Ultrasound showed high diagnostic accuracy. Modern PET equipment may also delineate vasculitic temporal, maxillary and vertebral arteries. The significance of follow-up imaging studies in LVV still needs to be determined.

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SP0073 DIAGNOSIS OF GASTROINTESTINAL VASCULITIS

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The gastrointestinal vasculature can be involved by a variety of vasculitis affecting large, medium, small or variable vessel size according to the 2012 revised Chapel Hill consensus nomenclature. Gastrointestinal involvement (GI) is one of the most severe complications of vasculitis requiring intensive immunosuppressive therapy and sometimes revascularization techniques or emergency surgery and may affect a variety of organs including large or small intestines, liver, biliary tract, pancreas, and less frequently, stomach or oesophagus. It can complicate the course of patient with known or with overt suspicion of vasculitis or be the main or even the only manifestation of these diseases.

Clinical manifestations range from abdominal pain, sometimes colicky, that usually worsens after eating to overt signs of bowel ischaemia or perforation with diffuse and constant abdominal pain, reduced peristalsis, abdominal defence and distension, and signs of peritoneal irritation. GI bleeding, frequently in the form of occult blood loss or haematochezia and less frequently melena or rectorrhagia, is common. Non-specific symptoms such as nausea, vomiting or diarrhoea may be observed. Pancreatitis, cholecystitis or appendicitis may also occur.

In large-vessel vasculitis, including giant-cell arteritis (GCA) and Takayasu disease (TAK), inflammation usually affects the proximal portion of the celiac trunk, hepatic or mesenteric arteries. GI involvement is rare in GCA with only isolated cases or small series having been reported. About 30% of patients with GCA have elevated hepatic enzymes as part of the acute phase response and rarely reflect involvement of the hepatic vasculature. Imaging techniques disclose stenotic or occlusive lesions in 25% of patients with TAK but clinically manifest involvement occur in about 16% of patients. Abdominal pain suggestive of vascular insufficiency is present in about 16% of patients; abdominal bruits can be perceived in 14% but only 4% present with overt signs of mesenteric ischaemia. Due to the involvement of the proximal part of the GI branches, the ischemic territories may be extensive but usually progression of vascular occlusion is not abrupt, and due to the size of vessels involved, patients may benefit from percutaneous revascularization procedures if immunosuppressive treatments are not sufficient. Interestingly 5% of patients with TAK also suffer from inflammatory bowel disease, particularly Crohn's disease.

In medium sized vasculitis, such as polyarteritis nodosa (PAN) or Kawasaki disease (KD), multiple muscular artery branches of the GI vasculature can be involved. Abdominal pain is frequent in PAN (35-95% of cases). GI involvement is more frequent in HBV-associated PAN than in idiopathic PAN. About 61% of patient with KD may have GI symptoms but these are severe in 4.6% only. Gallbladder hydrops and appendicular vasculitis may occur. The presence of prominent GI manifestations is more frequent in patients with coronary aneurysms indicating more severe vasculitic disease.

In small-vessel vasculitis, including anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV)(granulomatosis with polyangiitis [GPA], microscopic polyangiitis [MPA], and eosinophilic granulomatosis with polyangiitis [EGPA]) and immune-complex-mediated vasculitis (IgA vasculitis [Henoch-Shönlein purpura] and cryoglobulinemic vasculitis) the microvasculature is segmentally affected and the severity of involvement may be variable in different areas. Among AAV, GI involvement is more frequent in patients with EGPA (35-50%), than MPA (5-30%) or GPA (5-11%). In EGPA, GI involvement can be vascular as in other vasculitis or may be in the form of eosinophilic gastroenteritis or esophagitis. In GPA, in addition to vascular involvement, granulomatous hepatitis or pancreatitis may occasionally occur. Among immune-complex vasculitis, GI involvement is typical in IgA vasculitis, occurring in 50-75% of patients. Although it is usually mild and

self-limited, complications including bleeding, ischaemia, perforation or intussusception may appear in 3-5% of patients. Hepatobiliary involvement is uncommon (1.8%). GI involvement is rare in cryoglobulinemic vasculitis but is life-threatening when it occurs.

Among the category of variable vessel size vasculitis, GI involvement in Behçet's disease (BD) may consist of mucosal ulcers, frequently in the distal ileum, similar to inflammatory bowel disease, or involvement of mesenteric artery branches as in other vasculitis. Venous thrombosis involving the liver vasculature or GI tract may also occur in BD.

Vasculitis involvement may be restricted to different portions of the GI tract (gallbladder, appendix, intestines) in patients with no obvious evidence of systemic vasculitis. Gallbladder is the more frequent location and is usually incidentally discovered after surgery. Overt systemic vasculitis may develop in up to 25% of patients during follow-up.

Diagnosis of GI involvement in systemic vasculitis requires clinical suspicion. Imaging (computed tomography [CT]-angiography[CTA] or magnetic resonance angiography [MRA] are crucial in large vessel vasculitis evidencing concentric thickening with contrast enhancement of the vessel wall, along with lumen reduction. In medium vessel vasculitis CT may disclose hepatic, spleen or renal infarcts that reinforce clinical suspicion of vascular disease. However, CTA or MRA may not have enough resolution to evidence involvement of medium-sized branches and conventional angiography is usually needed, revealing vascular stenosis or occlusions as well as microaneurysms. In small-vessel vasculitis, and in all vasculitis conveying intestinal ischemia, CT may reveal thickening of the intestine wall, dilatation, and, in severe cases, pneumatosis of the intestine wall as well as gas in the portal system. Pneumo-peritoneum can be seen in the case of perforation. Ascites is not uncommon in medium and small vessel vasculitis. Occasionally, hemorrhage in the peritoneal cavity due to rupture of aneurysms can be observed. It is important to remark that imaging findings provide strong support to the clinical suspicion of vasculitis based on additional clinical, blood test or serologic findings or evidence of vasculitis elsewhere but similar findings can be observed in the context of a variety of vasculopathies or thrombophilic/embolic disorders that may mimic vasculitis.

Endoscopy is also a useful exploration in some settings. It may detect intestinal purpura, usually in IgA vasculitis, mucosal ulcers or mucosal inflammation. Endoscopic biopsies may reveal eosinophilic inflammation in EGPA but may not be deep enough to disclose inflamed blood vessels and its absence does not exclude vasculitis. In case of surgical complications, a throughout pathologic examination of the removed specimen should be requested in order to evidence of vascular inflammation.

GI involvement in systemic vasculitis is frequently a severe and potentially life-threatening complication. Prompt detection and intense immunosuppressive, revascularization or surgical treatment in the appropriate settings are crucial for a successful recovery and prevention of irreversible damage.

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SP0074 DIAGNOSIS OF CEREBRAL VASCULITIS

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Among all forms of vasculitis, perhaps that affecting the CNS (CNS-V) poses the greatest challenge in diagnosis given the lack of any non invasive high specificity test, the complex and extensive list of mimicking conditions, its inaccessibility for biopsy and its genuine rarity. The diagnosis still requires the presence of an unexplained neurologic sign or symptom(s) following an exhaustive evaluation, evidence of vascular involvement (by direct or indirect angiography) or biopsy and most importantly the meticulous exclusion of all those conditions capable of producing mimicking clinical, radiographic or histologic findings that would confound accurate diagnosis. Despite these challenges CNS-V has been increasingly reported due to a combination of increased diagnostic awareness and advances in diagnostics. Two of these major advances will be discussed including the use of direct vascular wall imaging to differentiate vascular inflammation from spasm or atherosclerosis and the use of next generations sequencing to identify infectious etiologies and obviate the use of biopsy. Clinical examples will be presented.

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THURSDAY, 13 JUNE 2019

15:30:00 – 17:00:00

How to manage and treat childhood onset lupus? A multidisciplinary point of view

SP0075 CASE PRESENTER: CHILDHOOD ONSET OF NEUROLUPUS

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Background: In the series of talks 'How to manage and treat childhood onset lupus? A multidisciplinary point of view' the first talk focuses on achievement of the initial diagnosis of Lupus within the childhood period, including some potential pitfalls and how these can be avoided. Initial and ongoing treatment within the childhood period, alongside paediatric specific therapeutic considerations will be discussed. The role of the multidisciplinary team of doctors, nurses, physiotherapists, occupational therapists, psychologists and sub-specialists will be highlighted. The impact of the diagnosis on the child, family, education and social activities will also be described. This talk will be followed by a subsequent talks following the patient through transition and into adulthood.

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SP0076 CASE PRESENTER: TRANSITION OF NEUROLUPUS PHASE INTO ADULTHOOD

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Childhood-onset SLE (cSLE) accounts for approximately 20% of all SLE cases and is often regarded as one of the most complex rheumatic diseases. With the increase of five-year survival rate of cSLE patients to over 95%, there is a growing number of adolescents and young adults (AYA) transferring from pediatric to adult care. Even in the best-case scenario, there is often considerable challenges regarding this transition. Hence, it is not surprising that morbidity, mortality and disease activity in cSLE, as well as in many other chronic illnesses and conditions, worsen during or just after the transition. The reasons for these deteriorations are numerous, with most prominent arising from the nature of the disease and changes during the tumultuous period of adolescence, but also the differences between pediatric and adult clinical and healthcare settings. Besides, the AYA patients with chronic conditions often feel they are not well prepared for the transition to adult care, while the adult rheumatologists commonly report concerns about assuming the responsibility of patients with the pediatric-onset disease. Some of these concerns are probably motivated by increased vulnerability of AYA patients with cSLE, who are more likely than their adult counterparts to develop lupus nephritis and neuropsychiatric manifestations, as well as other organs involvement and atherosclerosis. Moreover, due to the severity of the disease, patients with cSLE are treated more aggressively, accumulating more drug-related toxicity than patients with adult-onset SLE, which results in significant SLE-related damage and complications such as osteoporosis during the childhood. On the other hand, there is decreased medication adherence in AYA, further complicated by the neurocognitive and memory impact of the disease, as well as high rates of comorbid depression and anxiety. Some additional problems in cSLE patients at transition age are sexuality, fertility, and pregnancy. All this makes patients with cSLE require specialized and multidisciplinary care at the transition, that is capable of addressing medical, psychosocial, educational and vocational needs.

This presentation will be part of the comprehensive session discussing distinct features of the SLE during the different phases of life. It will emphasize specific challenges of the transition from pediatric to adult care, with the use of a compelling clinical case as an example.

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SP0077 CASE DISCUSSANT

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Background: Padiatric-onset systemic lupus (pSLE) is a severe multisystem autoimmune condition which mostly manifests after the age of 10 years. It is mainly a polygenic disorder, but a mutation in a single gene may predispose to the development of SLE (« monogenic lupus »). We will describe some unusual presentations which may lead to a delayed diagnosis or misdiagnosis and provide clues for the diagnosis of monogenic lupus.

Objectives: To discuss the difficult diagnosis and pitfalls in pSLE

Results: The most common initial manifestations of pSLE are cutaneous, musculoskeletal, renal, hematologic, and fever. However, atypical symptoms may reveal the diagnosis or occur during the course of SLE. In a French retrospective study¹, thirty-two percent of children had atypical symptoms, mainly including abdominal involvement in 26 patients, which lead to negative laparotomy for presumed appendicitis before the diagnosis of SLE could be established. Abdominal pain was related to lupus pancreatitis or peritonitis. The diagnosis of pSLE should be systematically searched for in a teenager who presents with unexplained mood disorders, psychosis or depression, which may reveal pSLE. In a patient