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A Large High-Flow Arteriovenous Malformation of the Distal Third of the Right Upper Arm (Cho Type IIIb): A Case Report and Literature Review

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ABSTRACT

Background. Peripheral arteriovenous malformations (AVMs) are rare congenital high-flow vascular lesions defined by a direct arteriovenous shunt lacking an intervening capillary bed. Misdiagnosis or inappropriate management may lead to catastrophic haemorrhage, high-output cardiac failure, trophic ulceration and functional loss of the limb. The angiographic Cho–Do–Yakes classification directly determines the endovascular strategy.

Case presentation. A patient was referred for imaging evaluation of a slowly enlarging, pulsatile swelling of the distal third of the right arm. Clinical examination revealed a palpable thrill, local warmth, a continuous systolo-diastolic bruit and dilated tortuous superficial veins, with mildly diminished ipsilateral distal pulses suggesting a moderate steal phenomenon (Schöbinger stage II). Triphasic CT angiography demonstrated a bulky dysplastic soft-tissue vascular network measuring 107 × 46 mm, organised around a complex nidus, fed predominantly by the distal brachial artery, with early venous opacification during the arterial phase through two dominant outflow axes — the cephalic vein and the ulnar venous network. Multiple intralesional phleboliths were present, without cortical erosion or medullary involvement of the distal humerus. The lesion was classified as Cho type IIIb.

Conclusion. This case illustrates the diagnostic power of multiphasic CT angiography in mapping a high-flow AVM and the necessity of rigorous classification to guide endovascular therapy. A painless, slowly progressive upper-limb mass may harbour a potentially devastating high-flow shunt whose control relies on the combined expertise of the diagnostic radiologist, the interventional radiologist and the vascular surgeon.

KEYWORDS

Arteriovenous malformation; Peripheral AVM; Cho classification; ISSVA classification; Nidus; CT angiography; Embolization; High-flow shunt; Schöbinger staging

MAIN ARTICLE

INTRODUCTION

Among vascular anomalies, arteriovenous malformations (AVMs) hold a distinct position: they are congenital, non-neoplastic, progressive, and typically silent for years before an abrupt clinical presentation. The reference nosology — the International Society for the Study of Vascular Anomalies (ISSVA) classification, last updated in 2025 — places them among high-flow anomalies, alongside arteriovenous fistulas (AVFs) and complex combined malformations [1,11].

Unlike infantile haemangiomas, which are true tumours that proliferate and then involute, AVMs are present at birth, grow proportionally with the patient, and may decompensate abruptly under identifiable triggers: puberty, pregnancy, hormonal contraception, trauma, inappropriate surgery, or misdirected percutaneous biopsy [2,4].

Their pathophysiological signature is the complete absence of a capillary network between arterial inflow and venous outflow. Blood shunts directly from the arterial to the venous side through a dysplastic tangle termed the **nidus**, the haemodynamic engine of the lesion [3,5].

This direct shunt generates a predictable cascade: chronic hyperflow with progressive dilatation of feeding arteries and draining veins and arterialisation of venous walls; a vascular steal phenomenon depriving distal territories of normal perfusion, causing ischaemic pain, trophic changes and occasionally digital necrosis; cardiac overload that may progress to high-output cardiac failure in major lesions; and mural fragility of dilated arterialised veins, exposing the patient to spontaneous or iatrogenic haemorrhage.

Extracranial AVMs represent fewer than 5% of all vascular anomalies, with a craniocervical predilection (~50%), followed by pelvic (~15%) and peripheral limb (~30%) locations [4].

The upper limb is a particularly hazardous site because of the density of critical structures (brachial neurovascular bundle, median, ulnar and radial nerves), the highly specialised function of the hand, which precludes mutilating surgery, and the major cosmetic and psychosocial impact.

Imaging plays a cardinal role: it confirms the diagnosis, distinguishes AVM from haemodynamically adjacent entities, stages and classifies the lesion, maps its architecture, and ultimately guides the endovascular procedure. Multiphasic CT angiography and MRI with dynamic sequences currently constitute the non-invasive foundation of pre-therapeutic assessment [9,10].

CASE PRESENTATION

A patient was referred to the department of diagnostic imaging for evaluation of a swelling of the distal third of the right arm, insidiously progressive over several years, with recent volumetric increase and the appearance of permanent, tortuous, engorged superficial veins over the antecubital fossa and proximal forearm. The patient reported local warmth, a sensation of heaviness on prolonged dependent positioning of the arm, and occasional distal paraesthesias.

Inspection revealed a poorly delimited, non-inflammatory mass without overt overlying skin change, associated with a discrete ectatic venous network. Palpation elicited a continuous thrill with systolic accentuation, and auscultation disclosed a systolo-diastolic bruit — the clinical hallmark of a high-flow arteriovenous shunt. Ipsilateral radial and ulnar pulses were present but mildly diminished compared with the contralateral side, consistent with a moderate steal phenomenon. Distal neurological examination was unremarkable. The lesion therefore corresponded to **Schöbinger stage II (expansion)**.

CT angiography of the right upper limb was performed on a multidetector scanner using an optimised triphasic protocol: a non-enhanced acquisition to detect calcifications (phleboliths) and osseous remodelling; an early arterial phase triggered by bolus tracking on the ipsilateral subclavian artery (150 HU threshold) ; and a delayed venous phase (60–70 s) to analyse drainage and shunt dynamics. Post-processing included maximum intensity projection (MIP) arterial phase (figure 1) and venous phase (figure 2), 3D volume rendering (VR) (figure 3) and curvilinear multiplanar reformations in axial, coronal and sagittal planes, to obtain a comprehensive three-dimensional map of the pathological vascular network.

Table 1. CT angiographic findings

Feature	Findings
Location	Soft tissues of the distal third of the right arm, predominantly anteromedial, without invasion of deep muscular compartments
Nidus	Bulky, tortuous dysplastic vascular network, 107 × 46 mm in greatest axis
Architecture	Tangle of vascular channels of irregular calibre, with ampullary dilatations and areas of relative stenosis
Arterial inflow	Predominant supply from the distal brachial artery, giving rise to several dilated tortuous feeders converging on the nidus

Venous outflow	Early venous opacification during the arterial phase, through two dominant axes — cephalic vein and ulnar venous network — both dilated secondary to chronic hyperflow
Calcifications	Multiple rounded intralesional phleboliths of frank calcific density, scattered within the nidus
Adjacent bone	No cortical lysis, periosteal erosion or medullary involvement of the distal humerus; preserved anatomical relationships
Adjacent structures	No visible neural involvement; discrete displacement of muscle bellies without destruction

The imaging appearance was consistent with a high-flow arteriovenous malformation of the distal third of the right arm, fed mainly by the distal brachial artery, with early efferent drainage through the cephalic and ulnar venous networks and multiple intralesional phleboliths. Based on the presence of a bulky complex nidus, multiple dilated shunts and multi-axial venous drainage, the lesion was classified as **Cho type IIIb**.

DISCUSSION

3.1 The high-flow versus low-flow dichotomy

The distinction between high-flow and low-flow lesions is the central pivot of vascular anomaly nosology [1,3,11]. Adopted from the seminal work of Mulliken and Glowacki (1982) and subsequently formalised by ISSVA, it rests on a simple, robust haemodynamic criterion. High-flow anomalies (AVM, AVF, pure arterial malformations) exhibit an active shunt, turbulent flow, a palpable thrill, an audible bruit and early venous opacification on dynamic imaging. Low-flow anomalies (venous, lymphatic, capillary malformations and their combinations) are non-pulsatile, compressible, bruit-free, and show no early arterial-phase opacification.

This distinction is not academic. Conventional low-pressure sclerotherapy applied to an AVM is both ineffective and hazardous: the sclerosant is immediately dispersed into the systemic circulation through the shunt, with risks of pulmonary embolism, distal ischaemia and cutaneous necrosis [3,4]. In our patient, high-flow features were unequivocal — thrill, continuous bruit, early venous opacification and hypertrophied draining veins — establishing the haemodynamic diagnosis definitively.

3.2 The nidus: the decisive therapeutic target

The nidus is the cardinal semiological concept of any AVM. It designates the dysplastic tangle of channels within which arteriovenous shunting occurs. Histologically it comprises vessels with hybrid arterial–venous walls lacking an organised internal elastic lamina, embedded in a fibrous stroma often showing low-grade chronic inflammation [3].

The therapeutic corollary is imperative: **any embolization that fails to reach the nidus is doomed to fail**[3,5,8]. Simple occlusion of feeding arteries triggers collateral recruitment — the so-called "Medusa's head phenomenon" — in which neighbouring muscular, cutaneous and osteoperiosteal branches dilate within weeks to reconstitute an equivalent, often more complex and less accessible afferent network. This pitfall explains the poor historical reputation of surgical ligation of feeding arteries, now formally contraindicated in AVM management.

In our patient, the considerable nidus size (107×46 mm) mandates meticulous planning: selection of the access route (transarterial via selective brachial catheterisation, retrograde transvenous via the cephalic vein, or direct percutaneous puncture under ultrasound or fluoroscopic guidance); selection of the embolic agent (absolute ethanol, Onyx®, n-BCA, or combinations); and staged treatment, as bulky AVMs almost invariably require several sessions separated by weeks to months.

3.3 Phleboliths: a diagnostic pitfall

Phleboliths are intravascular calcific concretions resulting from organised, calcified old thrombus within areas of relative stasis [6,7]. Classically described as a near-pathognomonic hallmark of venous malformations, they may also occur in bulky, long-standing AVMs, particularly when the nidus contains dilated slow-flow venular compartments. Their presence in our patient does not challenge the diagnosis of AVM; rather, it reflects the chronicity of the lesion, the coexistence of mixed haemodynamic compartments within a complex nidus, and possibly a combined malformative component (the ISSVA concept of *combined vascular malformations*). The cardinal rule is that phleboliths alone must never prompt a hasty diagnosis of "pure venous malformation" without assessment of flow dynamics: it is early arterial-phase venous opacification — present here — that establishes arteriovenous shunting and mandates classification as an AVM.

3.4 Classification frameworks

ISSVA 2025 provides a global, biologically and genetically oriented classification distinguishing vascular tumours; simple vascular malformations (capillary, venous, lymphatic, arteriovenous, fistulas); combined malformations; anomalies of named large vessels; and vascular malformations associated with other anomalies (Klippel–Trénaunay, Parkes–Weber, CLOVES syndromes) [1,11]. Our patient belongs to the category of *simple arteriovenous malformation*, without syndromic features.

Schöbinger staging describes the clinical natural history in four stages [4]: stage I (quiescence) — a pink, warm cutaneous lesion often mistaken for a haemangioma or port-wine stain, without functional impact; stage II (expansion) — growth, thrill and bruit, visible draining veins; stage III (destruction) — ulceration, haemorrhage, pain, necrosis and trophic changes; stage IV (cardiac decompensation) — high-output cardiac failure, life-threatening. Our patient was at stage II, with unequivocal haemodynamic signs but no trophic or cardiac complications, representing an optimal therapeutic window.

The Cho–Do–Yakes angiographic classification, proposed by Cho et al. and refined by Yakes, subdivides peripheral AVMs according to nidal architecture and the artery–nidus–vein relationship, and is today the reference framework for endovascular planning [3,5,8].

Table 2. Cho–Do–Yakes classification and corresponding endovascular strategy

Type	Architecture	Drainage	Preferred strategy
I	≤ 3 arteries draining into a single vein	Single-vein drainage	Coils or vascular plugs
II	Multiple arterioles draining into a dominant outflow vein	Dominant outflow vein	Retrograde transvenous embolization; venous occlusion with plugs/coils
IIIa	Multiple fine fistulous shunts, angiographic "striation" pattern	Multiple micro-shunts	Absolute ethanol or Onyx® via superselective transarterial route
IIIb	Multiple dilated shunts, bulky complex nidus	Multi-axial venous drainage	Ethanol / Onyx® ± direct puncture; multi-route approach often required

In our patient, the bulky complex nidus, multiple dilated shunts and venous drainage through two dominant axes (cephalic vein and ulnar venous network) place the lesion in **type IIIb** —

the most complex subtype, reputedly the most difficult to treat, yet amenable to expert endovascular management.

3.5 Differential diagnosis

Before a pulsatile upper-limb mass, several entities must be systematically considered and excluded: **post-traumatic arteriovenous fistula** (single-pedicle direct arteriovenous communication without a nidus, in a traumatic or iatrogenic context such as arterial puncture or catheterisation); **brachial artery aneurysm** (focal saccular or fusiform arterial dilatation without early venous drainage); **post-traumatic pseudoaneurysm** (perfused sac with a narrow neck and a single arterial communication); **hypervascular tumours** (sarcoma, glomus tumour, haemangioendothelioma, angiosarcoma — an enhancing solid soft-tissue mass without arteriovenous shunting, possibly with bone involvement or metastases); **pure venous malformation** (low flow, predominant phleboliths, no early arterial shunt, delayed opacification only); **lymphatic malformation** (absence of vascular enhancement, multiloculated cystic appearance, fluid–fluid levels); and **involuting infantile haemangioma** (paediatric context with a characteristic growth–regression kinetic). The cardinal semiological rule remains simple: early venous opacification during the arterial phase is the irrefutable radiological signature of arteriovenous shunting and permits confident diagnosis of AVM.

3.6 Imaging strategy

Doppler ultrasound is the first-line examination, identifying high-velocity turbulent flow, low resistive indices and venous arterialisation, but remains operator-dependent with limited three-dimensional mapping. **Multiphasic CT angiography** offers excellent 3D mapping, identification of inflow and outflow vessels, and detection of phleboliths and bone involvement, at the cost of ionising radiation and iodinated contrast nephrotoxicity. **MRI with dynamic MR angiography** (TWIST/TRICKS) provides superior soft-tissue characterisation and shunt dynamics without radiation, constituting an excellent pre-therapeutic assessment, but is limited by cost, availability and contraindications. **Digital subtraction angiography** remains the gold standard, with maximal temporal and spatial resolution, and allows treatment in the same session; being invasive, it is reserved for confirmed cases destined for intervention. CT angiography, as performed in our patient, is currently the pivotal pre-therapeutic tool, delivering a three-dimensional map immediately usable by the interventional team [9,10].

3.7 Management

The anatomical and haemodynamic complexity of AVMs requires systematic multidisciplinary discussion involving the diagnostic radiologist, interventional radiologist, vascular surgeon, reconstructive plastic surgeon, dermatologist and cardiologist (when haemodynamic repercussion exists), with geneticist and paediatrician input in syndromic or paediatric forms [2,4].

Endovascular embolization is the current first-line treatment, governed by three principles: target the nidus and not the feeding arteries; obliterate the shunt while preserving critical vascular axes; and stage the treatment across several sessions. Available agents include absolute ethanol — the most powerful endothelial destructive agent but also the most toxic (cutaneous necrosis, nerve injury, systemic cardiopulmonary effects), reserved for expert operators; Onyx® (EVOH), offering controlled nidal penetration, intrinsic radiopacity and excellent precision; n-BCA acrylic glue, with rapid polymerisation and durable occlusion; and coils or vascular plugs, restricted to type I lesions or to dominant outflow veins (type II). **Direct percutaneous nidal puncture** under ultrasound or fluoroscopic guidance is particularly valuable in type IIIb lesions such as ours, where endovascular access to the nidus is difficult.

Surgical resection is now reserved for well-circumscribed, surgically accessible AVMs; for secondary excision after preoperative embolic devascularisation; and for destructive or haemorrhagic forms threatening life or limb function. The golden rule is to operate after embolization, never before, and never partially — incomplete resection exposes the patient to more aggressive recurrence. **Active surveillance** with periodic clinical and ultrasound follow-up may be proposed for asymptomatic, stable Schöbinger stage I lesions, with patient education regarding decompensation triggers.

Potential complications include spontaneous or provoked haemorrhage, cutaneous ulceration progressing to necrosis, distal ischaemia from vascular steal, high-output cardiac failure in bulky lesions, disabling chronic pain, major psychosocial impact in visible locations, and post-embolization events (skin necrosis, peripheral nerve injury, paradoxical pulmonary embolism, ethanol-induced haemolysis). Prognosis depends on nidus size and complexity, endovascular accessibility, Cho type (IIIa/IIIb being the most challenging), Schöbinger stage at presentation, and above all the expertise of the interventional team. AVMs are not "cured" in the oncological sense — they are controlled; the concept of a chronic disease requiring long-term follow-up must be integrated from the first consultation.

CONCLUSION

This case of a large high-flow arteriovenous malformation of the distal third of the right arm, classified as Cho type IIIb, illustrates the semiological richness and architectural complexity that a congenital vascular lesion of the upper limb may reach. It underscores the radiologist's strategic role in three steps: **identifying** the lesion without being misled by a broad differential; **classifying** it according to Cho and Schöbinger criteria to communicate with clinicians and interventional radiologists in a shared language; and **mapping** the nidus, its inflow and its outflow with precision in order to guide targeted embolization.

In a field where each acquisition phase may reveal or conceal a shunt and where each embolic agent may either salvage or destroy tissue, imaging is not a passive witness but the operative blueprint. An AVM that is correctly interpreted, classified and mapped is already halfway treated. More broadly, this case supports the development of expert vascular anomaly centres, continuing education of young radiologists in this rare but formidable pathology, and wider adoption of modern haemodynamic and angiographic classifications within the radiological community.

FIGURES :

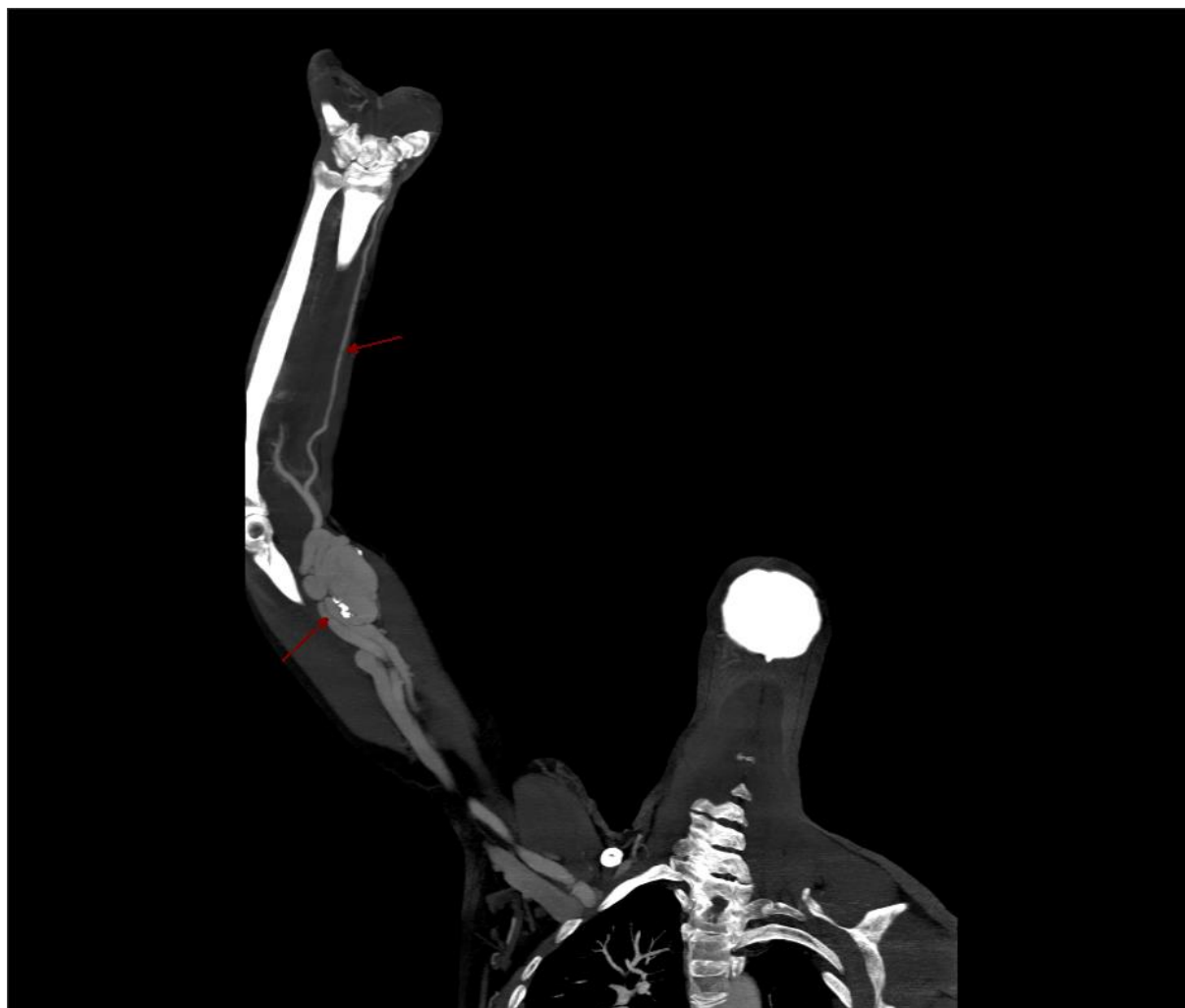


Figure 1 : Coronal Maximum Intensity Projection (MIP) - Arterial Phase
One arrow points to the nidus, and another indicates the brachial artery.

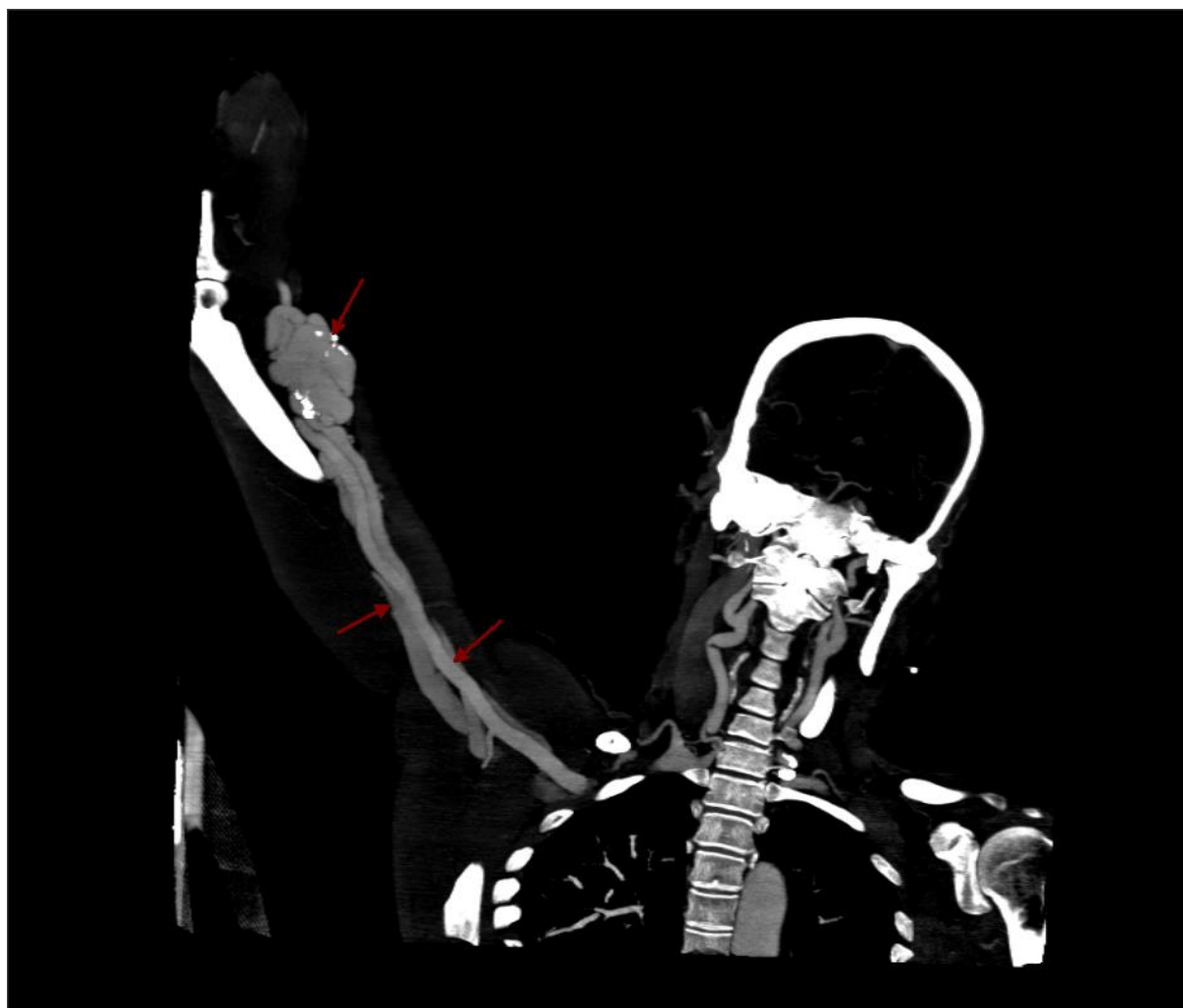


Figure 2 : Coronal Maximum Intensity Projection (MIP) - Venous Phase
Arrows highlight the nidus and its early drainage through the cephalic and axillary veins.



Figure 3 : 3D Volume-Rendered (VR) Reconstruction

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