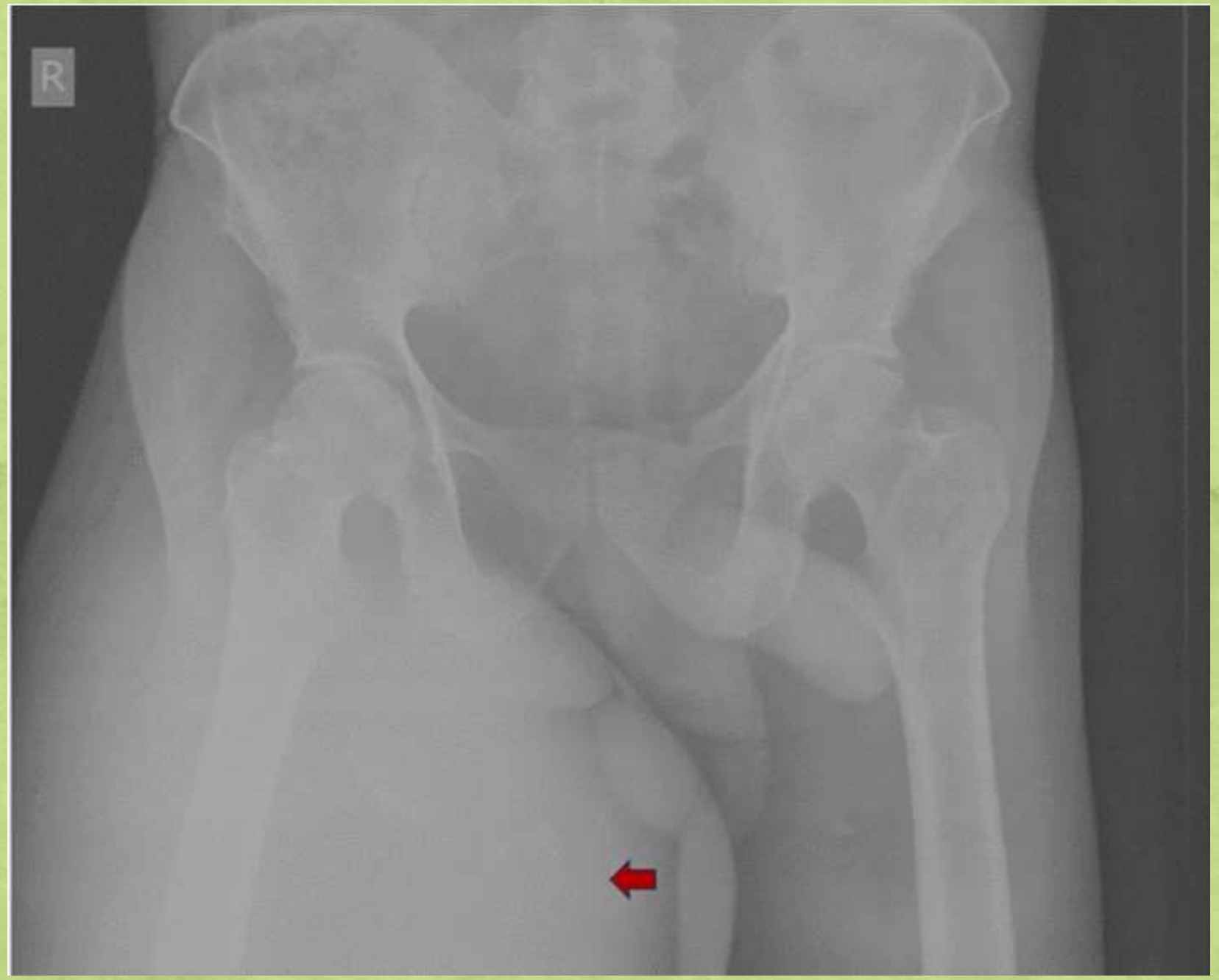
RADIOLOGY CLINICAL MEET FEBRUARY 2025

Sarcomatous Tumor with External Iliac Vein Aneurysm: A Rare Clinical Presentation

CLINICAL HISTORY

A 55 year old male came with the complaints of-

- Swelling in right inguinal region which was insidious in onset and has been gradually increasing in size over the past 12 years
- It was associated with a wound over a right inguinal region which was continuously bleeding for last 4 hours
- For the past 5 years, he also experienced diffuse swelling in both lower limbs (more pronounced on the right side), along with discoloration of the bilateral lower limbs .
- Additionally, he reported right flank pain radiating to the groin over the past year.
- The patient had no other significant clinical or familial history or history of trauma.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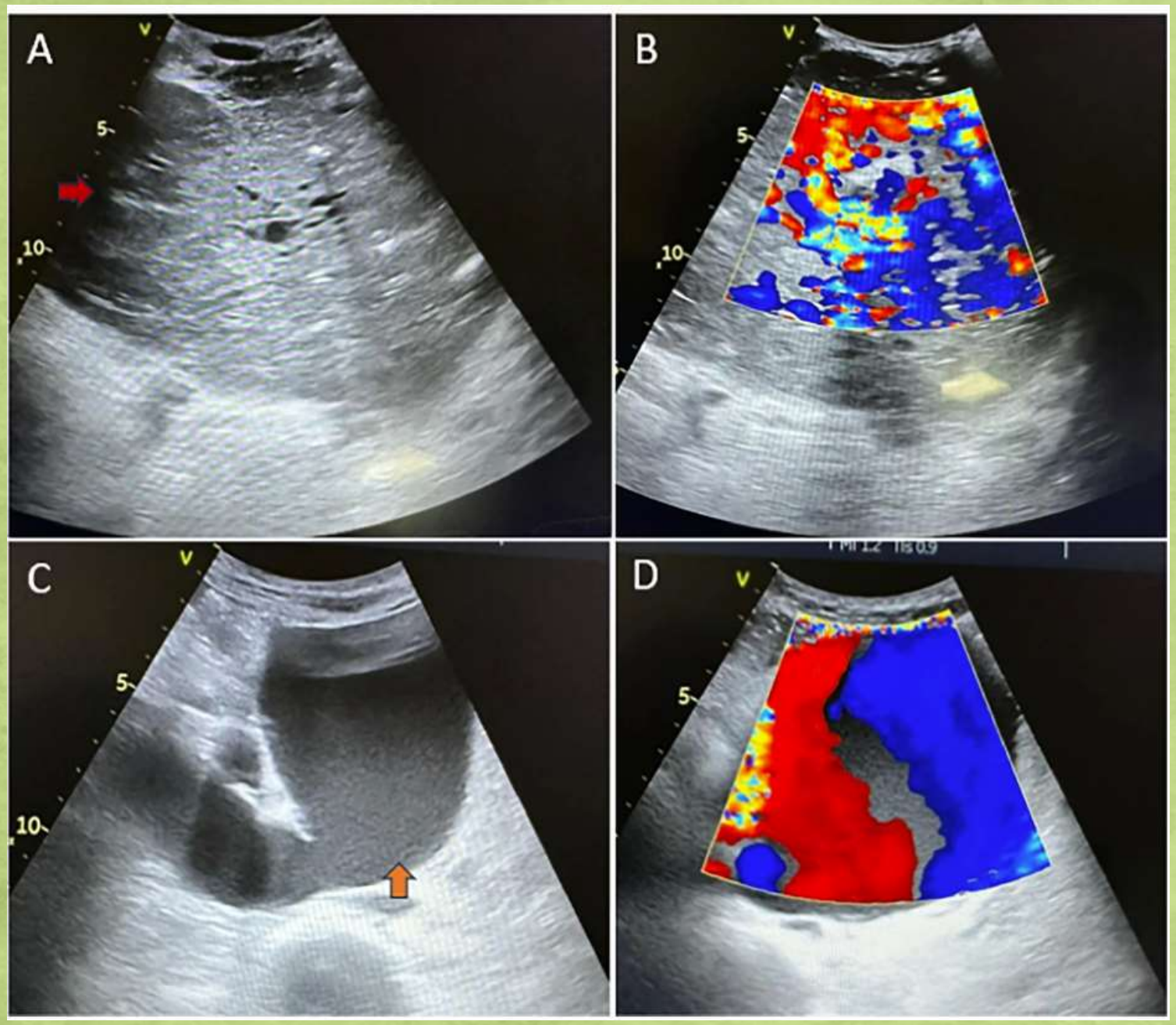
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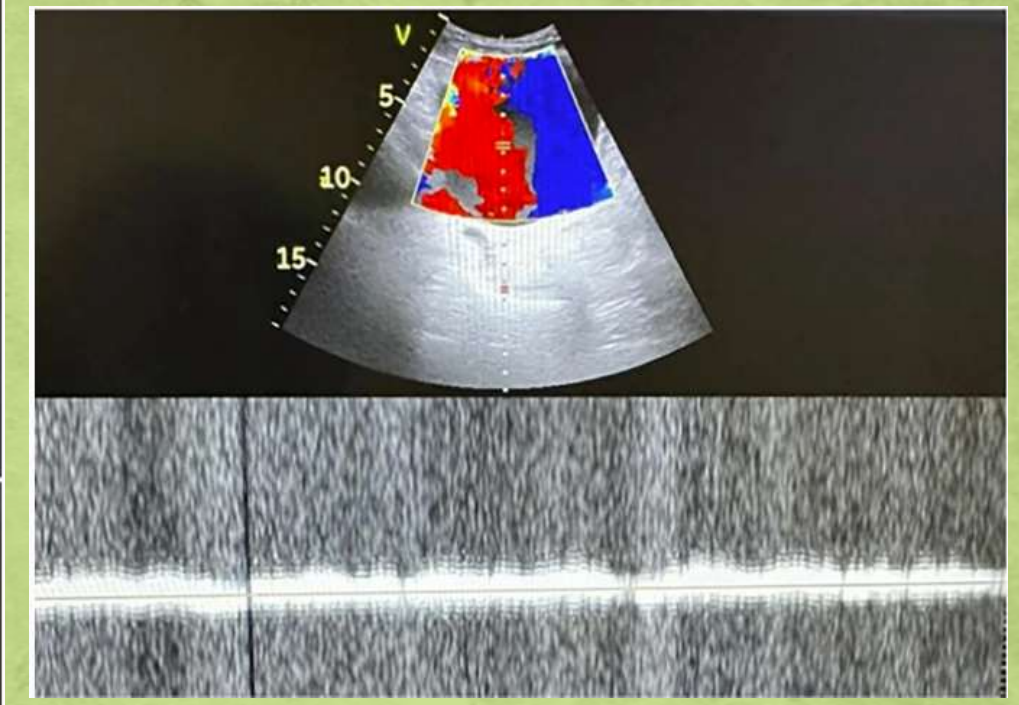

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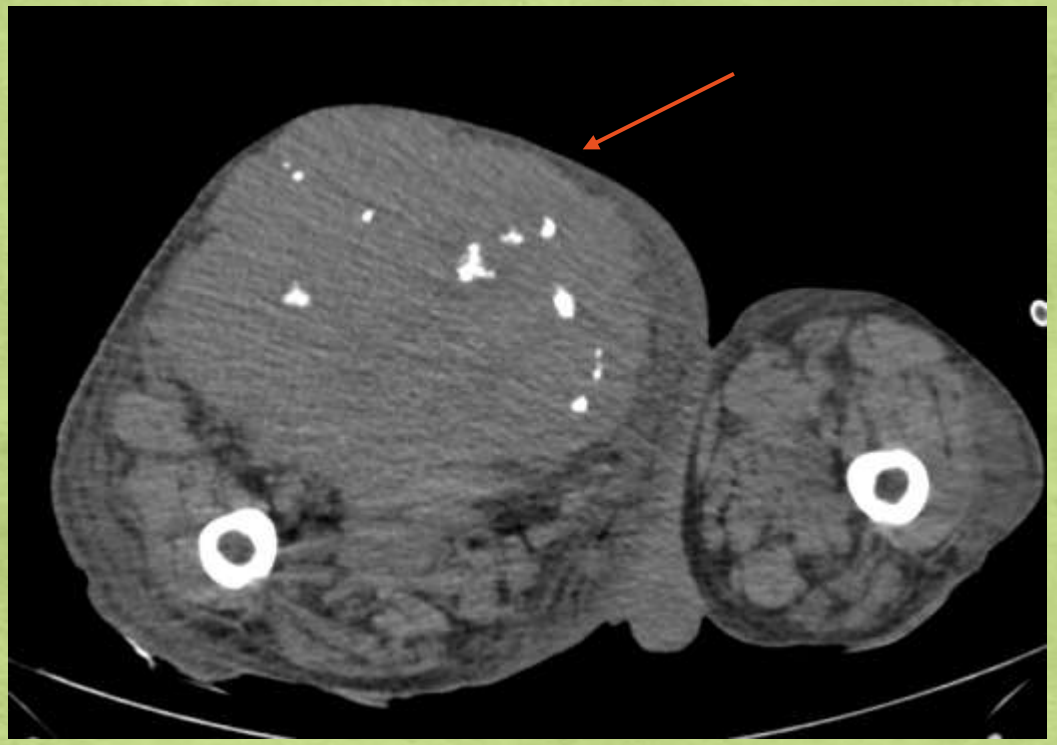

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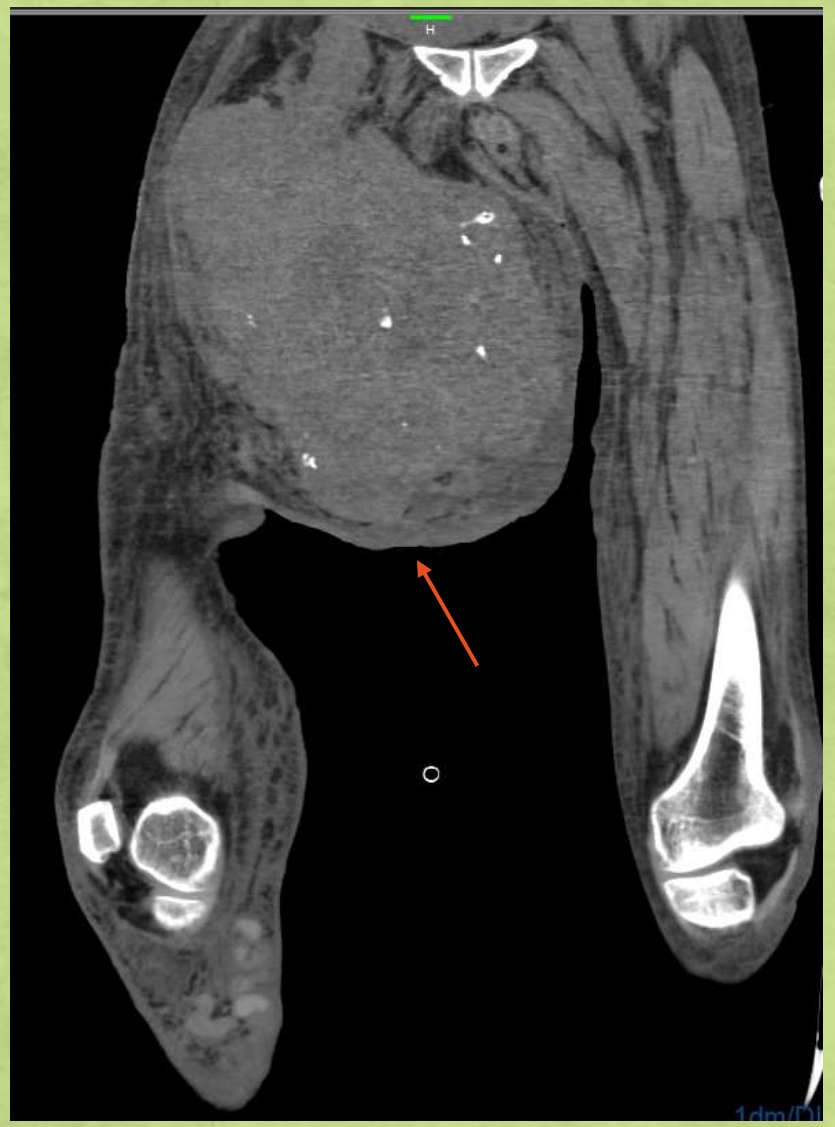

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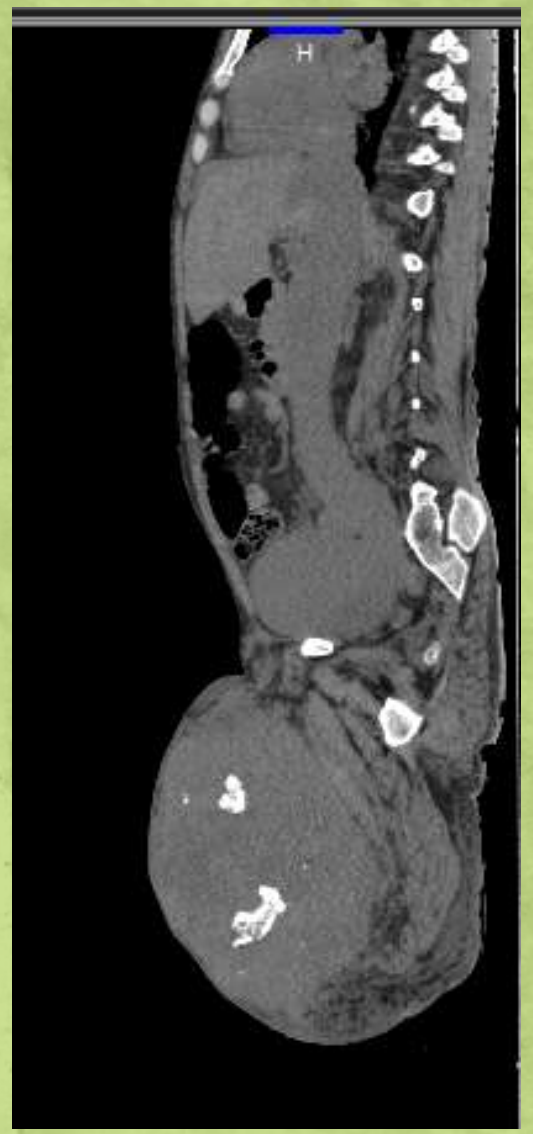

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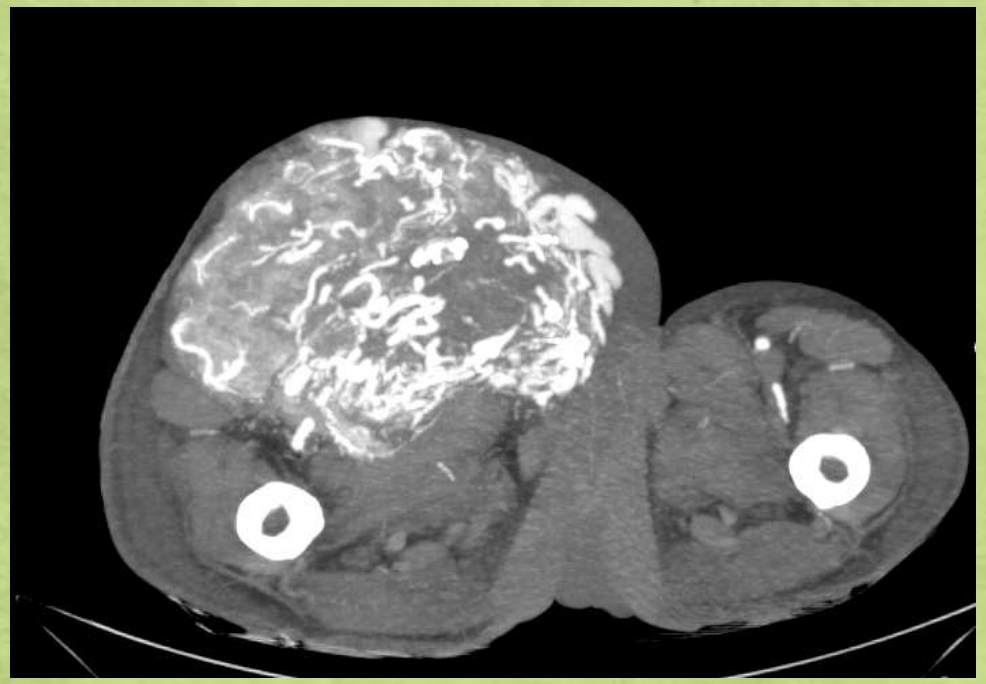

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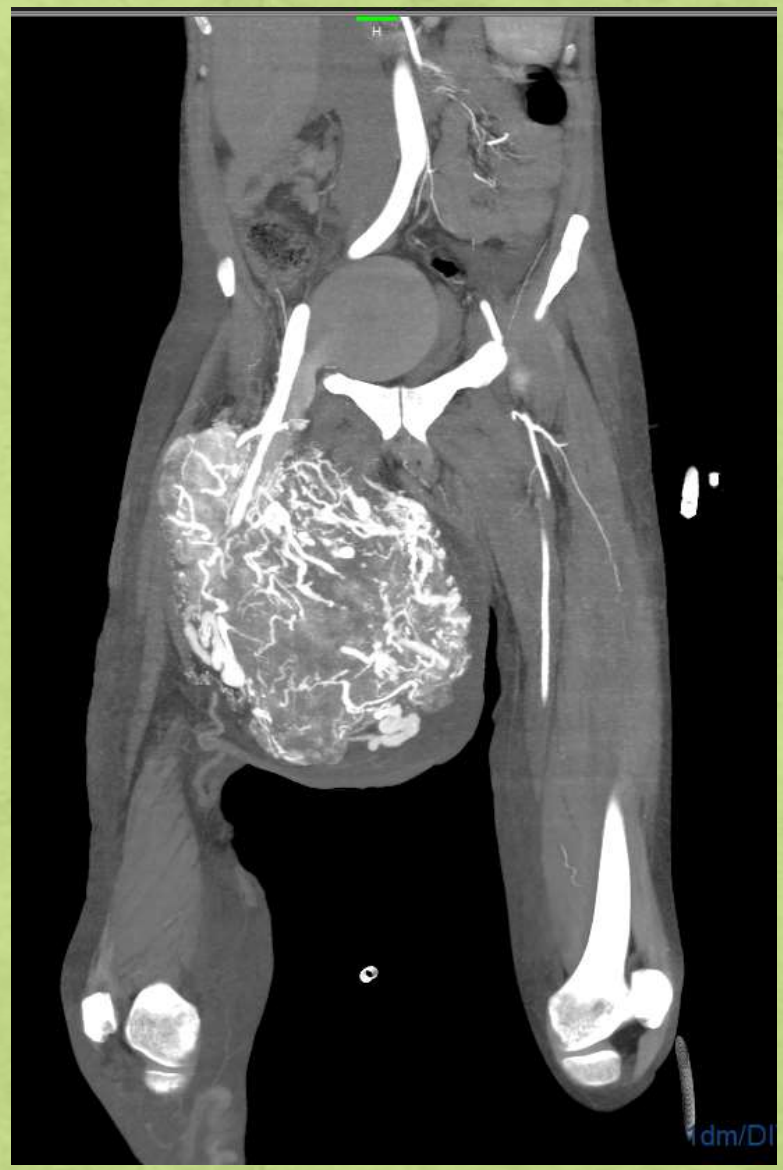

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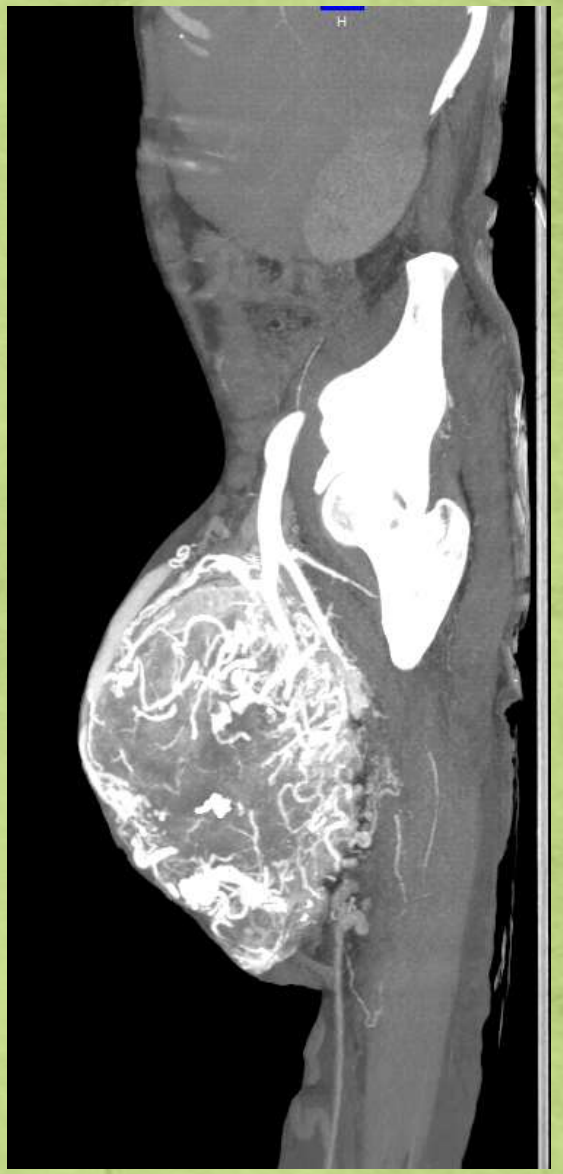

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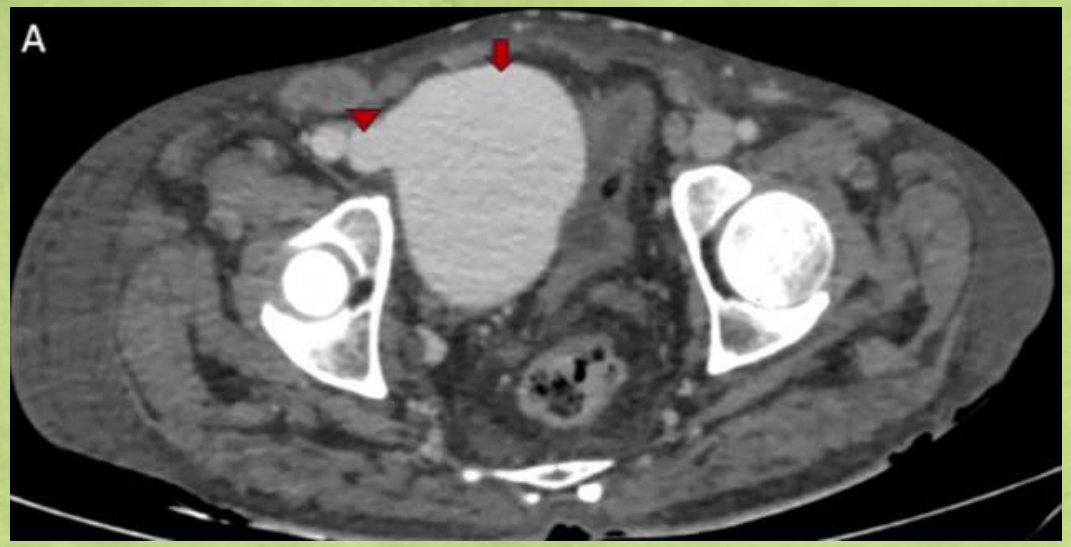
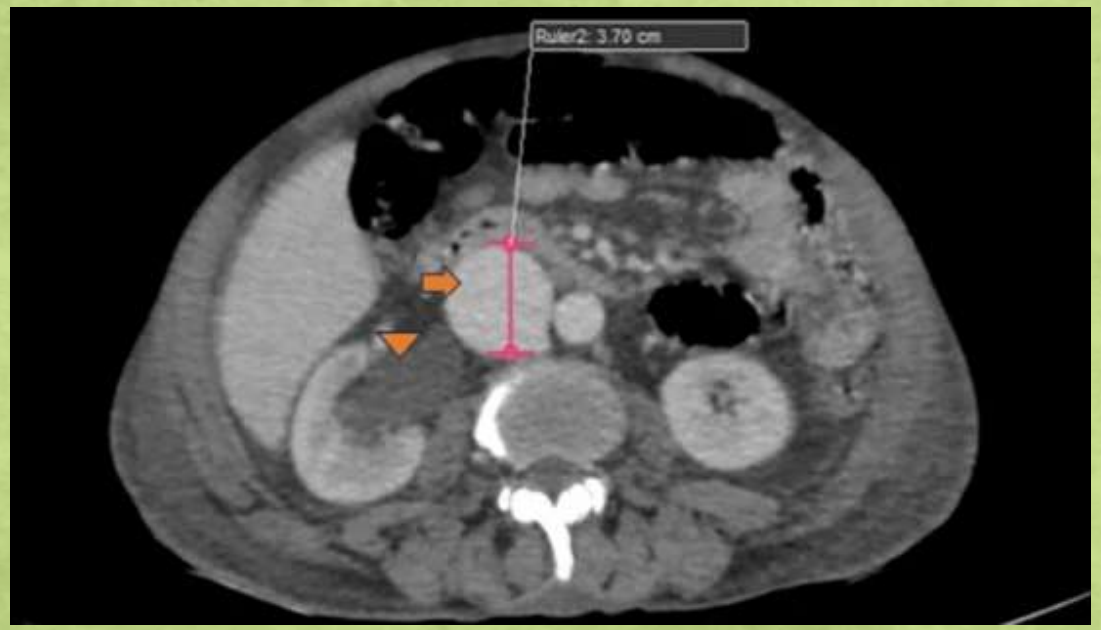
VENOUS





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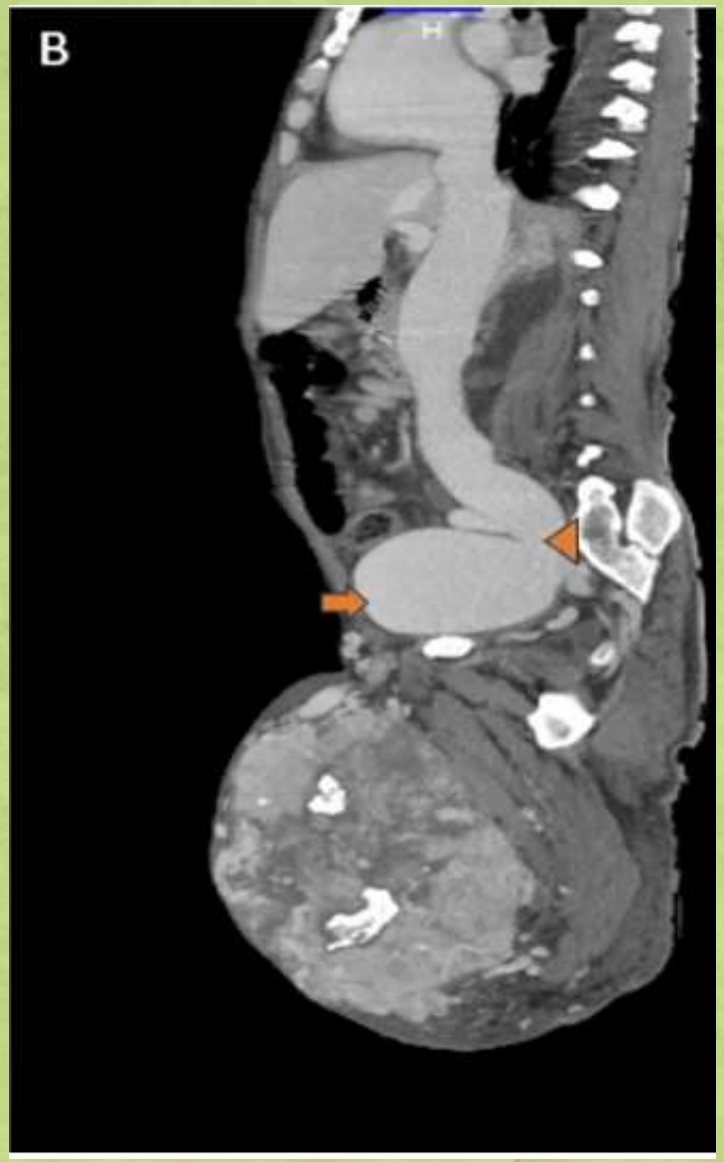

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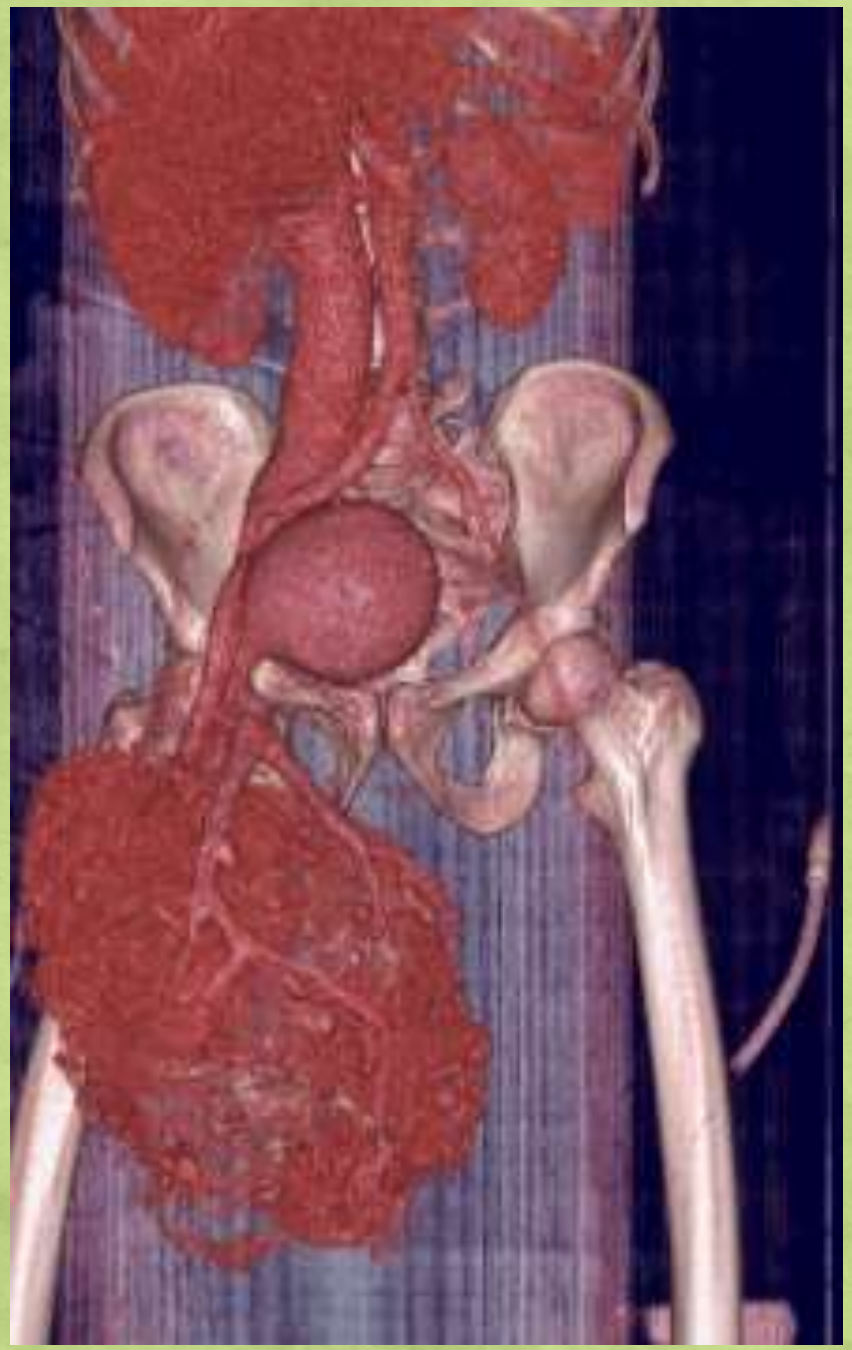

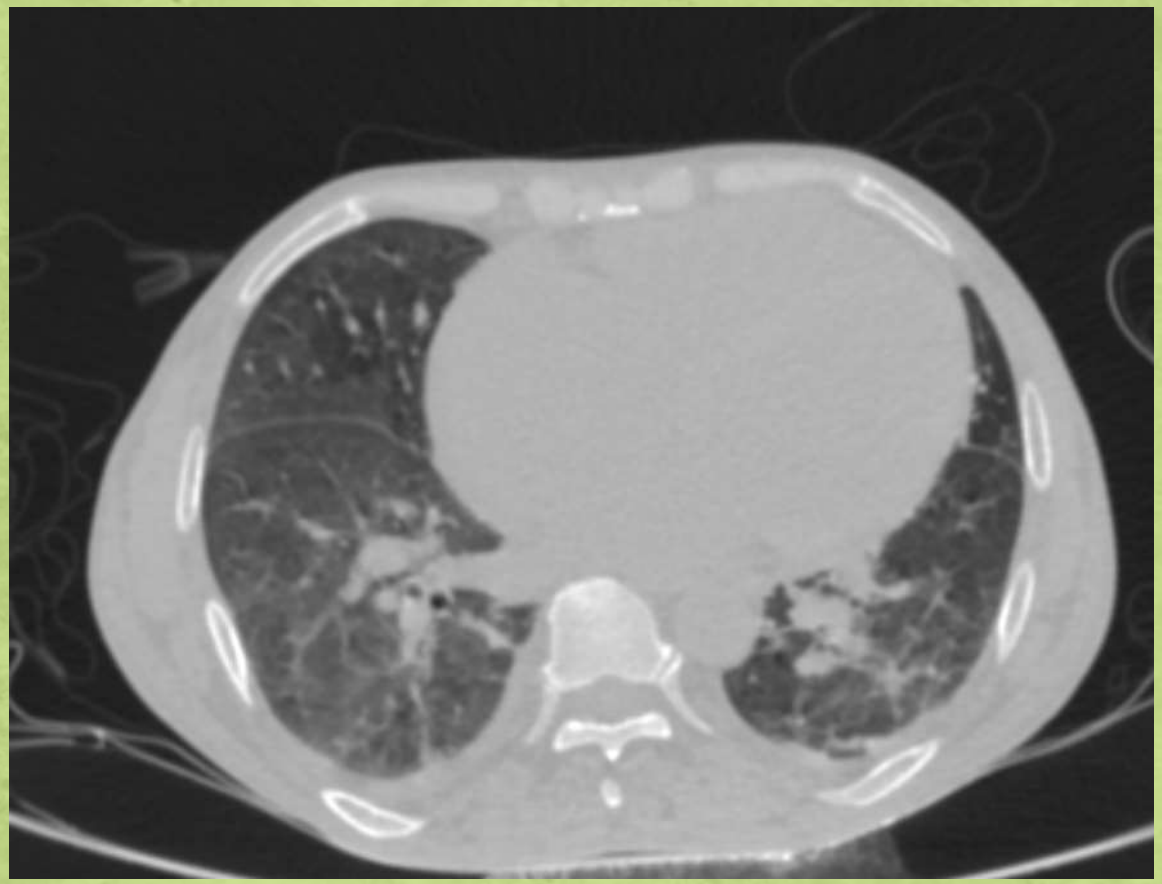

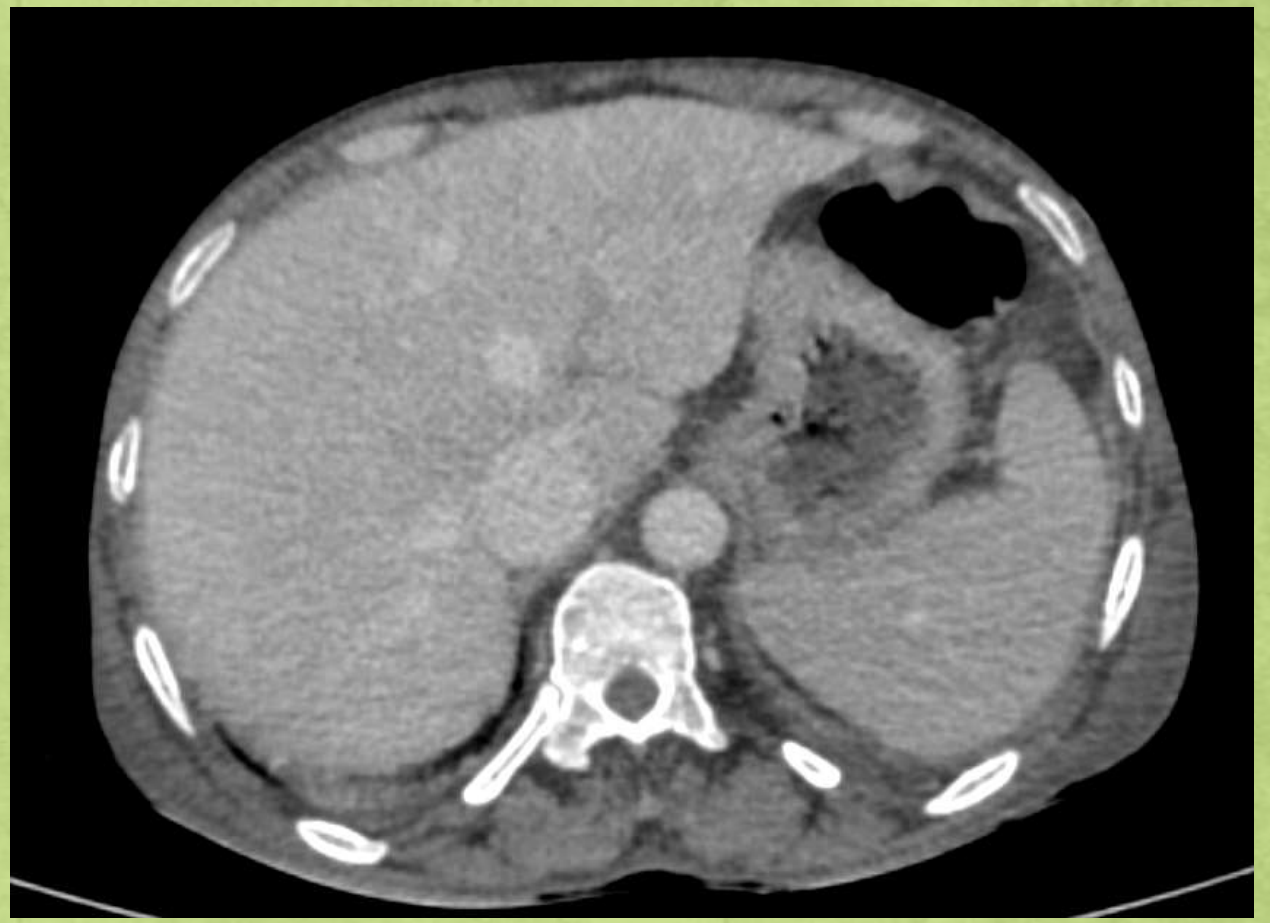

IMPRESSION

- **A large well-defined avidly enhancing mass lesion with multiple arterial feeders and draining veins showing intra tumoral arteriovenous shunting in the anterior and medial aspect of proximal right thigh – Findings suggestive of highly vascular soft tissue sarcoma.**

- **Right external iliac vein shows a large focal dilation at the level of urinary bladder –venous varix compressing right ureter resulting in proximal mild hydronephrosis with hydroureter.**

Feature	Angiosarcoma	Synovial Sarcoma (Vascular Subtype)	Epithelioid Hemangioendothelioma (EHE)	Malignant Peripheral Nerve Sheath Tumor (MPNST)	Metastatic Hypervascular Tumor	Pleomorphic Sarcoma with Possible Rhabdomyosarcoma
Location	Skin, subcutaneous tissue, deep soft tissues (extremities and trunk)	Near large joints (e.g., thigh, knee), usually deep	Extremities (most common), but can occur in liver, lung, and bone	Along major nerves (sciatic, brachial plexus), typically in the thigh	Variable—common in muscles, bones, or soft tissue (e.g., renal cell metastases)	Deep soft tissues (thigh, retroperitoneum, trunk)
Age Group	**50–70 years (older adults)**	**30–60 years (young to middle-aged adults)**	20–40 years (younger adults)	**30–60 years (adults), especially with NF1**	**Middle-aged to elderly (depending on the primary cancer)**	**Middle-aged to elderly (mean age ~50 years)**
Clinical Presentation	Rapidly growing, painless mass; skin discoloration (bruising); may ulcerate	Slow-growing mass, often with mild pain; can compress adjacent structures	Painless, slow-growing; can cause local pain if large	Painful, firm mass with neurological deficits (numbness, weakness)	History of primary malignancy (renal, thyroid, melanoma); rapid growth	Rapidly enlarging, painful mass, often with local invasion
Vascular Characteristics	**Highly vascular, arteriovenous (AV) shunting, and rapid flow**	Prominent vascularity with arterial feeders and venous drainage	Moderate vascularity; delayed contrast enhancement	Moderately vascular, especially in large tumors	Highly vascular, intense enhancement, often with feeding arteries	**Highly vascular, may show arteriovenous shunting**
CT Findings	**Heterogeneous, avid enhancement, necrosis, hemorrhage, AV shunting**	Well-defined, heterogeneous mass, strong contrast uptake, may show calcification	Well-defined, avid enhancement, with central necrosis	Ill-defined, heterogeneous with peripheral enhancement, necrosis	Hypervascular mass, rapid enhancement, and possible bone destruction	**Heterogeneous mass, avid enhancement, areas of necrosis and hemorrhage**
MRI Characteristics	**T1: Iso/hypointense, T2: Hyperintense with flow voids, strong post-contrast enhancement**	Triple sign (hypo-, iso-, hyperintense regions); T2: Hyperintense, post-contrast enhancement	T1: Isointense, T2: Bright with gradual contrast enhancement	T1: Isointense, T2: Heterogeneous with peripheral enhancement, may show target sign	T1: Isointense, T2: Hyperintense, with strong enhancement; often multiple lesions	**T1: Isointense, T2: Heterogeneous with necrosis, post-contrast enhancement**
Angiography	Early venous filling, AV shunting, tortuous vessels	Prominent arterial feeders with early venous drainage	Moderate vascular supply, delayed venous washout	Irregular vessels with slow arteriovenous flow	Hypervascular blush, prominent feeders and draining veins	Prominent feeders, arteriovenous shunting may be seen
Histopathology	Malignant endothelial cells, CD31+, CD34+	Biphasic (epithelial and spindle cells), SYT-SSX fusion	Epithelioid endothelial cells, CD31+, CAMTA1+	Spindle cells, S100-positive, pleomorphism, mitosis	Varies by primary tumor—renal (clear cells), thyroid (papillary structures)	Pleomorphic spindle cells, may show myogenic markers (desmin, myogenin in rhabdomyosarcoma)
Prognosis	Poor, high metastatic potential (lung, liver)	Intermediate, risk of metastasis to lung, bone	Variable, slow progression but can metastasize	Poor, especially in NF1 patients, with frequent recurrence	Depends on the primary tumor; poor if widely metastatic	Aggressive, poor prognosis due to high metastatic potential

DIFFERENTIAL DIAGNOSIS

- Angiosarcoma
- Pleomorphic Sarcoma with Possible Rhabdomyosarcoma

SOFT TISSUE SARCOMA

- Soft tissue sarcomas (STS) are rare malignancies of mesodermal origin. They make up 1% of all malignant tumours and commonly occur in the extremities or trunk.
- The etiology of STS is unknown but some factors are associated with a higher risk, including genetic and environmental factors (e.g., chemical carcinogens), irradiation, viral infections (HHV-8), and immune deficiency.
- STS usually present as painless soft tissue masses, often large at the time of diagnosis, and they metastasize hematogenously, mainly to the lung.
- **Radiographs** have limited utility in diagnosis; however, they can reveal soft tissue swelling and provide insight into the density of the underlying mass. They may also detect calcifications, phleboliths, ossifications, and any involvement of adjacent bones
- **Ultrasound** remains the primary imaging modality of choice and can broadly differentiate between benign and malignant lesions.
- **Malignant lesions typically exhibit a long axis diameter greater than 46 mm, hypervascularity, and heterogeneous solid composition with poorly defined margins.**
- **In contrast, benign lesions are usually smaller (< 46 mm), avascular subcutaneous masses with well-defined margins**

- **Computed tomography (CT)** has a limited role in STS imaging.
- Certain features on CT are considered markers for malignant behaviour, including margin irregularity, infiltration into adjacent organs, calcification, necrosis and hypervascularity.
- STS predominantly metastasises to the lung and so Chest CT is of great importance in STS staging and assessment of disease progression.
- **Magnetic Resonance Imaging (MRI) is the conventional imaging modality of choice** for evaluating soft tissue masses due to its superior soft tissue contrast and multiplanar imaging capabilities. MRI plays an important role in the pre- and post-operative assessment of STS.
- **Role of MRI in Soft Tissue Sarcoma**

1. Differentiates benign from malignant lesions based on signal intensity and enhancement.

Feature	Malignant Characteristics
Size	>5 cm (larger size increases suspicion of malignancy)
Margins	Irregular, infiltrative, or poorly defined borders
T1-Weighted Signal	Iso- to hypo-intense (relative to muscle)
T2-Weighted Signal	Heterogeneous hyperintensity (due to necrosis, hemorrhage)
Enhancement (Post-Contrast)	Heterogeneous, rapid, and irregular enhancement; peripheral rim enhancement (indicative of necrosis)
Internal Composition	Necrosis, hemorrhage, cystic degeneration ("triple sign" in some cases)
Invasion	Adjacent tissue invasion, fascial plane disruption, neurovascular encasement
Flow Voids	Present in highly vascular tumors (e.g., angiosarcoma)
DWI (Diffusion-Weighted Imaging)	Restricted diffusion (low ADC value due to high cellularity)
DCE MRI (Dynamic Contrast-Enhanced)	Rapid early enhancement with washout (indicates malignancy)

2.Tumor Staging & Extent

a) Evaluates tumor size, location, and involvement of surrounding tissues.
b) Assesses compartmental invasion and neurovascular involvement.

3.Preoperative Planning

a) Guides biopsy to viable areas.
b) Defines tumor margins for surgery and aids in radiotherapy planning.

4 .Post-Treatment Monitoring

a) Detects residual disease and recurrence.
b) Evaluates treatment response using size, enhancement, and diffusion patterns.

Patient with Suspicious Soft Tissue Mass

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Initial Evaluation & Clinical History

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Ultrasound (USG)(If superficial mass)
- Defines cystic vs. solid nature
- Limited for deep or large masses

-Helps to check for internal vascularity by color Doppler
- Benign versus malignant features

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MRI (Preferred Imaging Modality)
- Evaluates tumor size, extent, and tissue characteristics
- T1: Iso-/hypointense (relative to muscle)
- T2: Hyperintense (heterogeneous with necrosis)
- Contrast: Heterogeneous enhancement, necrosis, invasion

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CT Scan(If MRI unavailable or for staging)
- Useful for detecting calcifications and bone involvement
- Chest CT for lung metastasis evaluation

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Biopsy (Definitive Diagnosis)
- Image-guided core needle biopsy (preferred)
- Histopathological confirmation and grading

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Staging Workup
- Chest CT (lung metastases)
- PET-CT (if metastatic disease suspected)
- Regional lymph node evaluation (rare but critical)

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Multidisciplinary Management Decision –
- Surgery ± Radiation ± Chemotherapy
- Monitor with MRI for recurrence or treatment response

- **In our case, because of arteriovenous shunting of blood—potentially attributed to the high arterial vascularity in the soft tissue sarcomatous tumor—may explain the hemodynamically elevated venous flow and venous pressure that lead to the development of a venous aneurysm.**

- On examination, aneurysms can be misinterpreted as soft tissue tumors, adnexal masses, or inguinal hernias .

- Clinically, they may present with vague symptoms such as lower back pain and lower limb swelling, primarily related to local compression.

- The primary investigation for diagnosing iliac vein aneurysms includes duplex ultrasound scan with color Doppler. However, the preferred imaging modalities are CT venography, real-time venography, and MRI

- Complications of venous aneurysms include rupture and thromboembolism, which can occur due to embolus dislodgement from chronic thrombus formation within the aneurysm.

TREATMENT

- Treatment options for iliac vessel aneurysms include surgical excision followed by venorrhaphy, patch repair, or venous bypass .
- For smaller aneurysms, conservative management and post-surgical prophylaxis with anticoagulant therapy are accepted treatments
- Treatment for the sarcomatous tumor typically begins with neoadjuvant therapy with surgical removal.
- Surgical management involves wide local excision with 2-cm margins ,en bloc resection including involved vessels, and immediate vascular reconstruction using either autologous vein graft or synthetic materials.
- Postoperative care includes anticoagulation, careful wound management, and completion of adjuvant therapy if indicated.

Conclusions

This case highlights the rare presentation of soft tissue sarcoma complicated by external iliac venous aneurysm, illustrating the critical relationship between highly vascularized sarcomas and secondary venous complications. The fatal outcome due to aneurysmal rupture emphasizes the need for prompt, aggressive management.

Key learning points include

1) the necessity of comprehensive vascular imaging
2) Vigilance for venous complications in vascularized tumors
3) Urgent intervention to prevent catastrophic outcomes.

THANK YOU