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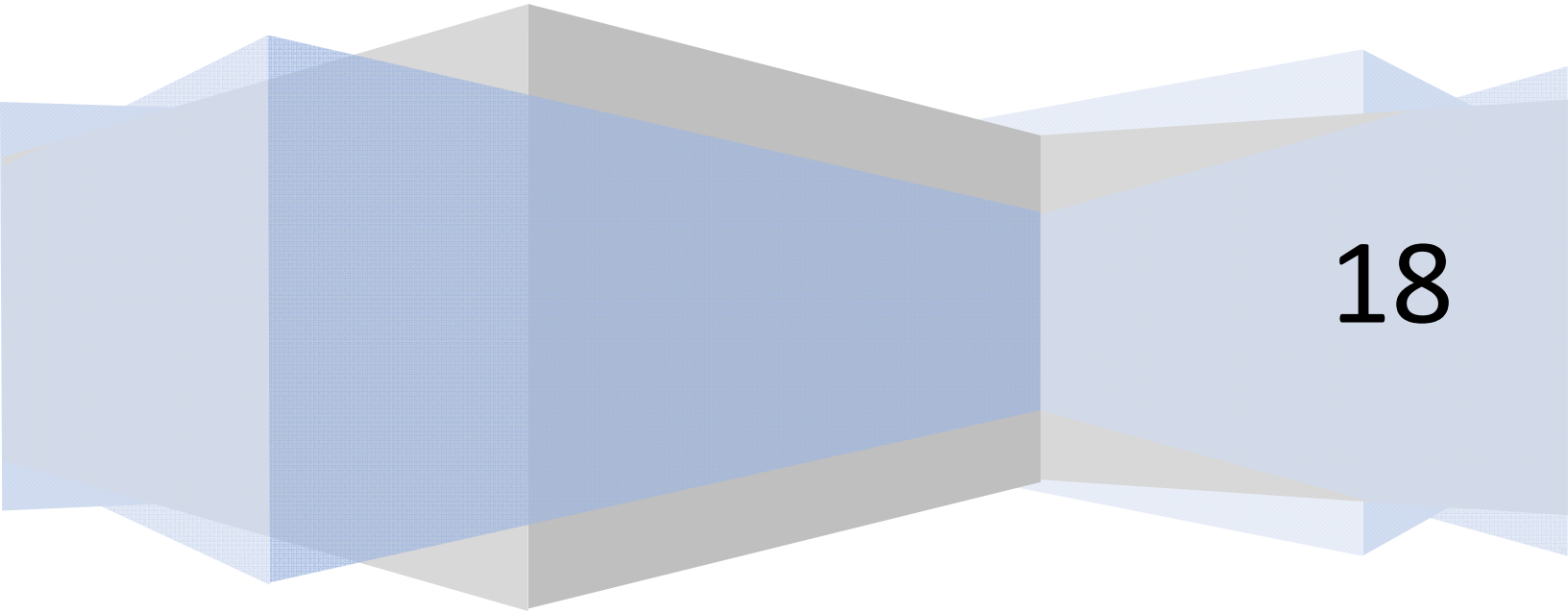
Sample Learning Material

USMLE Step 1

A Sample from Pathology.



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Microscopic polyangiitis (MPA)

MPA or microscopic polyarteritis is a clinical **variant of polyarteritis nodosa (PAN)** typified by prominent **lung involvement** and segmental necrotizing **glomerulonephritis**, sometimes **rapidly progressive**. These are absent, by definition, in classic PAN. Constitutional symptoms such as fever, asthenia, and myalgias are common in both PAN and MPA. Elevated acute-phase reactants, thrombocytosis, leukocytosis, and the anemia of inflammatory disease are common, although they are not uniformly present.

Both polyarteritis nodosa and microscopic polyarteritis (which affects arterioles, capillaries and venules, rather than medium- and small-sized vessels) can cause **neurologic deficits, livedo reticularis, renal compromise, and systemic symptoms of fatigue, fever, and weight loss**. However, because **polyarteritis nodosa affects larger vessels**, it can cause downstream **glomerular ischemia**, thereby activating the renin-angiotensin system and **raising blood pressure without causing an active urine sediment**. Microscopic polyarteritis, on the other hand, affects smaller vessels, causing glomerular necrosis and the resulting **active urine sediment of red cell casts and protein**, without raising blood pressure. Also, while both PAN and MPA cause mesenteric angina, **abdominal angiography is usually negative** in MPA because smaller vessels are involved (abdominal angiography is revealing in PAN). The Table below differentiates the two syndromes.

Table 5. Differentiating Polyarteritis Nodosa from Microscopic Polyangiitis

	Polyarteritis nodosa	**Microscopic polyangiitis**
Size of vessels involved	Medium-sized and small muscular arteries	Arterioles, capillaries, and venules
Pulmonary involvement	-	+
Neurological deficits	+	+
Renal compromise	+	+
Mesenteric angina	+	+
Constitutional symptoms	+	+
Hypertension	+	- *
Urine sediment	None	Active (red cell casts, protein, leukocytes)
Pauci-immune glomerulonephritis	-	+
ANCA	-	+
Abdominal angiography	+	-

*Hypertension is typically absent; however, it may be present in 34%.

MPA and granulomatosis with polyangiitis (formerly Wegener's granulomatosis) seem to be part of a clinical spectrum. However, an **absence of granuloma formation** and **sparing of the upper respiratory tract are features of MPA**. These features help to distinguish MPA from granulomatosis with polyangiitis, although the two conditions are occasionally difficult to distinguish.The renal biopsy tissue in **MPA**, as in **Wegener** and **Churg-Strauss syndrome**

(CSS), does not contain extensive immune complexes on immunofluorescent staining and electron microscopy (so-called **pauci-immune glomerulonephritis**). The presence of **serum p-ANCA** with antimyeloperoxidase specificity (found in 60% of MPA patients) supports the clinical diagnosis of MPA, but **p-ANCA is not specific** for this disease. ANCAs are not characteristic of PAN. **Treatment** is **corticosteroids**, cytotoxic agents added if no response.