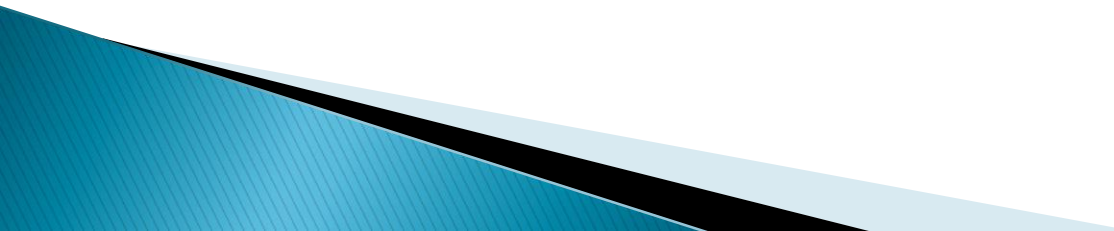
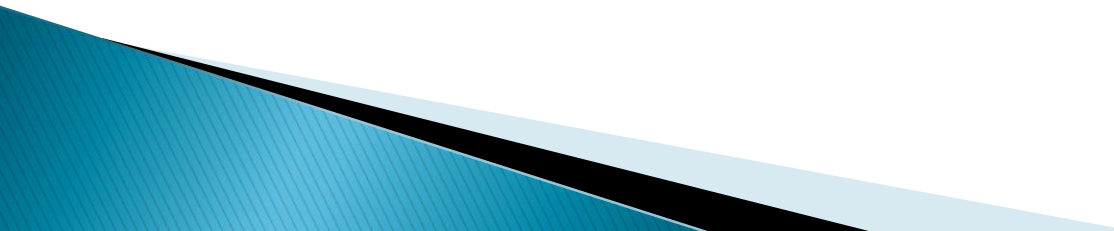


Fibromuscular Dysplasia (FMD)

FMD Definition

- ▶ Nonatherosclerotic, noninflammatory arteriopathy , narrowing of small and medium sized vessels
 - ▶ Segmental fibrosis and smooth muscle overgrowth of arterial wall
 - ▶ Mechanical ,genetic ,and hormonal factors involved in but cause of FMD remain unknown
- 

Epidemiology of FMD

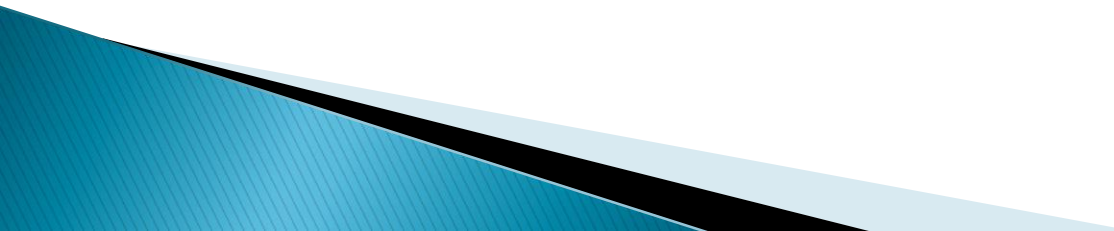
- ▶ Occur in any age but mainly affects children and individuals over the age of 50 years
 - ▶ Prevalence is 2–3%
 - ▶ 90% women
 - ▶ 10% familial
- 

Classification of FMD

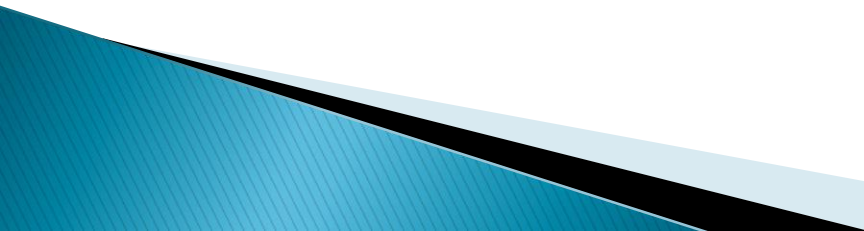
- ▶ FMD is classified in 5 main types, based on histology and location
- ▶ .1) medial fibroplasia (~90%)
- ▶ 2) intimal fibroplasia (~10%)
- ▶ 3) perimedial fibroplasia (<1%)
- ▶ 4) medial hyperplasia (<1%)
- ▶ 5) adventitial (periarterial) hyperplasia (<1%) .

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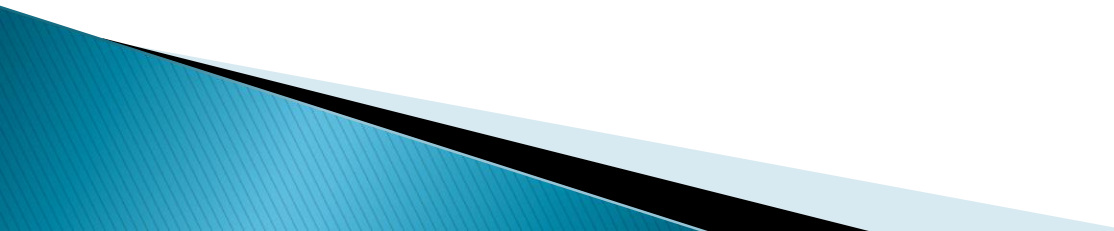
FMD Clinical Manifestation

- ▶ Asymptomatic
 - ▶ Renovascular hypertension 70%(35% bilateral)
 - ▶ CNS manifestation
 - ▶ Peripheral artery manifestation
 - ▶ Aneurysms and arterial dissections , more in men
- 

Segmental Arterial Mediolyysis

- ▶ Segmental involvement, characterized by dissecting aneurysms
 - ▶ Involvement of Abdominal aortic branches, especially the mesenteric arteries and carotid and vertebral arteries and iliac arteries .
 - ▶ The most common presentation is abdominal pain and hemorrhage in the middle and older age men
 - ▶ The acute phase of SAM carries a 50% mortality rate
- 

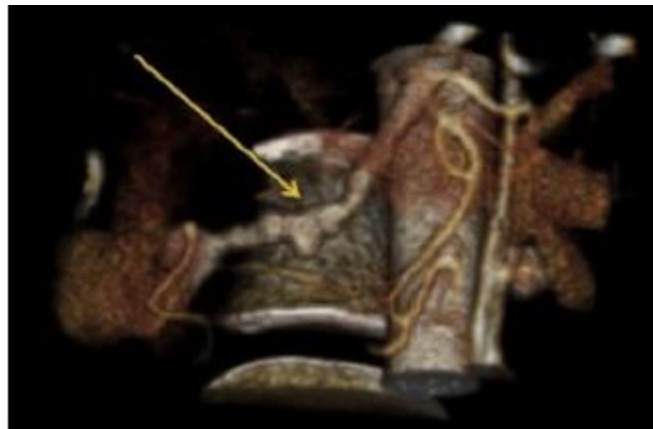
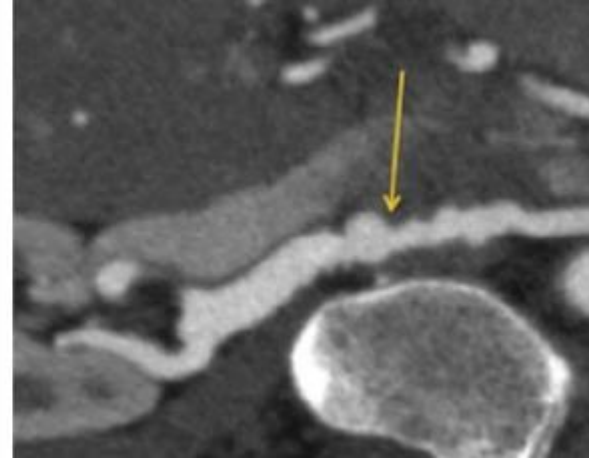
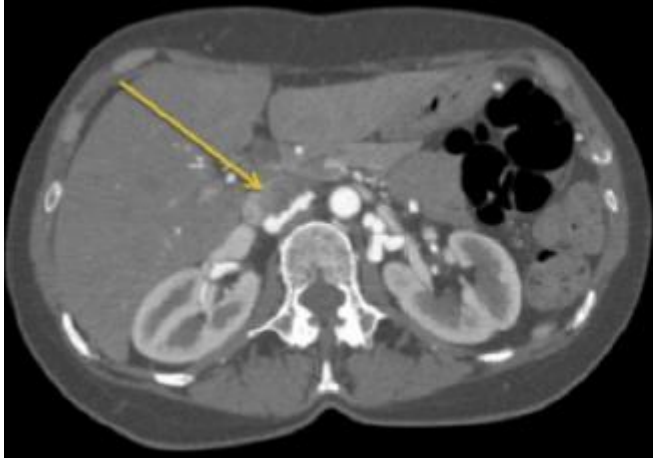
FMD Diagnosis

- ▶ Duplex ultrasonography
 - ▶ CTA
 - ▶ MRA
 - ▶ Conventional angiography, gold standard
 - ▶ The classic “string of beads”
 - ▶ Focal concentric” narrowing and “diffuse stenosis” in intimal and periadventitial fibroplasia FMD types
- 

Conventional angiography in a patient with FMD



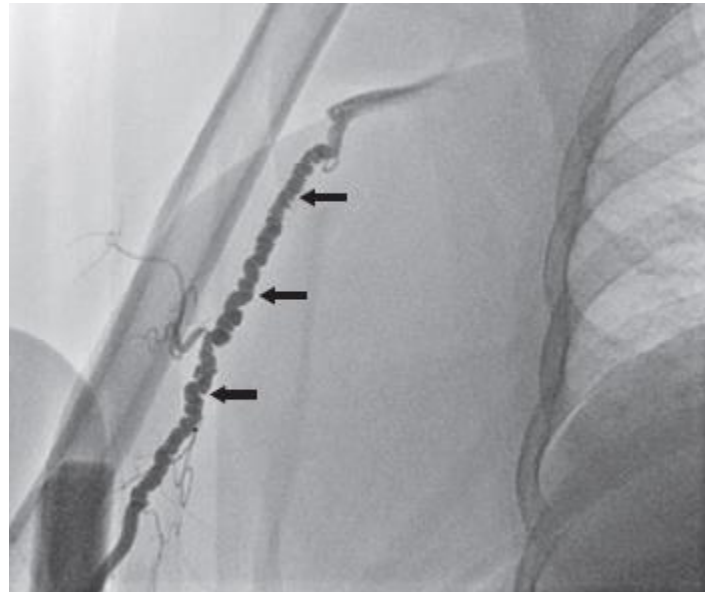
CT angiogram shows a beaded (string of beads) ,3-dimensional reconstruction of the renal artery.



MR Angiogram of 51-year-old woman with renovascular hypertension due to FMD

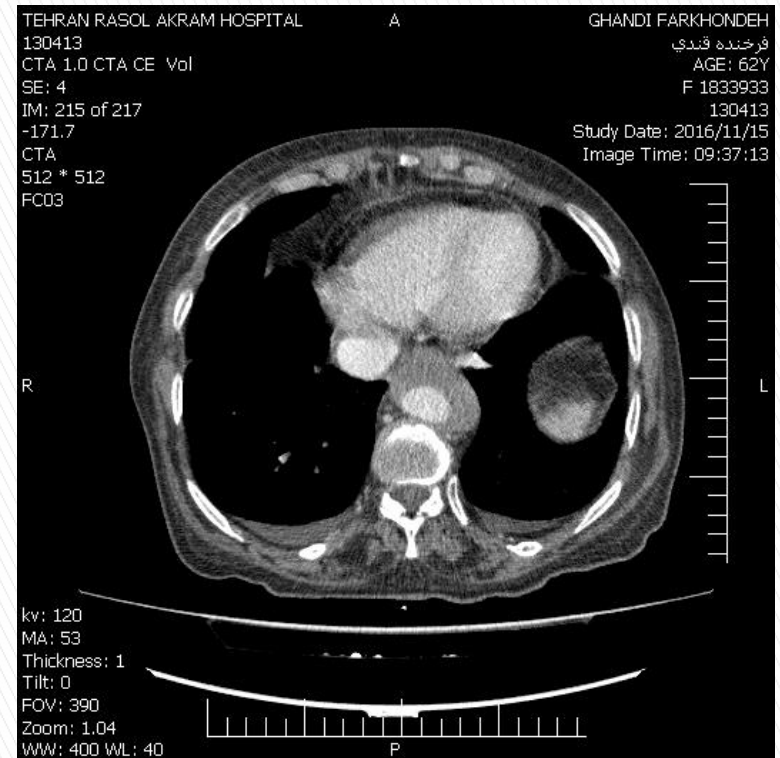


Fibromuscular Dysplasia of the Brachial Artery

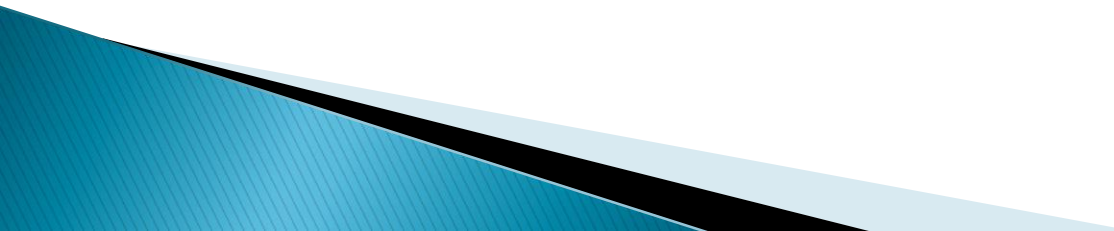


DDX of FMD

- ▶ Normal inflammatory markers (unless severe ischemia leads to tissue infarction)
- ▶ Absence of constitutional symptoms
- ▶ Absence of arterial wall thickening, edema or contrast uptake on cross sectional vascular imaging
- ▶ DDX: TAK, GCA and PAN



FMD Treatment

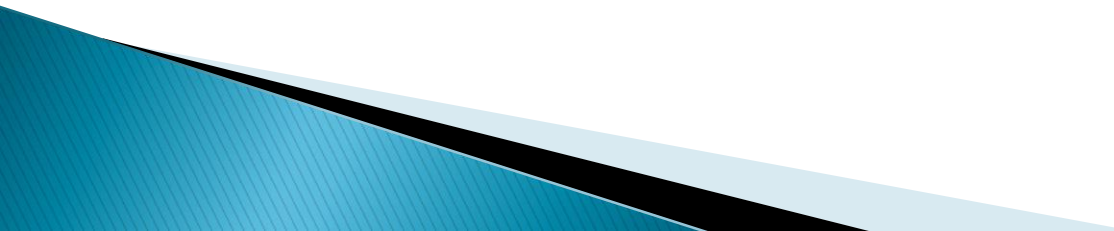
- ▶ Medical therapy: ARBs or ACEIs for RVH
 - ▶ Endovascular treatment : PTA for RVH ,coiling (e.g., aneurysms)
 - ▶ Surgical interventions for re-stenosis after PTA, complex lesions, or lesions difficult for PTA
 - ▶ Heparin therapy and 3–6 months warfarin for arterial dissection
 - ▶ Correction of risk factors
- 

Calciophylaxis

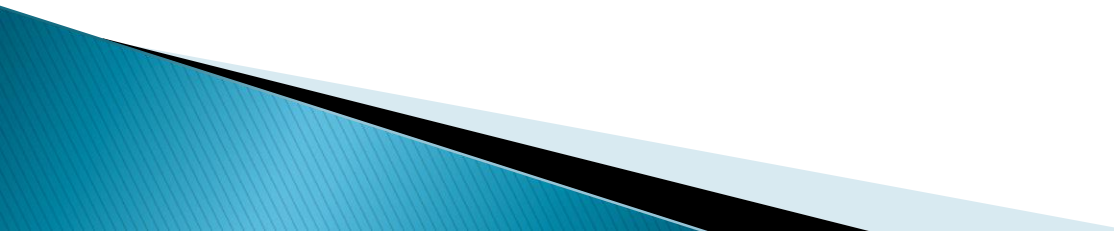
Calciophylaxis: Etiology

- ▶ 1% to 4% of hemodialysis patients
 - ▶ Primary hyperparathyroidism
 - ▶ Alcoholic liver disease, cirrhosis, IBD
 - ▶ Malignancy
 - ▶ Antiphospholipid antibody syndrome
 - ▶ Obese diabetic patients
 - ▶ Rheumatoid arthritis on long-term steroid and methotrexate use
 - ▶ Protein C and S deficiencies
 - ▶ Heparin, warfarin
- 

Claciphylaxis Pathogenesis

- ▶ Deregulation of vascular calcium deposition
 - ▶ The impairment of endogenous calcification inhibitors :osteoprotegerin and MPG
 - ▶ Gama glutamyl–carboxylase Vitk dependent enzyme that chagne inactive to active form of MPG
 - ▶ Low serum albumin, Systemic hypotension
 - ▶ Local trauma (e.g., from subcutaneous injections)
- 

Claciphylaxis Clinical Manifestation

- ▶ Skin and subcutaneous tissues
 - ▶ Areas with increased vascularity and significant subcutaneous adipose tissue: thighs, buttocks abdomen, and breasts and some times and distal extremities
 - ▶ Livedo reticularis
 - ▶ Violaceous, superficial, tender nodules ulceration and hemorrhage with dry necrosis
- 

Early and Late stage of Claciphylaxis



A, Early abdominal lesion

B, Advanced lower extremity lesions

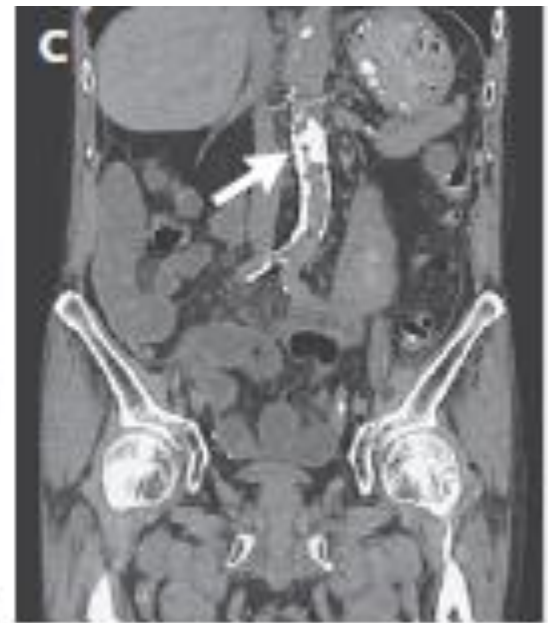


A

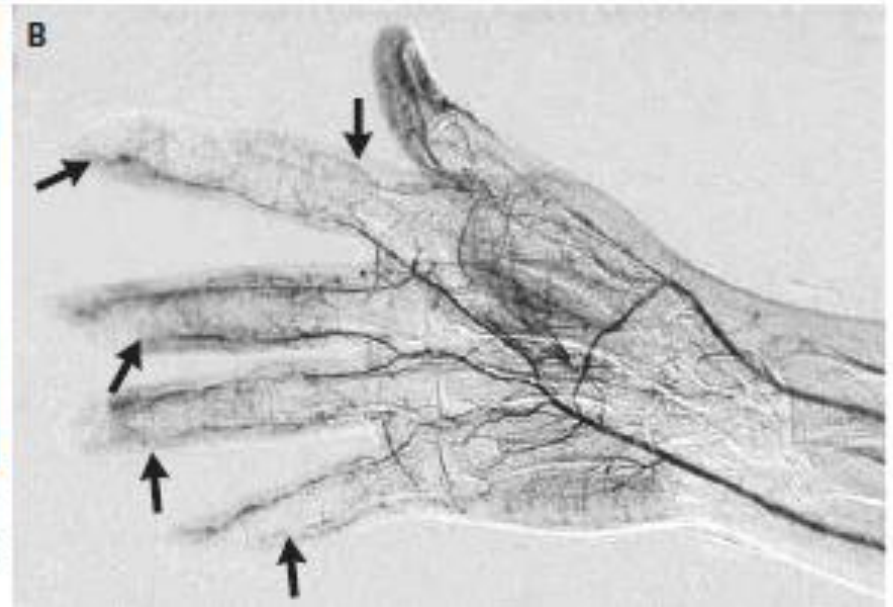


B

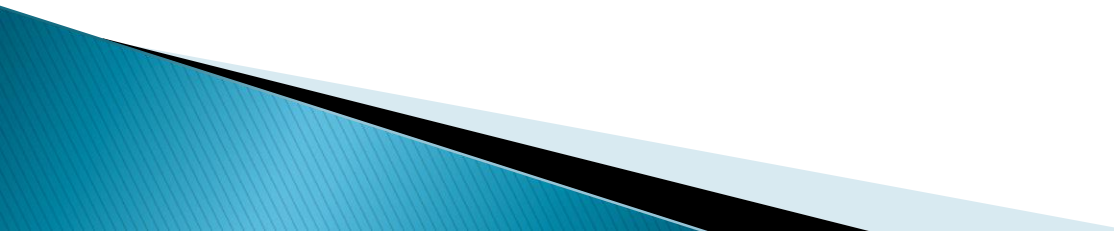
Aortic Calcification and Superior– Mesenteric– Artery Stenosis in patients with 6 years peritoneal dialysis



Calcific Arteriopathy

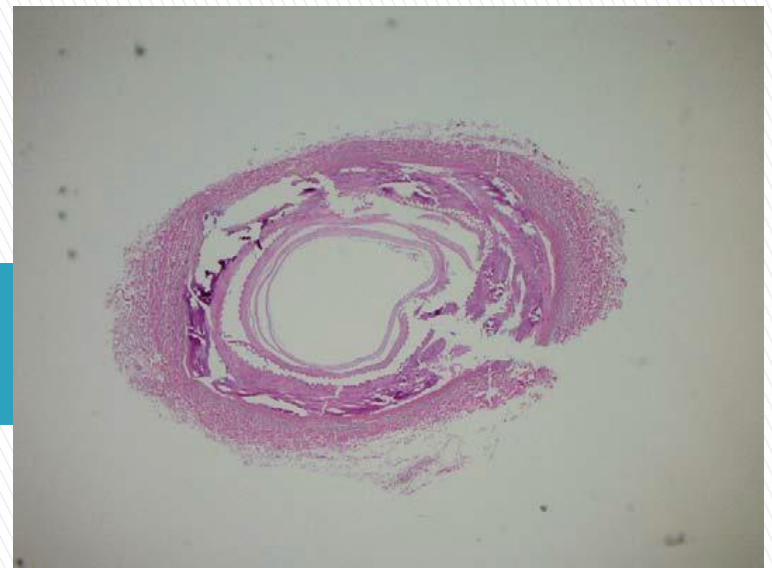
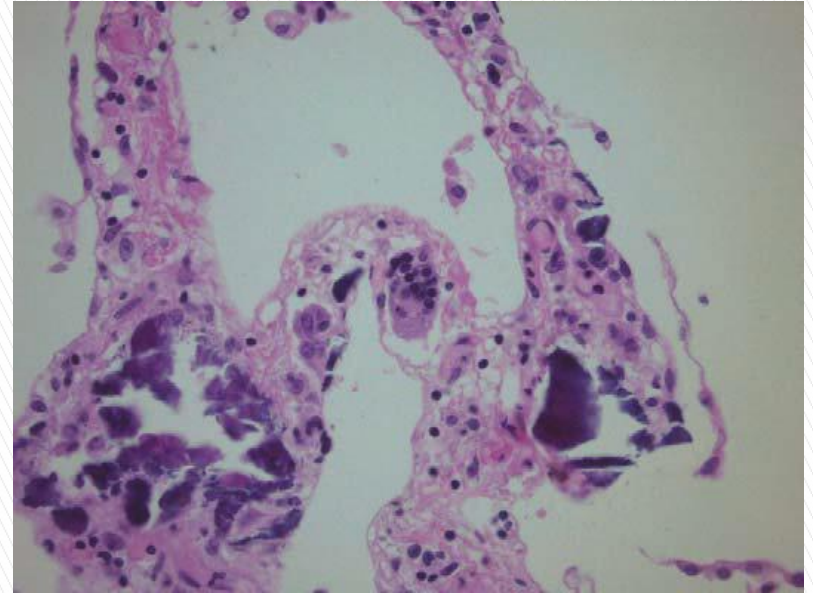


Claciphylaxis :Lab Data

- ▶ High serum phosphate
 - ▶ High calcium phosphorous product
 - ▶ High PTH
 - ▶ Transcutaneous oxygen saturation probes identify skin ischemia prior to the development of skin necrosis
 - ▶ Abnormal uptake on bone scan imaging
- 

Pathology of Claciphylaxis

- ▶ Intravascular calcium deposit within arteriols and venules
- ▶ Panniculitis
- ▶ Intimal proliferation or fibrosis
- ▶ Microthrombi
- ▶ Paucity of inflammation
- ▶ Giant cell reaction



Potential therapies for medial vascular calcification

