Polyarteritis nodosa

Polyarteritis nodosa, also known as panarteritis nodosa, periarteritis nodosa, Kussmaul disease, Kussmaul-Maier disease or PAN, is a systemic vasculitis of small- or medium-sized muscular arteries, typically involving renal and visceral vessels but sparing the pulmonary circulation.

Infantile polyarteritis nodosa is restricted to infants.

In polyarteritis nodosa, small aneurysms are strung like the beads of a rosary, therefore making "rosary sign" an important diagnostic feature of the vasculitis.

With treatment, five-year survival is 80%; without treatment, five-year survival is 13%. Death is often a consequence of kidney failure, myocardial infarction, or stroke.

Intracranial aneurysms in polyarteritis nodosa (PAN) are exceedingly rare lesions with unpredictable behavior that pose real challenges to microsurgical and endovascular interventions owing to their inflammatory nature.

Case reports

A 20-year-old man presented with diplopia, headaches, chronic abdominal pain, and weight loss. Diagnostic evaluations confirmed PAN, including bilateral giant cavernous carotid aneurysms. Cyclophosphamide and steroids achieved significant and sustained clinical improvement, with a decision to follow the aneurysms serially. Seven years later the left unruptured aneurysm enlarged, causing a sudden severe headache and a cavernous sinus syndrome. Treatment of the symptomatic aneurysm was pursued using flow diversion (PED) and the internal carotid artery was successfully reconstructed with a total of four overlapping PEDs. At 6 months follow-up, complete exclusion of the aneurysm was demonstrated, with symptomatic recovery. This is the first description of using a flow-diverting technique in an inflammatory vasculitis. In this case, PEDs not only attained a definitive closure of the aneurysm but also reconstructed the damaged and fragile arterial segment affected with vasculitis ¹⁾.

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Martinez Santos J, Kaderali Z, Spears J, Rubin LA, Marotta TR. Flow diversion in vasculitic intracranial aneurysms? Repair of giant complex cavernous carotid aneurysm in polyarteritis nodosa using Pipeline embolization devices: first reported case. J Neurointerv Surg. 2015 Jun 3. pii: neurintsurg-2015-011780.rep. doi: 10.1136/neurintsurg-2015-011780.rep. [Epub ahead of print] PubMed PMID: 26041095.

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