Temporal Artery Biopsy for Diagnosis of Giant Cell Arteritis

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ABSTRACT:

Giant cell arteritis is a vasculitis of the medium to large blood vessels. It is a true ocular emergency because it can rapidly cause severe and permanent loss of vision. The gold standard for diagnosis of giant cell arteritis is the temporal artery biopsy.

Keywords: Giant cell arteritis, temporal arteritis, temporal artery biopsy

INTRODUCTION:

Giant cell arteritis (GCA), also known as temporal arteritis, is the most common type of systemic vasculitis, with a lifetime risk of 1% in women and 0.5% in men in the United States.[1] This condition is seen exclusively in patients over the age of 50, and involves inflammation of large and medium sized blood vessels throughout the body.[2] The risk of this condition increases with age, with the greatest risk being for those individuals over the age of 70.[1] It is most commonly found in individuals of Northern European, and in particular Scandinavian descent.[1] GCA carries with it the risk of permanent blindness, so prompt diagnosis and management are vital.

DIAGNOSIS OF GIANT CELL ARTERITIS:

GCA is typically diagnosed by its presentation of a variety of ocular and systemic symptoms and signs. Ocular symptoms are present in up to 45% of GCA cases.[3]

The most concerning ocular complication that can occur in these patients is arteritic anterior ischemic optic neuropathy (AAION). This causes sudden painless vision loss that typically starts unilaterally.[4] This irreversible vision loss occurs in 15-20% of GCA patients, and if it occurs in one eye there is a 25-50% chance of the other eye becoming affected in one week without treatment.[1] In some cases, the permanent vision loss may be precedeamaurosis fugax, which describes a transient loss of vision in one eye.[1,3] If this happens it is an indication that immediate treatment is needed before the vision loss becomes permanent.

In addition to vision loss, GCA can present with several other significant ocular signs and symptoms. The presence of an afferent pupillary defect can be indicative of significant damage to the optic nerve and can be found in these patients unless both eyes are equally affected.[1,4] Patients with GCA may also present with visual field loss. This typically is an altitudinal or central defect.[4] In a small subset of these patients there can be presenting symptoms of diplopia and extraocular muscle dysfunction. This is because GCA can cause ischemic damage to any part of the oculomotor system in 5% of cases.[1] Ocular examination of a patient with GCA will typically reveal a swollen, pale optic nerve if AAION is occurring, and flame-shaped hemorrhages may also be present.[4] However, in some cases the fundus examination may reveal nothing out of the ordinary, especially if early in the condition.[1]

In addition to the ocular symptoms and signs of GCA, there are also numerous systemic findings that may be associated with it. The most common complaint of these patients is a new headache or facial pain.[3] Two other very common complaints are tenderness of the area around the temporal artery and jaw claudication, or pain in and around the jaw worsened by chewing.[1,3] These patients will often present with a fever, along with fatigue, anorexia, and weight loss.[1,4]

Finally, 40-50% of patients with GCA will also have polymyalgia rheumatica (PMR) concurrently.[1] Polymyalgia rheumatica is an inflammatory rheumatological condition found exclusively in patients over the age of 50, and most commonly in patients that are 70-80.[5] Patients will present with lingering pain and stiffness in the neck, shoulders and pelvic area for at least 30 minutes in the morning.[6] Given the frequency with which GCA and

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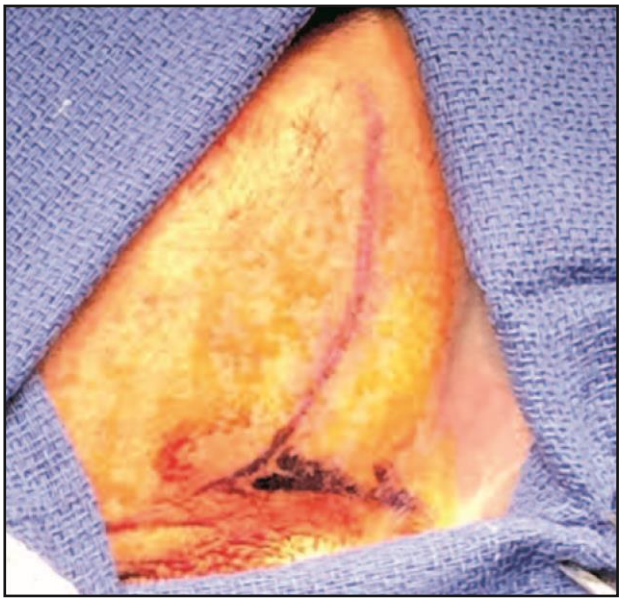

Fig. 1 Patient undergoing temporal artery biopsy showing marking of the frontal branch of the temporal artery prior to incision being made.

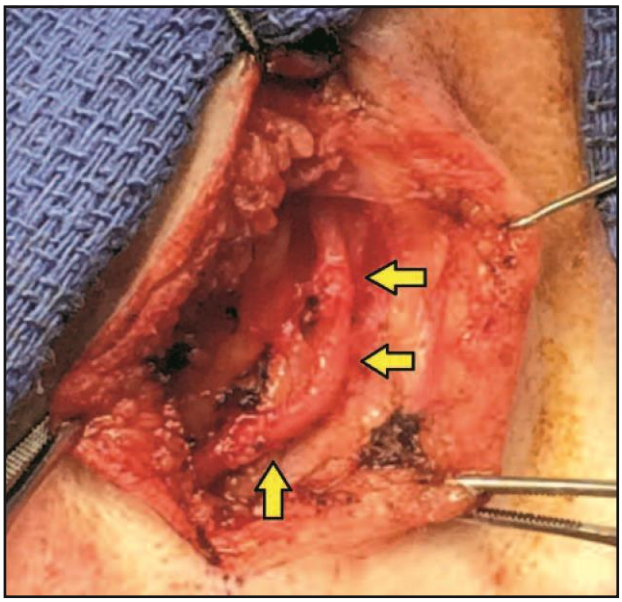

Fig. 2 Frontal branch of the temporal artery exposed in a sufficient section for biopsy (arrows).

PMR occur together, it has been suggested that they may in fact be two different stages of the same disease.[6]

While the symptoms mentioned previously should raise suspicion of GCA, its diagnosis is further delineated through laboratory testing. Patients suspected of having this condition should be sent for an erythrocyte sedimentation rate (ESR), C-reactive protein (CRP) and complete blood count (CBC).[4] ESR and CRP will both likely be elevated in a patient with GCA, but this is not enough to diagnose the condition, and normal values are not necessarily enough to rule it out.[1] CBC results will typically show significantly elevated platelet levels.[4] An elevated ESR increases the risk of a positive temporal artery biopsy by 1.5 times.[7] An elevated CRP increases it by 5.3 times, and an elevated platelet count increases it by 4.2 times.[7] If all three of the laboratory results are abnormally high the odds of a positive temporal artery biopsy become 8 times greater.[7] While all of these tests can help to raise suspicion of GCA, the only way to definitively diagnose the condition is with a temporal
artery biopsy.[8, 9]

TEMPORAL ARTERY BIOPSY: A temporal artery biopsy is the gold standard for the diagnosis of GCA.[8,10] Examination of the biopsy specimen would reveal mononuclear cell infiltrates and possibly giant cells along with vasculitis.[3] In performing the biopsy, it is important to take a longer portion of the artery, typically at least 2.5 cm. This longer specimen would avoid the problem

of skip lesions. These are present in approximately 10% of cases.[8] The biopsy should ideally be performed prior to starting treatment, but the vasculitis can still be seen within at least two weeks of starting treatment.[6,8]

The frontal branch of the temporal artery is typically used for the biopsy because it is a superficial vessel and located in an area with redundant blood supply.[11] The selected branch of the artery is then marked with a marking pen (Figure 1). The vessel is located through palpation, or if palpation is difficult, through the use of a portable Doppler ultrasound device.[9,11] An injection of 2% lidocaine is given to anesthetize the area, and an incision is made along the marking line through the skin and subcutaneous tissue.[9] Thermocautery is used to control bleeding. Blunt dissection with scissors is used to reveal the artery, while the adjacent tissue is held open with skin hooks.[11] Once the artery is exposed, adjacent tissue needs

to be expanded parallel to the artery to retrieve a sufficiently long specimen (Figure 2).[9] Sutures or surgical clamps are used to block blood flow through the vessel.[9,11] This section of the artery is then excised (Figure 3).[8]

Once the specimen has been removed, the incision can be sutured closed. The use of vicryl dissolvable sutures simplifies the healing process. To aid in holding the wound closed, cyanoacrylate skin adhesive and adhesive skin closure strips should be used. A pressure bandage is applied for 24 hours.[9] Recovery is typically quick following this procedure, but complications such as bleeding, hematoma, infection and nerve damage are possible.[11]

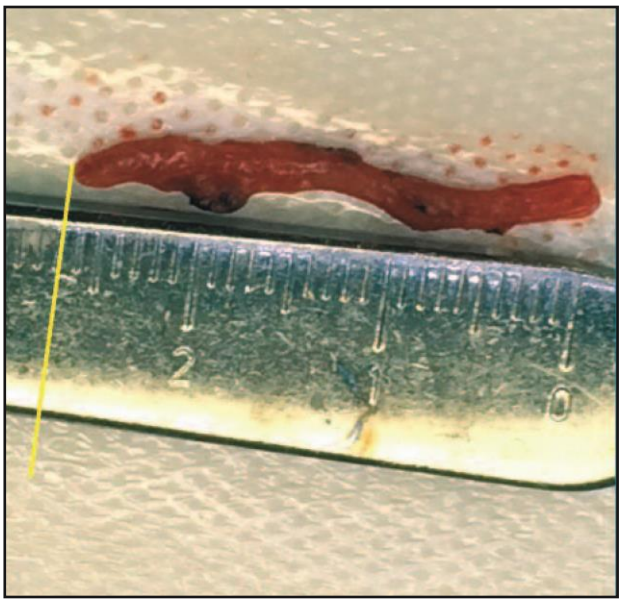

Fig. 3 Specimen of the frontal branch of the temporal artery measuring 2.7 cm in length.

It is important to remember that while a positive temporal artery biopsy will confirm the diagnosis of GCA, a negative biopsy does not necessarily rule it out. As mentioned previously, approximately 10% of cases of GCA present with skip lesions.[8] There have also been a number of cases of biopsy-negative GCA, in which the temporal artery biopsy was negative but a diagnosis of GCA was still made.[10]

Management of Giant Cell Arteritis:

GCA is treated with high-dose oral corticosteroids. The typical starting oral dose is 40-60 mg per day.[6] In patients experiencing vision loss, three days of intravenous methylprednisolone 500-1000 mg per day can be used as an initial treatment.10 GCA patients typically require a very slow taper of their steroids, in some cases lasting years.[12] This is at least partially due to the high rate of relapse as steroid levels are reduced.[2] Relapse rates as high as 65% have been reported.[3] The long-term use of steroids required by this condition are problematic for two reasons. First, steroids, especially oral steroids, have a number of significant side effects. Patients on long-term corticosteroids have reported poor blood sugar control, multiple bone fractures, hypertension, gastrointestinal bleeding, cataract development, glaucoma and infections as a result of their therapy.[12] Secondly, patients with underlying conditions related to the side effects such as diabetes, hypertension or osteoporosis, are at an even greater risk of these complications and ideally shouldn't be on steroids at all.

Until recently, there were no viable alternatives to steroid use for treating GCA.[2] Tocilizumab, a biologic, has been shown to be able to treat GCA as a monotherapy in a small pilot study.[2] It has been approved by Health Canada in November 2017, to be used with oral steroids to reduce the risk of recurrence of GCA and has been shown to significantly do so.[13] Tocilizumab is subcutaneously injected every 1-2 weeks to aid in effectively tapering off oral steroids.[13]

Conclusion:

GCA is a potentially sight-threatening condition that affects the elderly. Prompt diagnosis and treatment are required to preserve these patients' sight and to protect their quality of life. Multiple symptoms are associated with this condition, including sudden painless vision loss, afferent pupillary defect, jaw claudication, new-onset headache, tenderness of the temples on palpation, fever, weight loss and malaise. ESR, CRP and platelet levels tend to be elevated in these patients. However, the gold standard for diagnosing GCA is through temporal artery biopsy. Treatment is usually with high dose oral steroids, and while this will continue to be the case, tocilizumab provides a possible alternative or supplemental therapy.

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