Polyarteritis Nodosa

Subjects: Rheumatology
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Polyarteritis nodosa (PAN), also known as panarteritis nodosa, represents a form of necrotizing vasculitis that predominantly affects medium-sized vessels, although it is not restricted to them and can also involve smaller vessels. The clinical presentation is heterogeneous and characterized by a significant number of patients exhibiting general symptoms, including asthenia, fever, and unintended weight loss. Although PAN can involve virtually any organ, it preferentially affects the skin, nervous system, and the gastrointestinal tract. Orchitis is a rare but specific manifestation of PAN. The absence of granulomas, glomerulonephritis, and anti-neutrophil cytoplasmic antibodies serves to distinguish PAN from other types of vasculitis. Major complications consist of hemorrhagic and thrombotic events occurring in mesenteric, cardiac, cerebral, and renal systems. Historically, PAN was frequently linked to hepatitis B virus (HBV) infection, but this association has dramatically changed in recent years due to declining HBV prevalence.

PAN polyarteritis nodosa panarteritis nodosa monogenic VEXAS DADA2

1. Introduction

PAN was first described in 1866 by Kussmaul and Maier. They reported an “intermittent nodular appearance affecting arteries throughout the body, sparing large vessels (the aorta and its branches), small vessels (arterioles, capillaries, and venules) and pulmonary vessels” [1]. The distinct “pearl necklace” pattern led to the naming of this condition as polyarteritis nodosa. In 1952, pathologist Pearl Zeek laid out the first classification of PAN, distinguishing between hypersensitivity vasculitis, allergic vasculitis, PAN, and temporal arteritis, now, respectively, known as urticarial vasculitis, eosinophilic granulomatosis with polyangiitis (Churg–Strauss syndrome), PAN, and giant cell arteritis (Horton’s disease) [2]. A pivotal development came in 1982 when autoantibodies directed against neutrophil cytoplasm antigens (ANCA) were identified in eight patients with clinical characteristics of vasculitis, introducing ANCA-associated vasculitis (AAV) as a distinct category of vasculitis separate from PAN [3]. The most recent definition of PAN came from the 2012 Chapel Hill Consensus Conference (CHCC) where the disease was described as “necrotizing arteritis of the medium or small arteries without glomerulonephritis or vasculitis of arterioles, capillaries or venules and without ANCA” [4][5][6]. Over the past two decades, the medical community’s understanding of PAN has significantly evolved. While initially described as either primary vasculitis or Hepatitis B virus (HBV)-related, the current understanding recognizes PAN as a disease often secondary to genetic syndromes and malignant hematologic disorders [7].

2. Epidemiology

The prevalence of PAN varies greatly across different countries, ranging from 2 to 31 per million inhabitants in Europe [8][9]. There are notable north–south and seasonal gradients in the occurrence of the disease. Historically, PAN was frequently linked to HBV infection. With the advent of its vaccine and the enhancement of public health measures in response to the AIDS crisis, the incidence of PAN has seen a significant decline, turning it from one of the most common vasculitis in the 1990s to one of the least common today [8][10]. In addition, the improvement in laboratory techniques for the detection of ANCA in the 1980s led to the reclassification of certain vasculitis that were initially diagnosed as PAN [11]. The challenge in estimating the overall prevalence of PAN arises from a combination of factors, including the absence of serum markers, heterogeneous classification criteria, and a variety of predisposing genetic and environmental factors.

3. Physiopathology

PAN is typically characterized by a segmental, necrotizing, and transmural inflammation, predominantly involving small- to medium-sized arteries, although any arterial size could theoretically be susceptible. The disease most commonly impacts the visceral and muscular arteries, including their branches. In patient biopsies, it is common to observe co-existing lesions of diverse stages of inflammation and scarring within a single sample [12]. Because of the arterial inflammatory process, fibrinoid necrosis may develop, leading to the formation of microaneurysms. Over time, these complications can progress to chronic stages, characterized by fibrous scarring and vascular aneurysms, which can rupture and lead to severe bleeding [13][14]. During the acute phase, the cellular infiltrate, composed of macrophages, T lymphocytes, neutrophils, and eosinophils, is generally observed in the tunica media but can also invade the tunica interna and tunica externa [15]. One distinctive feature of PAN, compared to other vasculitis, is the absence of granulomas. This disease is also characterized by the coexistence of different stages of vascular inflammations at the same time.

The pathophysiology of PAN, not yet fully understood, may vary depending on the disease's specific etiology. Serum cytokine profile analysis in PAN patients has revealed an elevation in interferon-alpha (IFN-α), interleukine-2 (IL-2), tumor necrosis factor-α (TNFα), and IL-1-ß compared to healthy individuals and those with granulomatosis with polyangiitis (GPA) [16]. Immunohistochemical studies of muscle and nerve biopsies from patients showed the presence of macrophages (41%) and mostly CD4+ T lymphocytes (41%) [13][17]. Most of these studies, however, primarily focus on PAN associated with HBV.

Viral infections remain a common trigger of PAN and should be excluded in all cases. In PAN associated with HBV, the HBs antigen is responsible for the formation of immune complexes [18][19], as suggested by animal models of hepatitis B antigen-associated PAN, which show an accumulation of immune complexes in blood vessels [20][21]. Hepatitis C virus (HCV) has also been linked to PAN, with HCV-associated PAN tending to present more severe and acute symptoms [22][23]. However, this only concerns 5% of patients with PAN, and the distinction with cryoglobulinemic vasculitis can sometimes be challenging [24]. HIV infection has been associated with PAN, though HIV-associated PAN is generally less aggressive than HBV-associated PAN. The classical manifestation is mononeuritis multiplex and can occur at any stage of HIV infection [25]. Although parvovirus B19 has been

associated with PAN, a study using PCR tests found no higher prevalence of this infection in people with PAN compared to those without [26][27][28][29].

More recently, vasculitis has been associated with COVID-19 infection, but to date no cases of PAN have been reported [30][31]. COVID-19 vaccines have been associated with PAN manifestations [32][33][34]. Like other vasculitides, PAN can be induced by the use of certain drugs, such as minocycline [35]. The association between PAN and neoplasia is well established, especially for hematological malignancies, such as hairy T cell leukemia, or, more recently, myelodysplastic syndrome (MDS) [36][37][38][39][40].

More recently, genetic forms of PAN have been described. In the early 2000s, cases of PAN-like vasculitis were described in patients with Familial Mediterranean Fever (FMF) [41][42]. In a nationwide study in Turkey, PAN prevalence in patients with FMF was 0.9% [43]. FMF is caused by mutations in the MEFV gene that encodes for pyrin/marenostrin, which result in unregulated production of IL-1, leading to recurring inflammation, fever, and, sometimes, autoimmune manifestations [44]. Patients with PAN associated with FMF present a higher incidence of perirenal hemorrhages and elevated levels of inflammation [41][42][43][45]. Another condition related to genetic forms of PAN is STING-associated vasculopathy, with onset in infancy (SAVI), which is a type I interferonopathy. This condition is caused by mutations in the TMEM173 gene that induce the inflammation of endothelial cells in children. It often presents PAN-like symptoms in affected children [46]. A monogenic syndrome resulting from a deficiency in Adenosine Deaminase 2 (DAD2) has been described in familial cases of necrotizing vasculitis that resemble PAN [47]. Since 2014, over 60 bi-allelic loss-of-function mutations in the ADA2 gene have been documented [48][49]. Vascular inflammation in DAD2 patients is believed to be caused by an imbalance in macrophages, favoring the M1 type over the M2 type.

4. Clinical Manifestations

Signs and symptoms of PAN result from damage to the vascular walls, potentially affecting all organs. This section provides an overview of organ systems that can be impacted in PAN patients. Unless otherwise stated, the percentages and specifics of the manifestations come from the cohorts shown in **Table 1**.

Table 1. Characteristics of PAN patients reported in different cohorts (PNP: peripheral neuropathy, CNS: central nervous system, FFS: Five Factor Score, 1996). Results are expressed as percentages.

Characteristics	Pagnoux et al. (1963 to 2005) [50]	Sönmez et al. (1990 to 2015) [10]	Rohmer et al. (2005 to 2019) [50]	Georgin-Lavialle et al. (VEXAS) [51]	Meyts et al. (ADA2) [52]
General symptoms	93.1		85	95.7	50

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Fever	63.8	53.7	54	64.6	
Loss of weight	69.5	53.7	50	54.5	
Myalgia	58.6	46.2	50		
All cutaneous	49.7	67.2	59	83.6	75
Nodules	17.2				14
Purpura	22.1				
Livedo	16.7	17.9			50
Panniculitis			7.5	12.9	
Renal	50.6	47.7	20	9.5	
Hematuria	15.2				
Proteinuria	21.6				
Hypertension	34.8	41.7			21
Orchitis	17	14.9	16		4
Neurologic	79.0	43.2	59		

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PNP	74.1			5.2	9
Mononeuritis	70.7			2.6	
CNS	4.6				53
Digestive	37.9	22.3	28	13.8	33
Abdominal pain	35.6	37.3		8.6	12
Bleeding	3.4			0.9	
Perforation	4.3			0.9	2
Cardiovascular	22.4		39		
Pericarditis	5.5			4.3	
Distal necrosis	6.3	13.5			22
Thrombo-embolism				35.3	
Ophthalmic	8.6			40.5	

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Retinal vasculitis	4.3				
Pulmonary		2.9	8	49.1	
Cough	5.7				
Lung infiltrate	3.4			40.5	
Pleural effusion	3.4			9.5	
Chondritis				36.2	
Arthralgia	48.9	58.2		28.4	
Arthritis		17.9			

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5. Treatments

The treatment recommendations for PAN are primarily based on weak empirical evidence and are often drawn from recommendations for other forms of vasculitis, with modifications according to the disease severity. Mild PAN, characterized by non-life- or organ-threatening manifestations like constitutional symptoms, arthritis, or skin lesions, is differentiated from moderate to severe PAN, which involves more severe complications, such as arterial stenosis—particularly those involving the renal arteries and aorta—and ischemic complications that affect the heart, peripheral nervous system, and gastrointestinal system. To aid in risk stratification, the 1996 version of the Five Factor Score (FFS) can be used. This score assigns +1 point for each of the following: proteinuria greater > 1 g/day, serum creatinine > 140 µmol/L, cardiomyopathy, severe gastrointestinal involvement, and CNS involvement [53][54].

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