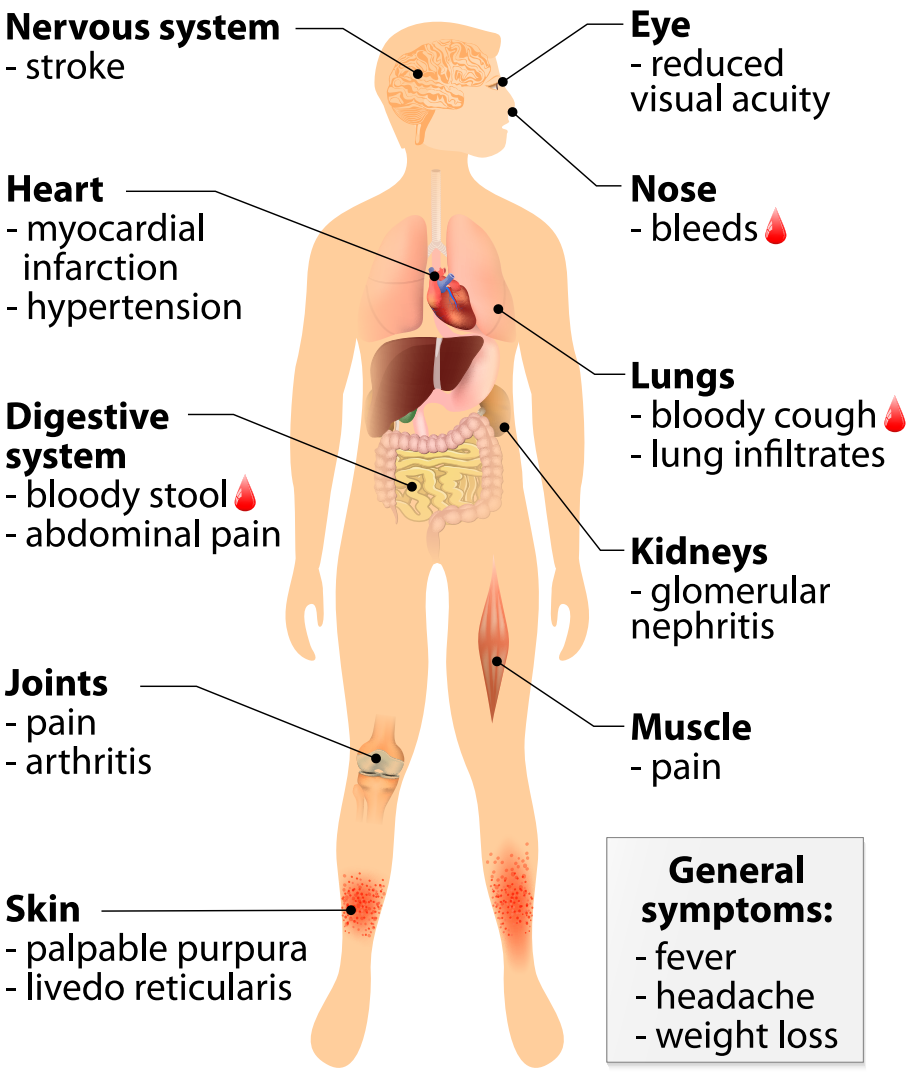
Vasculitis and the dental team

By **John Mills**[1]



What is vasculitis?

Vasculitis means, literally, inflammation of the lining of the blood vessels and is an umbrella term covering illness ranging from acute localised hypersensitivity reactions to severe, auto-immune systemic, incurable life-threatening diseases.

Primary Systemic Vasculitis (PSV) is an auto-immune disease where neutrophils in the blood become activated and start to attack the cells lining the blood vessels. The resulting inflammation of the blood vessel walls causes blood flow to be restricted or for vessels to be blocked, leading to damage or necrosis of the tissues or organs that they supply. Small vessels and capillaries become leaky – resulting in the characteristic rashes and purpura of vasculitis.

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The disease is incurable but can usually be controlled by medication and eventually brought into remission. There are 18 different types of PSV, some of which are extremely rare. They are classified by the size of vessels affected:

- Large (aorta and main arteries and veins)
- Medium (eg renal and gastro-intestinal arteries)
- Small (arterioles, venules and capillaries).

This last group particularly affects capillary beds in the lungs, kidneys, skin and mucosa – leading to necrosis and ulceration.

Dental patients

Members of the dental team can have especially good opportunities to influence the health and well-being of patients well beyond just their oral health. Unfortunately, years of increasing specialisation and 'payment by results' has tended to focus the attention of dental professionals ever more closely on the oral cavity and less on the patient as a whole. However, we all know the importance of having a comprehensive, accurate and up to date medical history as a means of getting a holistic picture of the patient.

Most 'healthy' people visit their dentist more frequently than they do their GP, so a carefully taken and thoughtfully scrutinised medical history taken in the dental practice may reveal early signs of unsuspected disease. In addition, many whole body, systemic illnesses have oral or facial signs and symptoms that, if recognised, may lead to the diagnosis of an otherwise unrecognised condition. Certain types of vasculitis fall into this category.

Types of vasculitis

Some of the most aggressive types of vasculitis are associated with a particular circulating antibody, anti-neutrophil cytoplasmic antibody - or ANCA. This type of vasculitis is relatively rare, with an annual incidence of around 1,200 new cases in the UK. This category includes Wegener's Granulomatosis (now called GPA), Churg Strauss syndrome (now EGPA) and Microscopic Polyangiitis (MPA).

In general terms, people of all ages and either gender can be affected, but some types affect predominantly either males or females. Some types such as Henoch Schonlein Purpura (HSP) affect mainly children; others such as Giant Cell Arteritis, mainly elderly patients.

Single or multiple organs can be affected by vasculitis and may include lungs, trachea, kidneys, skin, eyes, ears, joints, nose and sinuses, endocrine glands, heart, brain, central and peripheral nervous system.

Oral and facial signs

Several types of vasculitis have characteristic oral or facial symptoms that if recognised by someone in the dental team may lead to an early diagnosis.

The three types of vasculitis with most distinctive oro-facial signs are Wegener's Granulomatosis, now called Granulomatosis with Polyangiitis (GPA), Giant Cell Arteritis (GCA) and Behçet's Syndrome.

Wegener's Granulomatosis

Patients may have recurrent or persistent eroding ulcers on lips and mucosa which may become large. They frequently complain of nasal and sinus problems, such as persistent stuffy nose and blocked sinuses with nose bleeds and nasal crusting. This can progress to destruction of the nasal septum resulting in collapse of the bridge of the nose – the characteristic saddle nose. Vessels supplying teeth and periodontal tissues may be restricted or occluded resulting in phantom toothache and strawberry gingivitis. This may result in pulpal necrosis without obvious reason and periodontal damage. Eye signs may be red or bloodshot eyes due to scleritis or bulging eye due to formation of granulomas behind the eyeball.

Giant Cell Arteritis

Elderly patients complaining of headache or visual disturbance may show characteristically prominent veins in the temporal area. Patients may complain of scalp tenderness, but most significantly may complain of pain in the jaw or tongue during mastication.

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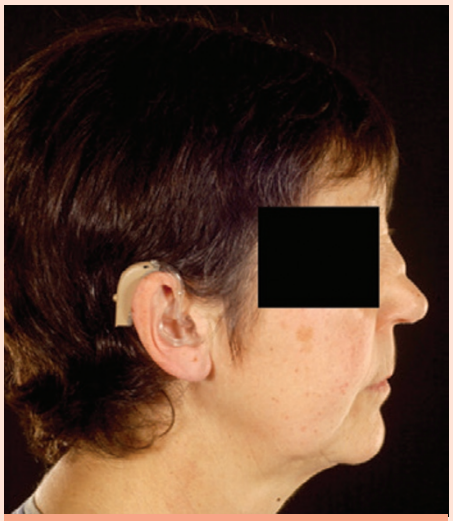

Fig. 1 Collapse in the bride of the nose caused by granulomatosis with polyangiitis (Wegener's). Note the hearing aid required for hearing loss due to concurrent middle ear disease

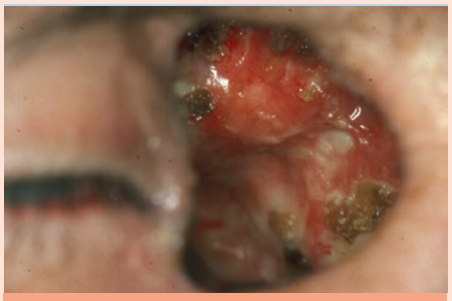

Fig. 2 Untreated GPA (Wegener's) eroding through the skin between the nose and the right eye

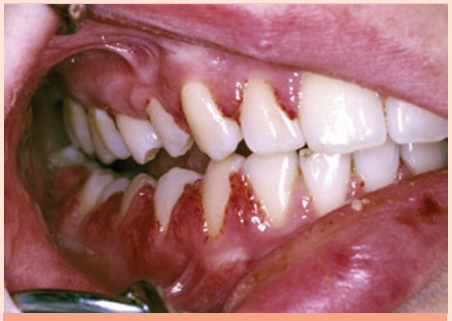

Fig. 3 'Strawberry gingivitis'. GPA (Wegener's) causing vasculitis of the gums

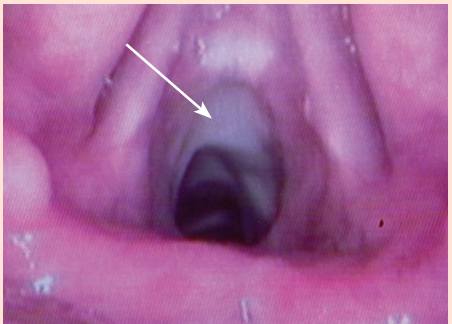

Fig. 4 Subglottic stenosis in GPA (Wegener's). Note the narrowing to the airway (arrowed) just below the larynx

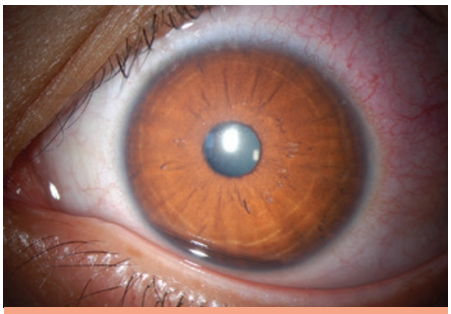

Fig. 5 Episcleritis in GPA

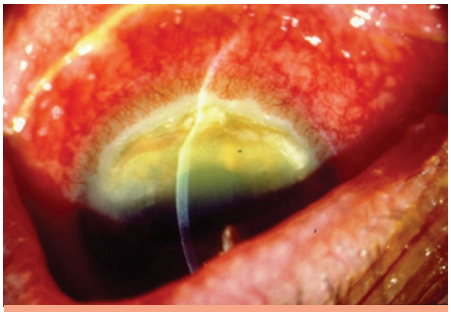

Fig. 6 Severe scleritis causing destruction of the cornea in GPA (Wegener's)

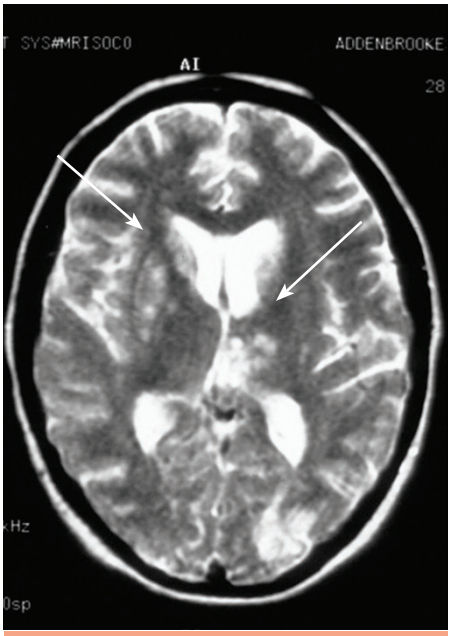

Fig. 7 Primary cerebral vasculitis confirmed by brain biopsy. Arrows indicate abnormal areas

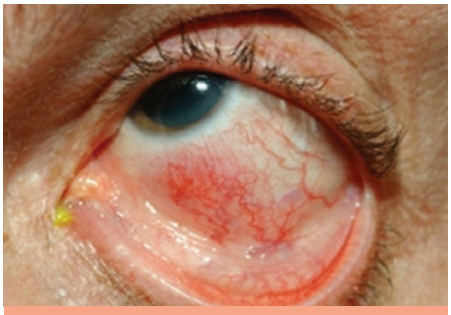

Fig. 8 Scleritis

Behçet's Syndrome

Persistent oral ulceration is very characteristic of this type of vasculitis and may be the first sign of the disease. The ulcers start as raised erythematous swellings which then break down into an ulcer with a grey pseudo membrane over. Usually on the mucosa, they may spread to the hard palate, pharynx and tonsils. Even worse are the associated genital ulcers which can be more painful and deeper than the oral ulcers. They can be very resistant to treatment. There may be associated facial acne and uveitis in the eye.

'VASCULITIS IS SOMETIMES ASSOCIATED WITH OTHER DISEASES SUCH AS CANCER OR RHEUMATOID ARTHRITIS.'

Treatment

Treatment is usually in two phases. Initial induction is with powerful immune-suppressing drugs such as cyclophosphamide, a chemotherapy drug most frequently administered as a series of six or eight pulsed IV infusions in conjunction with high dose steroids (prednisolone).

Once controlled, the disease is held in check by a maintenance regime using less powerful immune suppressing drugs (azathioprine, methotrexate, mycophenolate) in conjunction with lower dose prednisolone. This may continue for several years until full remission is achieved, but even then, relapse is common.

Prior to the introduction of cyclophosphamide in the early 1970s, some types of vasculitis such as Wegener's were invariably and rapidly fatal. However, cyclophosphamide has very much a 'scatter gun' effect, damaging all cells to some extent, thus causing serious side effects, such as loss of fertility, myeloid leukaemia and bladder carcinoma.

A more recent 'targeted' treatment regime, based on a manufactured monoclonal antibody, rituximab, is being used increasingly for both induction and maintenance. Rituximab suppresses the B cells in the bone marrow that produce the rogue neutrophils. It does not affect fertility and is very effective for treating relapse.

Members of the dental team should be very conscious of the consequences and side effects of treatment. The principal aim of treatment is to suppress the immune system, but of course this renders patients more vulnerable to infections. In some cases it causes neutropenia, a severe lack of white blood cells and thromcocytopenia, with consequent risk of clotting problems.

After the induction phase of treatment, patients may take other less potent immune suppressing drugs as maintenance therapy for many years. When checking lists of medication, names of drugs to look out for are azathioprine, methotrexate and mycophenolate (Cellsept).

Glucocorticoid steroids (such as prednisolone) in high doses are an initial key element in bringing active vasculitis disease under control. Prolonged use of artificial steroids suppresses natural cortisol production, with the result that the body cannot cope well with physical stress such as dental extractions, with consequent risk of surgical shock.

In addition, repeated use of high dose steroids or long term low dose steroid use may induce diabetes and osteoporosis. To counteract the osteoporosis risk, patients are frequently prescribed bisphosphonates. These affect bone metabolism, reducing osteoclast activity, but in the dental context the reduced vitality of the bone may increase the risk of serious postoperative infection.

Vasculitis is sometimes associated with other diseases such as cancer or rheumatoid arthritis. Other auto-immune diseases such as systemic lupus and Sjögren's disease can have a cross-over and thyroid disease or pituitary disease can be associated.

The early recognition and diagnosis of vasculitis can have a profound influence on the outcome. Many signs and symptoms are non-specific, but taken together can join up the dots and make a picture. The dental team can help to supply some of those dots that complete the picture.

The full range of dental implications and consequences of vasculitis is not well researched and would make a good subject for a postgraduate study.

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