

Contents

Part I Introduction

1 Definitions	3
1.1 General Introduction	3
1.2 Definitions	4
2 Classification and Epidemiology	7
2.1 Introduction	7
2.2 ACR (1990) Criteria	8
2.3 Chapel Hill Consensus Definitions	9
2.4 Epidemiology	9
2.5 Etiological Factors	10
References	11
3 General Presentation of the Vasculitides	13
3.1 Introduction	13
3.2 Pattern Recognition	13
3.3 Laboratory Investigations	16
3.4 Differential Diagnosis	18
References	19
4 General Principles of Treatment	21
4.1 Treatment	21
4.2 Remission Induction	21
4.2.1 Large Vessel Disease	22
4.2.2 Medium Vessel Disease	22
4.2.3 Medium/Small Vessel Vasculitis	22
4.2.4 Small Vessel Vasculitis	24

viii Contents

4.3	Maintenance Therapy	25
4.4	Long-Term Follow-Up	25
4.5	Relapsing Disease	25
4.6	Refractory Disease	26
4.7	Specific Drugs Used to Treat Vasculitis	27
4.7.1	Cyclophosphamide	27
4.7.2	Glucocorticoids	28
4.7.3	Methotrexate	28
4.7.4	Plasma Exchange	29
4.8	Monitoring	29
4.9	Patient Advice	30
4.10	Detection and Prevention of the Adverse Effects of Therapy	30
4.10.1	Osteoporosis	30
4.10.2	Vaccinations	31
4.10.3	Pneumocystis Jirovecii Infection	31
4.10.4	Cyclophosphamide-Induced Bladder Toxicity	31
	References	32

Part II Features of Individual Diseases

5	Giant Cell Arteritis	35
5.1	Introduction	35
5.2	Definition and Classification	35
5.3	Epidemiology	36
5.4	Etiology	36
5.5	Clinical Features	37
5.5.1	Systemic	37
5.5.2	Craniofacial	37
5.5.3	Ophthalmic	39
5.5.4	Neurologic	39
5.5.5	Extracranial Artery	39
5.6	Laboratory Features	40
5.6.1	Immunology	40
5.6.2	Imaging	40
5.6.3	Pathology	41

5.7	Diagnosis	42
5.8	Assessment of Disease Activity	43
5.9	Treatment	43
5.10	Prognosis	44
	References	45
6	Takayasu Arteritis	47
6.1	Introduction	47
6.2	Definition and Classification	47
6.3	Epidemiology	48
6.4	Etiology	48
6.5	Clinical Features	49
	6.5.1 Systemic	49
	6.5.2 Vascular	49
6.6	Laboratory Features	49
	6.6.1 Immunology	50
	6.6.2 Imaging	50
	6.6.3 Pathology	52
6.7	Diagnosis	53
6.8	Assessment of Disease Activity	53
6.9	Treatment	53
	6.9.1 Pharmacological Therapy	53
	6.9.2 Surgery	54
6.10	Prognosis	54
	References	55
7	Wegener's Granulomatosis	57
7.1	Introduction	57
7.2	Definition and Classification	57
7.3	Epidemiology	58
7.4	Etiology	58
7.5	Clinical Features	59
	7.5.1 Systemic	59
	7.5.2 Pulmonary	59
	7.5.3 Cutaneous	59
	7.5.4 ENT	60
	7.5.5 Gastrointestinal	61

x Contents

7.5.6	Neurological	61
7.5.7	Renal	61
7.5.8	Eye	61
7.5.9	Musculoskeletal.	62
7.5.10	Cardiac	62
7.6	Laboratory Features	62
7.6.1	Immunology.	63
7.6.2	Imaging.	63
7.6.3	Pathology.	65
7.7	Diagnosis.	66
7.7.1	Differential Diagnosis	66
7.7.2	Assessment of Organ Involvement.	66
7.8	Assessment of Disease Activity	67
7.9	Treatment	67
7.9.1	Remission Induction.	68
7.9.2	Maintenance	68
7.10	Prognosis.	68
	References	70
8	Churg–Strauss Syndrome	71
8.1	Introduction.	71
8.2	Definition and Classification.	71
8.3	Epidemiology	72
8.4	Etiology.	73
8.5	Clinical Manifestations	73
8.5.1	Pulmonary	73
8.5.2	Cutaneous	74
8.5.3	Gastrointestinal.	75
8.5.4	Neurological	75
8.5.5	Cardiac	76
8.5.6	Renal	76
8.5.7	ENT	76
8.5.8	Ophthalmic	76
8.5.9	Musculoskeletal.	77
8.6	Laboratory Features	77
8.6.1	Immunology.	78
8.6.2	Imaging.	78
8.6.3	Pathology.	78

8.7	Diagnosis	78
8.7.1	Differential Diagnosis	78
8.7.2	Assessment of Organ Involvement. . .	79
8.8	Assessment of Disease Activity	79
8.9	Treatment	80
8.9.1	Remission Induction.	80
8.9.2	Maintenance	80
8.10	Prognosis	81
	References	81
9	Microscopic Polyangiitis	83
9.1	Introduction	83
9.2	Definition and Classification	83
9.3	Epidemiology	83
9.4	Etiology	84
9.5	Clinical Features	84
9.5.1	Systemic	84
9.5.2	Renal	84
9.5.3	Pulmonary	84
9.5.4	Cutaneous	86
9.5.5	Neurological	86
9.5.6	Cardiac	86
9.5.7	Gastrointestinal.	86
9.5.8	Otorhinolaryngeal.	86
9.5.9	Ocular.	87
9.5.10	Venous Thromboembolism	87
9.6	Laboratory Features	87
9.6.1	Immunology.	87
9.6.2	Pathology	87
9.7	Diagnosis	88
9.7.1	Differential Diagnosis	88
9.7.2	Assessment of Organ Involvement. . .	89
9.8	Assessment of Disease Activity	89
9.9	Treatment	90
9.9.1	Remission Induction.	90
9.9.2	Maintenance	91
9.10	Prognosis	91
	References	92

10 Polyarteritis Nodosa	95
10.1 Introduction	95
10.2 Definition and Classification	95
10.3 Epidemiology	95
10.4 Etiology	97
10.5 Clinical Features	97
10.5.1 Systemic	97
10.5.2 Cutaneous	97
10.5.3 Musculoskeletal	97
10.5.4 Neurological	98
10.5.5 Renal	99
10.5.6 Gastrointestinal	99
10.5.7 Cardiac	99
10.5.8 Orchitis	100
10.5.9 Other	100
10.6 Laboratory	100
10.6.1 Immunology	101
10.6.2 Viral Serology	101
10.6.3 Imaging	101
10.6.4 Pathology	102
10.7 Diagnosis	103
10.7.1 Differential Diagnosis	103
10.7.2 Assessment of Organ Involvement	103
10.8 Assessment of Disease Activity	104
10.9 Treatment	104
10.9.1 Non-HBV-PAN	104
10.9.2 Virus-Associated PAN	104
10.10 Prognosis	105
References	106
11 Kawasaki Disease	107
11.1 Introduction	107
11.2 Definition and Classification	107
11.3 Epidemiology	108
11.4 Etiology	108
11.5 Clinical Features	108
11.5.1 Mucosal	109
11.5.2 Cutaneous	109

11.5.3	Ophthalmic	110
11.5.4	Cardiovascular.....	110
11.6	Laboratory Features	111
11.6.1	Cardiac Investigations	111
11.6.2	Immunology.....	111
11.6.3	Pathology.....	111
11.7	Diagnosis.....	112
11.8	Assessment of Disease Activity	112
11.9	Treatment	112
11.10	Prognosis.....	113
	References	114
12	Henoch–Schönlein Purpura	115
12.1	Introduction.....	115
12.2	Definition and Classification.....	115
12.3	Epidemiology	116
12.4	Etiology.....	117
12.5	Clinical Features	117
12.5.1	Cutaneous	117
12.5.2	Gastrointestinal.....	117
12.5.3	Musculoskeletal.....	119
12.5.4	Renal	119
12.6	Laboratory Features	119
12.6.1	Renal Function	120
12.6.2	Immunology.....	120
12.6.3	Pathology.....	120
12.7	Diagnosis.....	121
12.8	Assessment of Disease Activity	121
12.9	Treatment	121
12.10	Prognosis.....	122
	References	123
13	Behçet’s Disease	125
13.1	Introduction.....	125
13.2	Definition and Classification.....	125
13.3	Epidemiology	125
13.4	Etiology.....	126
13.5	Clinical Features	126

13.5.1	Orogenital	126
13.5.2	Cutaneous	127
13.5.3	Ophthalmic	128
13.5.4	Neurological	129
13.5.5	Vascular	130
13.5.6	Musculoskeletal.	130
13.5.7	Gastrointestinal.	130
13.5.8	Renal	130
13.6	Laboratory Features	131
13.6.1	Immunology.	131
13.6.2	Genetic.	131
13.6.3	Synovial Fluid Examination	131
13.6.4	Cerebrospinal Fluid	131
13.6.5	Imaging.	131
13.6.6	Pathology.	132
13.7	Diagnosis.	132
13.8	Assessment of Disease Activity	132
13.9	Treatment	133
13.9.1	Mucocutaneous.	133
13.9.2	Ocular.	133
13.9.3	Vasculopathy	134
13.9.4	Gastrointestinal.	134
13.9.5	Musculoskeletal.	134
13.9.6	Neurological	134
13.10	Prognosis.	135
	References	136
14	Cryoglobulinemic Vasculitis	137
14.1	Introduction.	137
14.2	Definition.	137
14.3	Epidemiology	137
14.4	Etiology.	138
14.5	Clinical Features	138
14.5.1	Systemic	138
14.5.2	Cutaneous	138
14.5.3	Neurological	138
14.5.4	Renal	139
14.5.5	Musculoskeletal.	139

14.6	Laboratory Features	139
14.6.1	Viral Serology	140
14.6.2	Immunology.	140
14.6.3	Pathology.	140
14.7	Diagnosis.	140
14.8	Assessment of Disease Activity	141
14.9	Treatment	141
14.10	Prognosis.	141
	References	142
15	Vasculitis Mimics	143
15.1	Introduction.	143
15.2	Cholesterol Crystal Embolism	143
15.2.1	Clinical Features	144
15.2.2	Laboratory Features.	146
15.2.3	Treatment.	147
15.3	Calciophylaxis.	147
15.4	Cardiac Myxoma.	147
15.4.1	Clinical Features	147
15.4.2	Treatment.	148
15.5	Infective Endocarditis	148
15.6	Fibromuscular Dysplasia.	148
15.7	Chronic Ergotism	149
15.8	Köhlmeier–Degos Disease	149
15.9	Cryofibrinogenemia.	150
15.10	Radiation Vasculopathy.	151
15.11	Cocaine Abuse	151
15.12	Scurvy	151
15.13	Sweet’s Syndrome.	153
	References	154
16	Secondary Vasculitis	155
16.1	Introduction.	155
16.2	Definition.	155
16.3	Epidemiology	155
16.4	Etiology.	156
16.5	Infection and Vasculitis	156
16.6	Vasculitis and Malignancy	158
16.7	Drug-Induced Vasculitis	160

16.8	Systemic Rheumatoid Vasculitis	161
16.9	Systemic Lupus Erythematosus	163
16.10	Sjögren’s Syndrome	164
16.11	Spondyloarthropathies	164
	References	165
17	Primary Angiitis of the Central Nervous	
	System Vasculitis	167
17.1	Introduction	167
17.2	Definition and Classification	167
17.3	Epidemiology	168
17.4	Etiology	168
17.5	Clinical Features	168
	17.5.1 Systemic	168
	17.5.2 Neurological	168
17.6	Laboratory Features	169
	17.6.1 Cerebrospinal Fluid	169
	17.6.2 Imaging	169
	17.6.3 Pathology	170
	17.6.4 Electroencephalography	170
17.7	Diagnosis	170
17.8	Assessment of Disease Activity	170
17.9	Treatment	170
17.10	Prognosis	171
	References	172
18	Relapsing Polychondritis	173
18.1	Introduction	173
18.2	Definition and Classification	173
18.3	Epidemiology	173
18.4	Etiology	173
18.5	Clinical Features	174
	18.5.1 Systemic	174
	18.5.2 Cartilage Inflammation	174
	18.5.3 Pulmonary	175
	18.5.4 Ophthalmic	176
	18.5.5 Musculoskeletal	177
	18.5.6 Cutaneous	177
	18.5.7 Cardiovascular	177

18.6	Laboratory Features	177
18.6.1	Immunology.	177
18.6.2	Imaging.	177
18.6.3	Pathology.	178
18.7	Diagnosis.	178
18.8	Assessment Disease Activity	178
18.9	Treatment	178
18.10	Prognosis.	179
	References	179
19	Cogan’s Syndrome	181
19.1	Introduction.	181
19.2	Definition and Classification.	181
19.3	Epidemiology	181
19.4	Etiology.	181
19.5	Clinical Features	182
19.5.1	Systemic	182
19.5.2	Ocular.	182
19.5.3	Vestibular and Auditory.	182
19.5.4	Vascular	183
19.5.5	Neurological	183
19.6	Laboratory Features	183
19.6.1	Immunology.	183
19.6.2	Pathology.	183
19.7	Diagnosis.	184
19.8	Assessment of Disease Activity	184
19.9	Treatment	184
19.10	Prognosis.	184
	References	185
	Index	187