results, since a pathologic study looking at many specimens found the shortest area of inflamed vessel was 3.2 mm.

The dilemma is that the treatment for this disease (high dose immunosuppressives, long term tapering regimens) is sometimes worse than the disease itself for an elderly patient. The complications from steroids are numerous, and many of these patients already have fragile health (osteoporosis, diabetes, GI bleeding risk, etc). So tissue biopsy proof of the disease is very important. In patients who are very highly suspect, bilateral temporal artery biopsy should be considered.

Treatment regimens are controversial. How high should the initial dose of Prednisone be (60, 80, or 100 mg)? Some neuro-ophthalmologists even advocate IV pulse steroids as the initial regimen. And how slowly should you taper? Can some patients eventually be tapered off completely? Review of the literature reveals no “right answers” to these questions. But multicenter trials have looked at appropriate initial oral dosing of between 0.5-1.0 mg/kg of Prednisone (I personally use 1.0 mg/kg), or IV Solumedrol between 250 mg to 1 gm initial bolus (I personally consider using 1 gm bolus treatment in suspicious cases with amaurosis symptoms). Also steroid-sparing agents may be used in diabetics or patients in whom steroids are contraindicated, but keep in mind that their onset of action is delayed (a week or more). So high dose steroids initially are still recommended. Most clinicians initiate treatment immediately in patients who are highly clinically suspicious, even before the biopsy can be obtained, and maintain the high dose steroids until the biopsy results are known. The biopsy will be most useful (more easily interpreted if showing classic inflammation) if it is performed within 1-2 weeks of initiating treatment.

There is still a lot of new information being learned about TA/GCA. It has been long suggested that this inflammatory disease might be triggered by an infectious agent. Definite cyclical variations in incidence have been shown in studies out of Minnesota, Scotland, France, and Israel, with peaks in incidence being simultaneous to respiratory infections. (I personally noted that most of my positive biopsy cases occur between November and February!) And a group out of UCLA recently isolated gene fragments from cells in GCA temporal arteries with high homology to microbial genes. And much research is being done to compare the relative significance of the different laboratory abnormalities in predicting the diseases (questions such as: is CRP better than...
ESR? Is thrombocytosis a risk factor for vision loss? Is IL-6 a more-sensitive marker?). And clinical trials are underway to investigate if regimens of steroid-sparing agents might be useful in the tapering phase to lessen the relapse rate of the disease. And there was a promising study (however with small numbers) which showed a possible future role for duplex ultrasonography to aid in the diagnosis of TA/GCA (the presence of a hyperechoic halo around the temporal artery, indicating signs of edema and vessel stenosis, was highly correlated with subsequent positive temporal artery biopsies). This may or may not prove to be useful diagnostically, however, since the negative consequences of the high-dose steroid treatment, as well as the risk of not treating an unknown patient whom may have the disease, will most likely still rely on the results of a tissue biopsy.

TA/GCA is a high risk disease, presenting with a constellation of symptoms and frequently abnormal labs, and the clinician can determine a high rate of suspicion for the disease in any one patient. Yet still this is one disease where truly “the tissue is the issue” for clinching the diagnosis.

Dr. Keefe received her medical degree from Case Western University, School of Medicine. She completed an Internal Medicine residency at the National Naval Medical Center, Bethesda MD, and her Ophthalmology residency at the Naval Medical Center in San Diego, CA. Dr. Keefe did her Fellowship on Ophthalmic Pathology at the Armed Forces Institute of Pathology (AFIP), Washington DC. Her areas of expertise include Oculoplastic and Orbital Surgery, Ocular Pathology, and Cataract Surgery.

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