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Vasculitis





Common types of Vasculitis



 Behcet's disease which results in blood vessel inflammation

Buerger's disease which affects arteries and veins in the arms and legs

- Churg-Strauss syndrome
 which leads to inflammation and reduced
 blood flow
- Cryoglobulinemia
 which involves abnormal blood proteins

Giant cell arteritis which affects the lining of arteries

- Granulomatosis with polyangiitis
 which affects the nose, sinuses, and throat
- Henoch-Schonlein purpura which affects small vessels in the skin and joints
- Kawasaki disease which affects the coronary arteries
- Takayasu's arteritis which can affect the aorta

Not intended as Medical Advice

- This lecture is informational only and not intended to diagnose or suggest treatments to any individual listening to this lecture.
- We advise you to seek medical direction with your licensed primary care provider.



Learning Objectives:

- What is Vasculitis?
- Common Types
- Mainstream Medical Treatments
- Alternative Treatments

Definition of Vasculitis

- Vasculitis is an autoimmune disorder heralded by blood vessel involvement. Blood vessels large and small, arteries, veins and capillaries can all be involved.
- The diseases are usually classified by the size of the blood vessels involved and/or the organs that are affected. There are several triggers and different regions of the body that can be affected.
- In general, inflammation develops in blood vessel walls which cause swelling, narrowing of the lumen and weakness of the structural wall.

Pathophysiology

Cause:

- Inflammation is sparked by some type of trigger, whether it is caused by an underlying infection (virus, microbe) or another environmental agitatent. The tissue of the blood vessel's inner lining (endothelium) become inflamed when immune cells become hyperactive and attack native tissue. This attack is perceived as a defensive maneuver to destroy a foreign intruder like a virus, but the body's immune system is tricked, and the immune cells attack the "self" (autoimmunity).
- When the site of attack is a blood vessel, we call this a vasculitis. While most vasculitis is due to inflammation and autoimmune disorders, occasionally cancers such as lymphoma and leukemia can be the root cause. Cases of drug-induced vasculitis exist.

Cause:

- Problems occur when distal organs or systems are affected. A severely narrowed blood vessel or even occluded due to severe inflammation will not allow the passage of oxygen containing RBCs and anything downstream suffers hypoxia. This leads to necrosis or death of tissue starved of oxygen. End organs such as kidneys and lung tissue can be destroyed. When the vessel wall becomes weakened to the point of dilating or expanding, the integrity may be so stressed the vessel ruptures. Surgical intervention is sometimes necessary to repair a damaged vessel before it ruptures.
- There are different types of vasculitis and different end organs affected. While the pathological and histological appearance may remain the same, the variance occurs with the size of the vessels involved and the end organs involved. For example, when large vessels are involved you have conditions such as Behcet's disease, which involve painful ulcers in the mouth and genital area with the uvia of the eyes often involved. It is usually specific to men in their 30's and of Mediterranean descent.
- Another is Cogan's syndrome which is more systemic and can lead to skin changes and eye inflammation and even hearing
 loss. Giant Cell arteritis occurs in the artery of the temple of our head and usually affects older folks. Headaches, scalp tenderness
 and pain are harbingers of this condition and without treatment can lead to sudden vision changes or loss.
- Polymyalgia Rheumatica (PMR) affects the joints and muscles causing pain, stiffness and weakness, mostly of the proximal muscle groups of the shoulder and pelvic girdle. Takayasu's arteritis (KA) while rare usually affects Asian teenagers and women. KA can cause destruction of the aortic arch. Malaise, fever, weight loss and sore joints usually precede vessel involvement.

Cause:

- Medium sized vessels are affected in a few other disorders such as Buerger's disease and Raynaud's phenomenon. Buerger's is also known as thromboangitis obliterans which can cause disruption of blood flow to fingers and toes with necrosis and loss of those affected limbs. Seen very often in men in their 30's and associated with cigarette smoking and elevated serum homocysteine levels. Raynaud's is a commonly seen malady where extremities such as fingers get cold and turn "white" with decreased blood flow when exposed to a trigger (like touching a cold object).
- A rare childhood disease known as Kawasaki disease (KD) where the walls of vessels systemically are inflamed and all sizes are involved and can be damaged with aneurysms. Also known as muscocutaneous lymph node syndrome as a child presents very often with red patches on mucous membranes and a red rash on the skin. Some serious cardiac issues may occur in these children if not treated early. It is successfully treated with medication if caught early and rarely do surgeons get involved to repair enlarged weak blood vessels.
- Polyarteritis nodosa can present with lace like rash, bumps under the skin, anemia is yet another manifestation of a vasculitis. Other conditions such as Eosinophilic granulomatosis with polyangiitis (EGPA) aka Chung-Strauss Syndrome, affecting skin, lungs, kidneys and nervous system. Cryoglobulinemic vasculitis, IgA vasculitis (Henoch-Schonlein Purpura), hypersensitivity vasculitis and microscopic polyangiitis are others. Wegener's granulomatosis (GPA) is still another variety that affects the upper airway, lung and kidney.
- Signs and symptoms can include fever, loss of appetite, weight loss, fatigue and generalized pain. Very often there are skin manifestations. Organs involved can include skin, joints, the lung, the GI tract, upper airway tree (nose ears), eyes, the brain and nervous system tissue. It can be quite disturbing for a parent to find their child with a very high fever, mucous membrane lesions and peeling skin. These are some classic signs of Kawasaki Disease, a rare type of vasculitis in children.

Work Up

Making the diagnosis:

Tests that are used to diagnose vasculitis are: CBC to rule out anemias; an antineutrophilic cytoplasmic antibodies test (ANCA) where certain antibodies are produced in some vasculitis. The Erythrocyte Sedimentation Rate (ESR, aka Sed Rate) is a very common and important test in evaluations. Another inflammatory marker often ordered by doctors is the C-reactive Protein (CRP).

As an inflammatory disease, inflammatory biomarkers are important tests for diagnosing these disorders. Sometimes biopsy of an affected vessel is important as histologic evaluation can help make a diagnosis. Other studies such as an EKG and ECHO to measure cardiac function, a Chest x-ray for pulmonary issues and a urinalysis to check for renal function and involvement are ordered. Ultrasound, CT and MRI scans may be helpful as well as angiography which visualizes blood vessels that may be narrow, swollen, deformed or obstructed.



Incorporating a holistic approach utilizing natural treatments and lifestyle changes will build up the immune system while alleviating fatigue and other symptoms.

Antioxidants are found in dark colored berries, dark leafy vegetables, and can be obtained through vitamin supplements such as vitamins A, C, and E. Another very useful antioxidant for HSP is coQ10, which can directly reduce the inflammation of the endothelial lining of the blood vessel wall.

A major class of antioxidant compounds are flavonoids, which are found in berries, green tea, and citrus fruits. A 2012 study demonstrated that flavonoids isolated from a Chinese herbal medicine *Bidens bipinnata* reduced inflammatory messengers in the serum of HSP patients, including interleukin-8, tumor necrosis factor-alpha, and nitric oxide. These inflammatory messengers are usually high in the blood of HSP patients. The flavonoids from this plant also reduced the expression of NF-kB, which is an upstream regulator of many inflammatory messengers.

Fish oil is high in Omega-3 Fatty Acids which have been shown to reduce inflammatory cytokines in the body by enhancing the production of anti-inflammatory prostaglandins. A 2004 study showed that fish oil supplementation combined with an ACEI hypertensive medication helped protect the kidneys of children with HSP by reducing the amount of protein lost in the urine.



Treatment

Treatments in MSM



- Treatments vary according to what type and the severity of the disease. Also
 considered in a medical regimen is what end organs are involved. Most treatment is
 by medication, but on occasion surgery is called for. Many can be treated with overthe-counter pain medications in the class called NSAIDS (Motrin, Aleve or aspirin).
- Other choices are corticosteroids like prednisone. Some physicians may prescribe cytotoxic medications. These types of medications will suppress or kill cells that cause inflammation. Some examples are cyclophosphamide, methotrexate and azathioprine.
- In pediatric cases of Kawasaki disease, high doses of aspirin are used in combination with intravenous immune globulin. To repair bulging vessels, vascular surgeons may have to go in and remove or repair aneurysms.



Treatments in MSM



- There are no set guidelines for prevention of vasculitis, however, keeping a healthy immune system, proper diet, avoiding environmental toxins and stimulants such as tobacco smoking and doing what it takes in higher risk patients to avoid triggers can help lower the risk of getting this disorder.
- As wellness professionals, being observant with clients when they are ill appearing and may present with skin manifestations, should be directed to have an evaluation.



Alternative Therapies



NATURAL THERAPIES

Follow an anti-inflammatory diet as well as the measures listed below that can bring about dramatic improvement in patients with autoimmune disorders:

- Eliminate cow's milk and cow's milk products (substitute other calcium sources)
- Eat more fruits and vegetables (make sure that they are organically grown)
- Eliminate polyunsaturated vegetable oils, margarine, vegetable shortening, all partially hydrogenated oils, all foods (such as deep-fried foods) that might contain trans-fatty acids. Use extra-virgin olive oil as your main fat

 Increase your intake of omega-3 fatty acids. Take two to three grams of a molecularly distilled fish oil supplement each day

• Take anti-inflammatory herbs such as ginger and turmeric. Dr. Weil recommends unsweetened turmeric tea, as well as a combination of ginger, turmeric, and other botanicals with anti-inflammatory properties.

[Note: for those interested in a more natural approach to therapy for vasculitis let me suggest: Dr. Andrew Weil weighs in with the above]



Alternative Therapies



NATURAL THERAPIES

Consider taking grape seed extract, a source of powerful antioxidant compounds called OPCs (oligomeric proanthocyanidins). Research has shown these compounds to be useful in protecting blood vessels, making them more elastic and less likely to leak.

EarthClinic suggests: Apple Cider Vinegar Tonic - ACV Tonic is a wonderful natural remedy for many of the vasculitis symptoms, with reported natural cures for, among other ailments, chronic fatigue, flu and allergies (an immune system problem). The acetic and malic acids in ACV have antiviral, antibiotic and anti-fungal properties. It's important to use natural organic apple cider vinegar with the 'mother' as that contains the healing enzymes.

Herbal Remedies - Garlic, Echinacea and Goldenseal are all excellent herbal treatments to boost the immune system. Garlic is particularly useful because of its anti-inflammatory and astringent properties. In addition, garlic is a useful home remedy as it has effective antibacterial, antiviral and anti-fungal properties. In addition to strengthening the immune system, Echinacea enhances lymphatic function while Goldenseal is an antibacterial that also detoxifies and cleanses the body.



Whole Body Health



LDN Conditions that are helped by Low Dose Naltrexone (LDN) LDN Research Trust - The Low Dose Naltrexone Charity ITP made the list under HemOnc list

Rapamycin a Very Promising Therapy: AntiInflammatory and promotes autopagy by impact on mTOR **Amlexanox** a possibility: <u>Kinase inhibition in autoimmunity and inflammation - PMC (nih.gov)</u>

So, if one of MY patients has a vasculitis and did not want HD-prednisone or biologics; I would select LDN + Rapamycin and possibly Amlexanox (in effort to avoid using HD-prednisone)

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