Hereditary hemorrhagic telangiectasia

- Osler-Weber-Rendu syndrome
- The classic triad on presentation is:
  - epistaxis
  - multiple telangiectasias
  - positive family history

HHT

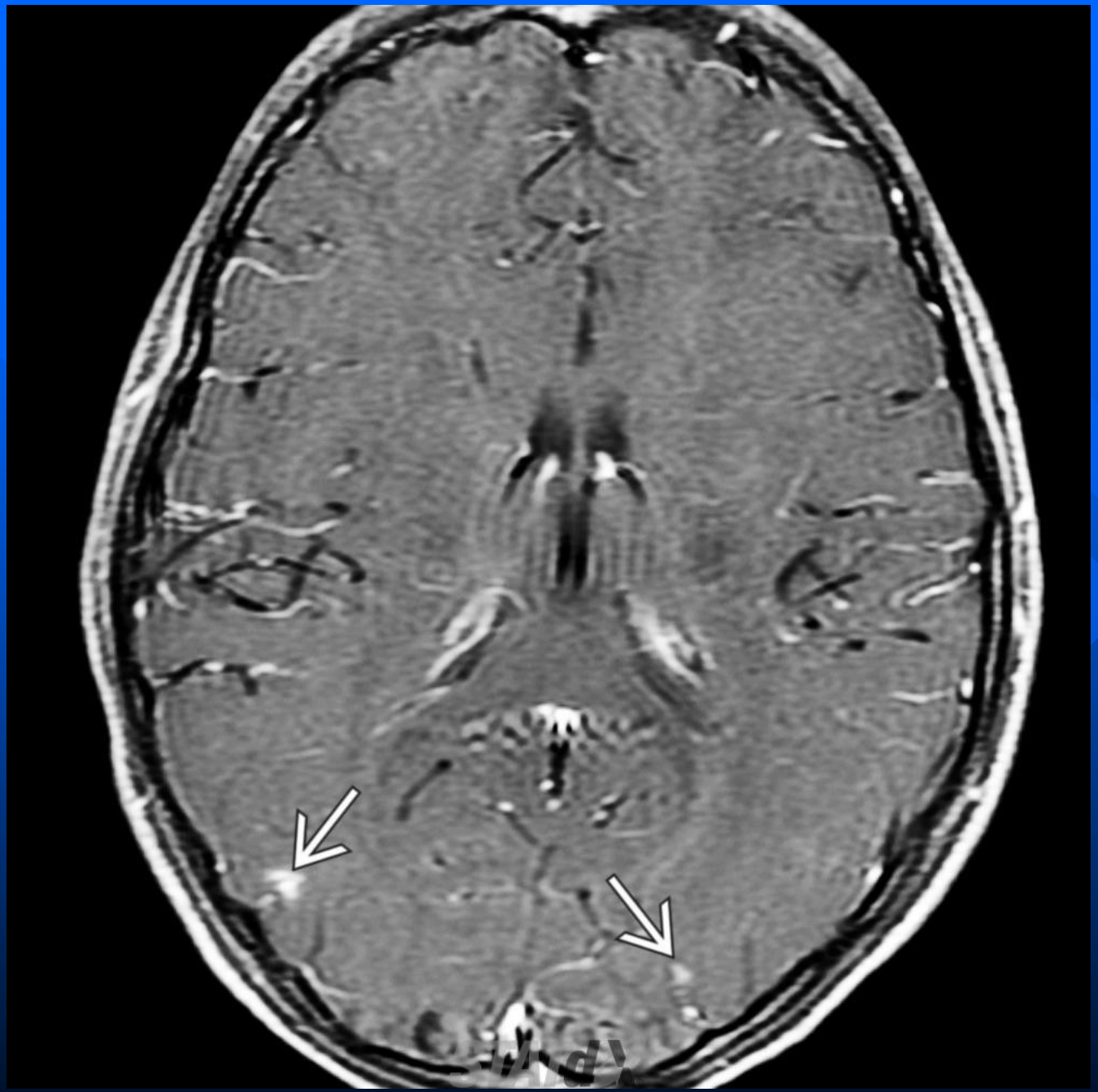
- Multiple arteriovenous malformations (AVMs) that lack an intervening capillary network.
- Telangiectasias (small superficial AVMs) are particularly common

CNS

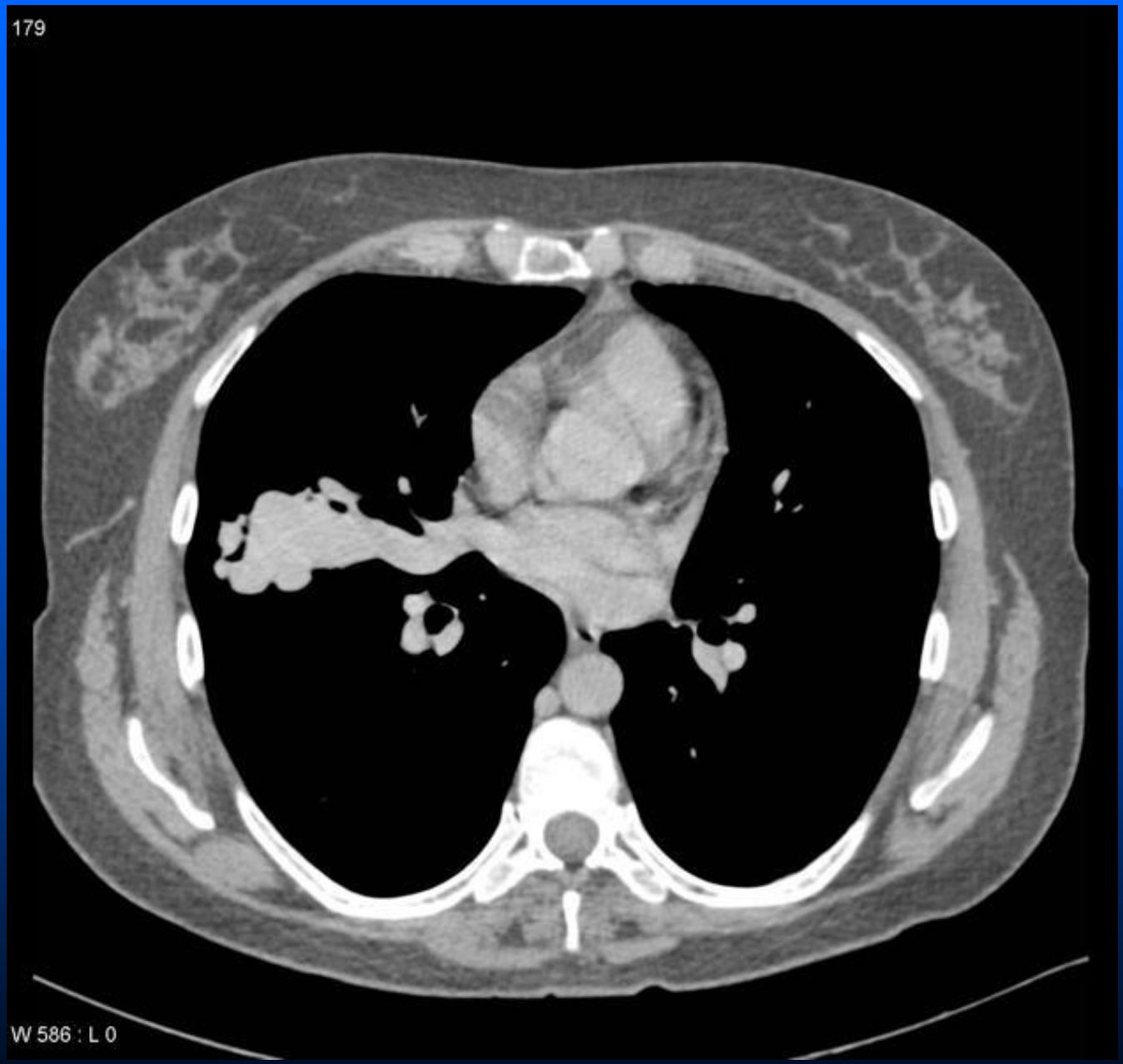
- **5-10%** cerebral AVMs, spinal AVMs or cerebral aneurysms
- One-third of cerebral complications in HHT are due to cerebral AVMs or aneurysms, and two-thirds are due to paradoxical emboli from pulmonary AVMs.
- Increased incidence of capillary telangiectasia and developmental venous anomalies

Best diagnostic clue

- Multiple pulmonary (pAVM) or cerebral arteriovenous malformations (cAVM) in patient with recurrent epistaxis
- 50% have nosebleeds by 10y, 80-90% by 21 years; 95% lifetime
- Recurrent epistaxis from nasal mucosal telangiectasias
  - Intracranial bleed from AVM/AVF
  - TIA, stroke, abscess secondary to pAVMs



A screening T1 C+ FS 3T MR scan was obtained in this 11-year-old female with a family history of HHT shows 2 tiny "fluffy" foci of enhancement (white solid arrow) consistent with capillary vascular malformations (sometimes called "micro-AVMS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