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PELVIC SOLITARY FIBROUS TUMOR, HISTORICALLY CLASSIFIED AS HEMANGIOPERICYTOMA, PRESENTING WITH VENOUS COMPRESSION AND PELVIC CONGESTION: A CASE REPORT

Dejan SVILAR[1,2], Jovana ĐOŠIĆ[2], Anđela ĐURIĆ[2], Bojan STOJANOVIĆ[3,4]*

[1]Department of Radiology, Faculty of Medical Sciences, University of Kragujevac, Kragujevac, Serbia, [2]Department of Radiological Diagnostics, University Clinical Centre Kragujevac, Kragujevac, Serbia, [3]Department of Surgery, Faculty of Medical Sciences, University of Kragujevac, Kragujevac, Serbia. [4]Clinic for General Surgery, University Clinical Centre Kragujevac, Kragujevac, Serbia

Corresponding Author
Dejan Svilar
Department of Radiology, Faculty of Medical Sciences, University of Kragujevac, Department of Radiological Diagnostics, University Clinical Centre Kragujevac, Kragujevac, Serbia
Email: swilar@gmail.com

ABSTRACT

Introduction: Solitary fibrous tumour is a rare fibroblastic mesenchymal neoplasm historically classified as hemangiopericytoma. Although it can occur at almost any anatomical site, pelvic involvement is uncommon, and symptoms often develop late as a consequence of progressive mass effect.

Case presentation: A 69-year-old man presented with abdominal pain, progressive swelling of the right lower limb, and paraesthesia. Laboratory investigations revealed mild anaemia, elevated fibrinogen levels, and mildly increased C-reactive protein, while serum tumour markers remained within normal limits. Multidetector computed tomography demonstrated a large lobulated hypervascular abdominopelvic mass with heterogeneous enhancement and central necrosis. Computed tomographic angiography revealed compression of the right iliac vein, femoral venous dilatation, and marked pelvic venous congestion. Colour Doppler ultrasonography demonstrated abnormal venous flow without definitive evidence of thrombosis.

Conclusion: Solitary fibrous tumour is a rare but clinically significant entity that should be considered in the differential diagnosis of large hypervascular pelvic masses. A multidisciplinary approach is essential for optimal treatment planning, while surgical resection remains the primary therapeutic modality.

Keywords: solitary fibrous tumour; hemangiopericytoma; pelvis; multidetector computed tomography; Colour Doppler ultrasonography.

Introduction

Solitary fibrous tumour (SFT) is an uncommon fibroblastic mesenchymal neoplasm characterised by a wide anatomical distribution, variable biological behaviour, and a distinctive immunohistochemical and molecular profile. The term hemangiopericytoma was historically used to describe highly cellular spindle-cell tumours with prominent branching vascular channels. However, contemporary classifications place many of these lesions within the SFT spectrum rather than as a separate clinicopathological entity (1,2). This shift in terminology is clinically important because the diagnosis now relies not only on morphology and vascular architecture but also on immunohistochemistry, particularly nuclear STAT6 expression, which serves as a reliable surrogate marker of the characteristic NAB2::STAT6 fusion (3,4).

Pelvic SFTs are rare. Their deep location and slow growth often allow them to reach considerable size before diagnosis. Symptoms are therefore often non-specific and may include abdominal or pelvic pain, urinary complaints, bowel symptoms, lower-extremity swelling, or neurological symptoms caused by compression of pelvic organs, vessels, or nerves. Large pelvic lesions may mimic other soft-tissue tumours, including leiomyosarcoma, liposarcoma, gastrointestinal stromal tumour, paraganglioma, or vascular malformations, making preoperative diagnosis challenging.

Imaging plays a central role in lesion characterisation and operative planning. Contrast-enhanced multidetector computed tomography (MDCT) may reveal a well-demarcated, lobulated, hypervascular soft-tissue mass with heterogeneous enhancement and, in larger tumours, necrotic or cystic degeneration. Computed tomographic angiography (CTA) and Doppler ultrasonography may provide additional information on vascular supply, venous compression, altered flow dynamics, and the risk of thrombotic or congestive complications. Nevertheless, definitive diagnosis requires tissue sampling and histopathological confirmation.

We present a case of a large pelvic SFT, historically classified as hemangiopericytoma, that manifested with abdominal pain, right lower-limb swelling, and radiologically evident pelvic venous congestion. The case highlights the importance of integrating cross-sectional imaging, vascular assessment, and immunohistochemistry in the diagnostic work-up of rare hypervascular pelvic tumours.

Case Presentation

A 69-year-old man presented with abdominal pain of several months' duration. He also reported swelling and tingling in the right lower limb. During hospitalisation, he underwent baseline laboratory testing and imaging investigations.

Routine haematological analysis demonstrated mild anaemia, with haemoglobin of 126 g/L (reference range, 138-175 g/L) and haematocrit of 0.375 L/L (reference range, 0.415-0.530 L/L). Further laboratory testing revealed elevated fibrinogen levels of 6.06 g/L (reference range, 2.00-4.50 g/L) and a mild increase in C-reactive protein to 14.20 mg/L (reference range, 0.00-5.00 mg/L). Serum tumour markers, including carcinoembryonic antigen, alpha-fetoprotein, prostate-specific antigen, and neuron-specific enolase, were within reference limits.

Imaging Findings

Native and contrast-enhanced MDCT of the abdomen and pelvis demonstrated a large lobulated tumour occupying much of the abdominopelvic cavity. The lesion was highly vascular, with marked post-contrast enhancement and a central area of necrosis. It measured 145 x 110 x 123 mm in the anteroposterior, transverse, and craniocaudal dimensions, respectively. The mass exhibited an expansive growth pattern, compressing adjacent structures without unequivocal CT signs of direct infiltration. On imaging grounds, the main differential diagnoses included soft-tissue sarcoma, leiomyosarcoma, liposarcoma, and gastrointestinal stromal tumour. A smaller presacral lesion with similar imaging characteristics was also noted along the right rectal wall, measuring 30 x 22 mm in the axial plane.

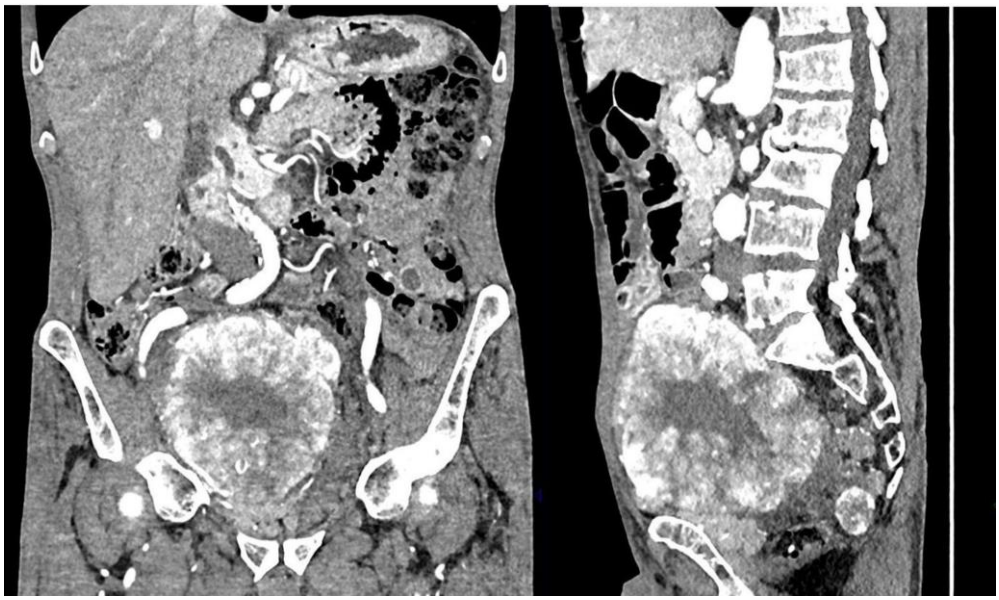

Figure 1. Contrast-enhanced MDCT showing a large hypervascular abdominopelvic mass with heterogeneous enhancement and expansive growth.

CTA demonstrated marked pelvic venous congestion. The tumour compressed the right iliac vein, whereas the right common femoral vein was dilated, measuring approximately 22 mm in diameter and irregularly opacified, raising a strong suspicion of thrombosis on CT. Congestion of both spermatic veins and both dorsal penile veins was also present. The remaining vascular findings were within expected limits for the patient's age.

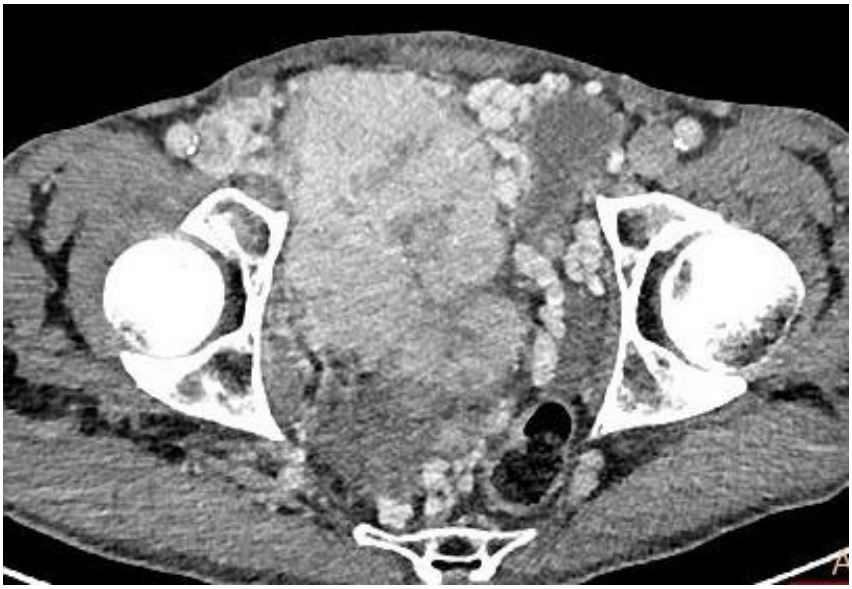

Figure 2. Axial CT image illustrating the pelvic component of the mass and associated vascular congestion.

Given the marked venous abnormalities affecting the lower limbs, Colour Doppler ultrasonography was performed to evaluate the deep and superficial venous systems. Although the veins were moderately dilated, the examination revealed no definitive evidence of superficial or deep venous thrombosis. Venous blood flow was markedly slowed and reversed direction during the Valsalva manoeuvre. Instead of the expected continuous, monophasic flow pattern modulated by respiration, spectral Doppler showed discontinuous flow with peaks above and below the baseline, indicating alternating, non-physiological changes in flow direction associated with respiration.

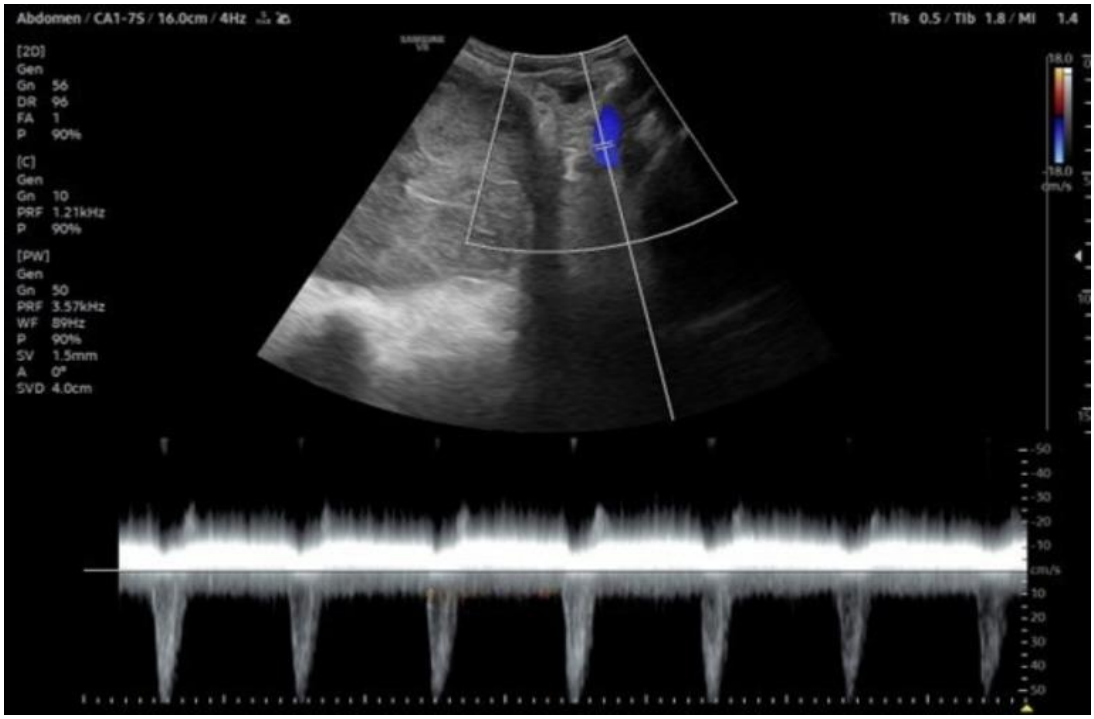

Figure 3A. Color Doppler ultrasonography demonstrating altered flow dynamics in the iliac venous system.

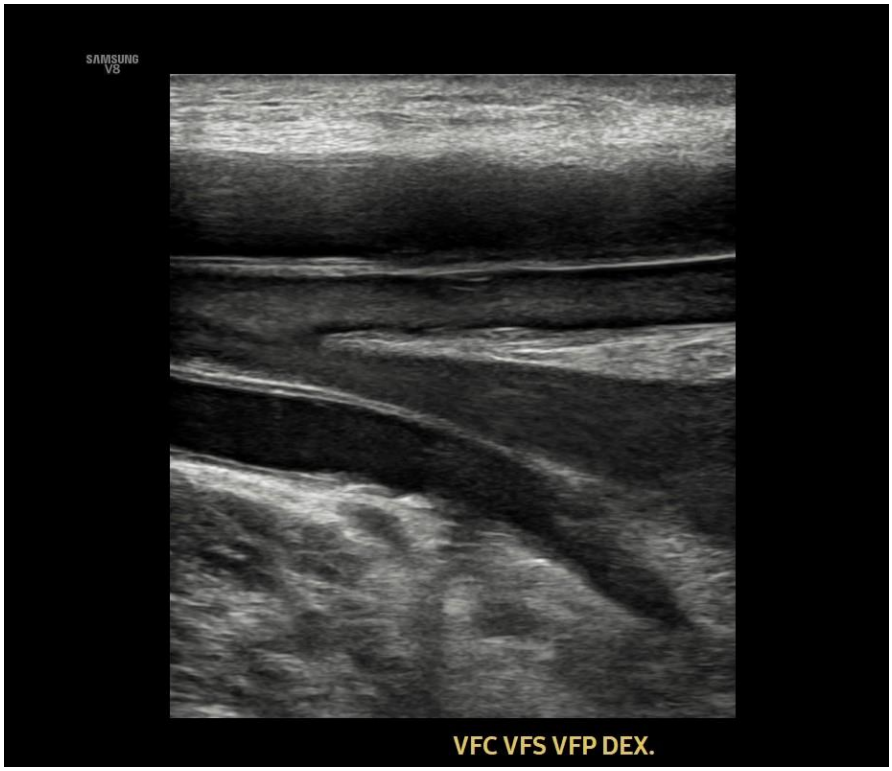

Figure 3B. Ultrasonographic image of lower-extremity venous dilatation without definitive evidence of thrombosis.

Histopathological and Immunohistochemical Assessment

To establish the histological diagnosis and confirm the nature of the lesion, an ultrasound-guided percutaneous core needle biopsy was performed using a 16-gauge needle, yielding four tissue cores. Immunohistochemical analysis demonstrated expression of STAT6, CD34, vimentin, CD99, CD10, and CD57. Tumour cells were negative for CD117, desmin, S-100, SMA, BCL-2, CD31, DOG1, and HMB-45. The Ki-67 proliferation index was approximately 5%. Taken together, the morphological and immunohistochemical findings supported the diagnosis of a solitary fibrous tumour, historically classified as hemangiopericytoma, WHO grade I.

Multidisciplinary Decision

A multidisciplinary team comprising an internist-oncologist, a radiation oncologist, and an abdominal surgeon reviewed the case. Given the size, pelvic localisation, hypervascular appearance, and clinically significant venous compression, they recommended referral to a specialised oncological surgery centre for definitive surgical management.

Discussion

This case illustrates several features that make pelvic SFT diagnostically and therapeutically challenging. First, the tumour reached a considerable size before diagnosis, consistent with the behaviour of deep-seated pelvic and retroperitoneal lesions. Such tumours may remain clinically silent for prolonged periods and become symptomatic only after compressing adjacent structures. In the present case, the predominant clinical manifestation was not a palpable painless mass but abdominal pain accompanied by right lower-limb swelling and paraesthesia, reflecting the mechanical and haemodynamic consequences of a large pelvic lesion.

Second, the radiological appearance was suggestive but not pathognomonic. The combination of a well-defined, lobulated contour, strong contrast enhancement, central necrosis, and a prominent vascular component raised suspicion for a hypervascular mesenchymal tumour. However, considerable imaging overlap exists with leiomyosarcoma, liposarcoma, gastrointestinal stromal tumour, paraganglioma, and other vascular tumours. Consequently, radiological findings should be interpreted as part of a broader diagnostic framework rather than relied upon as a sole basis for diagnosis.

The venous findings merit particular attention. CTA suggested compression of the right iliac vein and raised suspicion of thrombotic involvement of the right common femoral vein. In contrast, Colour Doppler ultrasonography showed no evidence of established deep or superficial venous thrombosis at the time of examination.

Instead, Doppler assessment revealed venous dilatation, markedly reduced flow velocity, and alternating flow direction influenced by respiratory dynamics and the Valsalva manoeuvre. This discrepancy highlights the complementary value of anatomical and functional vascular assessment in patients with large pelvic tumours. In such cases, lower-limb swelling may result from extrinsic venous compression and pelvic venous congestion even in the absence of confirmed thrombosis.

The final diagnosis relied on histopathological and immunohistochemical evaluation. Nuclear STAT6 expression, together with CD34 positivity and exclusion of GIST, smooth muscle, neural, endothelial, melanocytic, and other spindle-cell neoplasms, strongly supports the diagnosis of SFT. The low Ki-67 index and WHO grade I classification suggest relatively indolent biological behaviour. Nevertheless, SFTs are well recognised for their unpredictable clinical course. Several risk-stratification systems have identified large tumour size, necrosis, increased mitotic activity, positive surgical margins, advanced age, and recurrent disease as factors associated with a higher risk of local recurrence or metastasis (5,6). Accordingly, even apparently low-grade tumours require careful risk assessment and long-term surveillance.

Complete surgical excision with negative margins remains the cornerstone of treatment for localised SFT and offers the best chance of durable disease control (7,8). In pelvic tumours, surgical management may be technically demanding because of tumour size, hypervascularity, and proximity to the iliac vessels, ureters, rectum, bladder, and pelvic nerves. Preoperative planning should therefore include detailed vascular mapping, evaluation of potential venous obstruction, and assessment of whether selective embolisation may reduce the risk of intraoperative bleeding. The roles of radiotherapy and systemic therapy are less clearly defined and are generally reserved for selected cases, including unresectable disease, positive margins, recurrence, or malignant and aggressive histological features.

This case further emphasises the importance of multidisciplinary management. Radiologists, pathologists, oncologists, vascular specialists, and surgeons each contribute essential information regarding lesion morphology, vascular consequences, tissue diagnosis, oncological risk, and operative feasibility. For rare tumours such as pelvic SFT, such collaboration is not merely desirable but fundamental to safe, effective, and rational treatment planning.

The key diagnostic, radiological, therapeutic, and clinical findings are summarised in Table 1. The table highlights their clinical significance and provides practical insights that may assist clinicians in evaluating similar rare pelvic tumours.

Table 1. Summary of the key diagnostic, radiological, therapeutic, and clinical features of the present case of pelvic solitary fibrous tumour (historically classified as hemangiopericytoma).

Aspect	Findings in the present case	Diagnostic relevance	Radiological relevance	Therapeutic implication	Clinical significance	Take-home message
Tumor localisation	Large abdominopelvic/ pelvic mass.	Pelvic SFT/haemangiopericytoma is rare and may be difficult to recognise clinically.	MDCT precisely defined tumour size, extension, and relationship to surrounding structures.	Referral to a specialised surgical oncology centre was recommended.	Pelvic localisation increases the risk of compression of vessels, nerves, the urinary tract, and bowel.	Rare pelvic masses should be evaluated with both oncological and vascular considerations.
Tumor morphology	Lobulated, expansile, well-defined soft-tissue mass with central necrosis.	Suggests a mesenchymal tumour, but it is not specific enough for definitive diagnosis.	Strong post-contrast enhancement supports a hypervascular lesion.	Preoperative planning must consider tumour vascularity and possible bleeding risk.	Hypervascularity may complicate biopsy and surgical excision.	Imaging suggests the diagnosis, but histopathology confirms it.
Vascular effects	Compression of the right iliac	Explains the patient's clinical symptoms	CTA and Colour	Surgical planning	Venous compression may	In pelvic SFT,

	vein with pelvic venous congestion and right-leg swelling.	and supports the mass-effect mechanism.	Doppler demonstrated altered venous haemodynamics.	should include assessment of the major pelvic vessels.	mimic thrombosis or primary vascular disease.	vascular compression can be a major clinical manifestation.
Colour Doppler findings	Venous dilation with a non-physiological alternating flow pattern.	Demonstrates the functional consequence of extrinsic venous compression.	Adds dynamic information that CT alone cannot fully provide.	Helps evaluate the need for vascular monitoring and perioperative thrombosis prevention.	Useful in patients with limb swelling or suspected venous obstruction.	Doppler ultrasonography complements CT in assessing the haemodynamic impact.
Differential diagnosis	Leiomyosarcoma, liposarcoma, GIST, desmoid tumour, and other pelvic soft-tissue tumours.	A broad differential diagnosis requires tissue confirmation.	Imaging overlap is considerable among pelvic soft-tissue tumours.	The treatment strategy depends on accurate tumour classification.	Misclassification may lead to inappropriate therapeutic decisions.	Large hypervascular pelvic tumours require a multidisciplinary diagnostic work-up.
Immunohistochemistry	STAT6, CD34, vimentin, CD99, CD10, and CD57 positivity; CD117, DOG1, desmin, SMA, S-100, HMB-45 negativity.	STAT6 nuclear expression strongly supports the diagnosis of SFT.	Correlates with imaging features of a hypervascular mesenchymal tumour.	Confirms that treatment should be planned as SFT rather than GIST or smooth-muscle tumour.	Immunohistochemistry is essential for the final diagnosis.	STAT6 is a key diagnostic marker in modern SFT classification.
Proliferative activity	Ki-67 index approximately 5%.	Suggests relatively low proliferative activity.	Does not exclude clinically relevant behaviour.	Complete excision and follow-up remain necessary.	Low Ki-67 is favourable, but tumour size and necrosis still require caution.	SFT behaviour may be unpredictable even when proliferation is low.
Risk assessment	Advanced age, large tumour size, and necrotic areas.	These features may increase concern for recurrence or aggressive behaviour.	Necrosis and size are visible on imaging and should be reported.	Long-term surveillance is required after treatment.	Risk stratification should not rely only on histological grade.	Even apparently low-grade SFTs require prolonged surveillance.
Treatment approach	Multidisciplinary recommendation for surgical referral.	Surgery remains the main therapeutic option for localised disease.	Imaging defines resectability and relationship to pelvic structures.	Complete resection with negative margins is the goal.	Complex pelvic localisation requires experienced surgical teams.	Multidisciplinary management is essential for optimal outcomes.
Overall value of the case	Rare pelvic SFT with venous compression and haemodynamic consequences.	Demonstrates the need for integrated radiological and pathological diagnosis.	MDCT, CTA, and Doppler provided complementary information.	Supports individualised surgical planning.	The case expands awareness of unusual presentations of pelvic SFT.	Pelvic SFT should be considered in large hypervascular masses with vascular compression.

Conclusion

This case highlights pelvic solitary fibrous tumour, historically classified as hemangiopericytoma, as a rare but important diagnostic consideration in the evaluation of large, hypervascular abdominopelvic masses with compressive vascular manifestations. In the presented patient, MDCT, CTA, and Colour Doppler ultrasonography provided complementary information on tumour morphology, vascularity, central necrosis, pelvic venous congestion, and altered lower-limb venous flow. Definitive diagnosis ultimately relied on histopathological and immunohistochemical confirmation, particularly STAT6 and CD34 positivity. Although the low Ki-67 index suggested relatively low proliferative activity, the patient's advanced age, large tumour size, and necrosis indicate the need for careful risk assessment, complete surgical excision whenever feasible, and prolonged postoperative surveillance. This case, therefore, emphasises the importance of a multidisciplinary approach integrating radiology, pathology, vascular assessment, and surgical oncology in the diagnosis and management of rare pelvic mesenchymal tumours.

Learning Points

Large pelvic SFTs may present with venous compression and congestion rather than with tumour-specific clinical manifestations.

MDCT, CTA, and Colour Doppler ultrasonography provide complementary information on tumour morphology, vascular relationships, and haemodynamic consequences.

STAT6 and CD34 positivity, together with the exclusion of key spindle-cell mimics, are central to establishing the diagnosis of SFT.

Surgical excision remains the cornerstone of treatment, but preoperative planning should account for tumour hypervascularity, venous compression, and potential involvement of pelvic structures.

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REVIEW ARTICLE

SOLITARNI FIBROZNI TUMOR KARLICE, RANIJE KLASIFIKOVAN KAO HEMANGIOPERICITOM, KOJI SE MANIFESTOVAO KOMPRESIJOM VENA I VENSKOM KONGESTIJOM U KARLICI: PRIKAZ SLUČAJA

Dejan SVILAR[1,2], Jovana ĐOŠIĆ[2], Anđela ĐURIĆ[2], Bojan STOJANOVIĆ[3,4]*

[1] Odeljenje za radiologiju, Fakultet medicinskih nauka, Univerzitet u Kragujevcu, Kragujevac, Srbija
[2] Odeljenje za radiološku dijagnostiku, Univerzitetski klinički centar Kragujevac, Kragujevac, Srbija
[3] Katedra za hirurgiju, Fakultet medicinskih nauka, Univerzitet u Kragujevcu, Kragujevac, Srbija
[4] Klinika za opštu hirurgiju, Univerzitetski klinički centar Kragujevac, Kragujevac, Srbija

SAŽETAK

Uvod: Solitarni fibrozni tumor (SFT) predstavlja retku mezenhimalnu neoplazamu, ranije klasifikovanu kao hemangiopericitomom. Iako se može javiti gotovo na bilo kojoj anatomskoj lokaciji, karlična lokalizacija je retka, a simptomi se obično javljaju kasno zbog progresivnog efekta mase.

Prikaz slučaja: Šezdesetdevetogodišnji muškarac javio se, zbog višemesečnog abdominalnog bola, otoka i trnjenja desne noge. Laboratorijski nalazi pokazali su blagu anemiju, povišen fibrinogen i blago povećan C-reaktivni protein, dok su tumorski markeri bili uredni. Multidetektorska kompjuterizovana tomografija abdomena i karlice, prikazala je masivnu lobuliranu hipervaskularnu tumorsku masu sa heterogenim postkontrastnim pojačanjem i centralnom nekrozom. Color-Doppler ultrasonografija vena donjih ekstremiteta, pokazala je usporen i nefiziološki venski protok, bez jasnih znakova tromboze dubokih vena u trenutku pregleda.

Zaključak: SFT male karlice predstavlja retku, ali klinički značajnu dijagnozu u diferencijalnom razmatranju velikih, hipervaskularnih tumorskih promena karlice. Multidisciplinarni pristup je od ključnog značaja za planiranje optimalnog lečenja, pri čemu hirurška resekcija ostaje osnovni terapijski modalitet.

Ključne reči: solitarni fibrozni tumor; hemangiopericitom; karlica; multidetektorska kompjuterizovana tomografija; kolor dopler ultrasonografija.