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Case Report

A rare case of asymptomatic retroperitoneal and thigh femoral nerve schwannoma ☆,☆☆

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ABSTRACT

We present a rare case of an asymptomatic extensive tumor in the retroperitoneum and thigh, which has not been published in the scientific literature so far. During the preventive examination, in an otherwise healthy 29-years old man, a tumor mass in the lower left abdominal quadrant was observed on ultrasound. A computed tomography scan and magnetic resonance imaging revealed a large, lobulated, multiseptated retroperitoneal mass stretching from the level of the lower left kidney pole to the distal third of the left thigh. Positron emission tomography/ computed tomography showed weak metabolically active mass with low probability of malignancy, and radiological differential diagnosis included lymphangioma, less likely lymphoma and as least likely sarcoma. Due to the assumed liquid content of the tumor, no preoperative biopsy was performed. A demanding and long-lasting operation was performed on Department of abdominal surgery at Clinical Hospital Merkur. The tumor was removed completely along with most part of the femoral nerve. Pathohistological analysis showed a benign tumor of the nerve sheath- schwannoma. A novel aspect in the presentation of this clinical entity is the extensiveness of the neoplasm to multiple regions without symptoms. According to the available literature, only 2 similar case reports are described, which were symptomatic.

☆ Competing Interests: The authors of case report "A Rare Case of Asymptomatic Retroperitoneal and Thigh Femoral Nerve Schwannoma" declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper. One of authors is the patient's sister, which further motivated her to present an interesting and rare case to other colleagues.

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Introduction

Schwannoma is rare tumor, affecting predominantly females and mostly found in patients 20 to 60 years of age, originating from Schwann cells which form sheath that provides protection and support of peripheral nerves [1]. It usually affects nerves in the head, neck and extremities. Retroperitoneal location is rare, approximately 3 % of all schwannomas [2]. Only 4 % of all retroperitoneal tumors are schwannomas [3]. A finding that is not specific but can point to a neural origin of a tumor is a mass in the retroperitoneum, originating proximally to the spine, in the psoas muscle, or involving the neural foramina [4]. It is usually a benign tumor and rarely undergoes malignant transformation unless associated with neurofibromatosis, which is the case with 5 % to 18 % of all retroperitoneal schwannomas [2]. There are no radiological characteristics that can differentiate between benign and malignant form of schwannoma [4]. Generally, schwannomas are clinically silent and discovered incidentally and symptoms, if present, are unclear and nonspecific [5].

The preoperative diagnosis is usually difficult to obtain and the definitive diagnosis is made upon complete surgical excision, as the gold standard for the management of these tumors, and histopathological examination [6]. In primary adult retroperitoneal tumors of mesenchymal origin, there is often a mismatch between tumor volume and the small number of symptoms when the mass is found [4].

Case presentation

The preventive systematic examination was performed in a healthy 29-years old man. His mother died at the age of 40 from aortic dissection, grandfather and uncle from chronic lymphatic leukemia, other uncle from liver failure of unknown cause. At the age of 3 he had inguinal hernia operation, right leg UGFS (ultrasound guided foam sclerotherapy) of superficial varicose veins at the age of 19, and left varicocele operation at the age of 20. He did not take any medications. Patient's physical examination was without notes. In laboratory findings increased values of total cholesterol, total bilirubin, urate and iron were found. On abdominal ultrasound there were observed a border finding of fatty liver and a tumor mass in the lower left abdominal quadrant. Since the examination was performed in a private institution, no ultrasound figures were available, and the patient received only the written report. The mass was described as inhomogeneous, scantly vascularized tumor, 12×7 cm in size. Patient did not have any abdominal pain or leg swelling. Only after prolonged periods of standing, approximately 6-7 h at work, did he experience pain in both legs. Key clinical observation was an undefined tumor mass in lower left abdomen which had to be differentiated quickly.

The multiphasic (non-contrast phase, followed by contrast enhanced late arterial and venous phase) computed tomography scan of the proximal abdomen, pelvis and upper legs was performed (Figs. 1-3). The CT scan revealed large, slightly lobulated left retroperitoneal mass, spreading from the level of the lower left kidney pole to the distal third of the left thigh (Fig. 2). The mass was 90 mm in the largest transverse diameter, sharply and well marginated from adjacent major psoas and iliacus muscle, iliopsoas, sartorius and vastus medialis muscle, as well as common and superficial left artery and vein, with no clear signs of infiltration (Fig. 1A-C). It had relatively homogenous structure, with few visible septa showing no signs of significant contrast enhancement (Fig. 1C). The tumor had dense fluid density (HU between 17 and 24) with no signs of significant vascularization. According to anatomical spread and other radiological characteristics, the differential diagnosis primarily suggested for retroperitoneal lymphatic malformation, less likely lymphoma and least likely myo-/liposarcoma.

Compared to the right leg, there was significant hypotrophy of sartorius muscle and atrophy of vastus medialis muscle (Fig. 3). All vascular structures of abdomen, pelvis and left thigh were patent and in normal size, and there were no pathological morphological findings of abdominal and pelvic organs and structures. In all presented regions lymph nodes were normal in size. Laboratory work-up to rule out lymphoma was done with values within referent range. Positron emission tomography computed tomography (PET/CT) whole body scan found tumor mass to be weakly metabolically active with FDG standardized uptake value (SUV) of maximal 3,5, which was primarily assumed to be result of low uptake in the lymph and was not indicative for malignancy. In the other scanned areas, there were no signs of pathological FDG uptake. Magnetic resonance imaging (MRI) completed

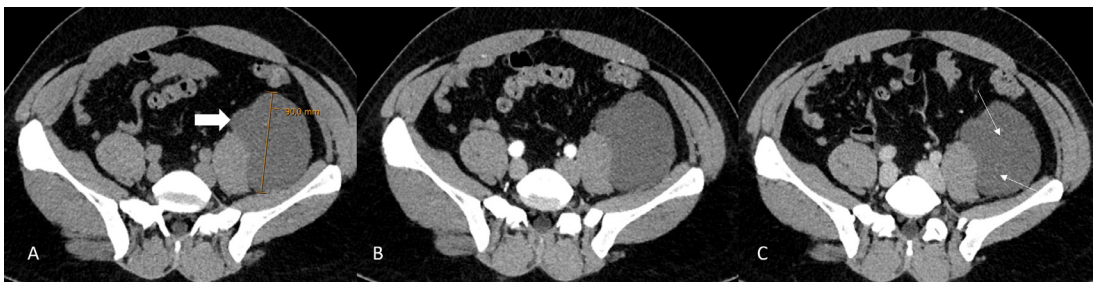

Fig. 1 – CT- Axial non-enhanced (A) and multiphasic contrast- enhanced (B late arterial phase, C venous phase) An expansive, slightly lobulated mass in the left retroperitoneum, with the largest transverse diameter of 90 mm (white thick arrow on A). The mass is sharply marginated from the adjacent psoas and iliac muscle, has a homogeneous structure, with few thin septa (white thin arrows on C), with no significant contrast enhancement or vascularization.

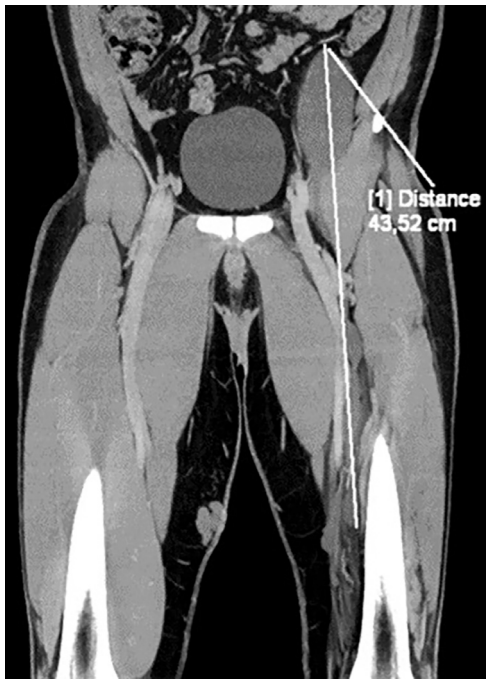

Fig. 2 – Contrast-enhanced CT oblique coronal MIP reconstruction showing polilobulated retroperitoneal mass spreading along common and superficial artery and vein to the level of the distal third of the left thigh (craniocaudal longest diameter 43,5 cm).

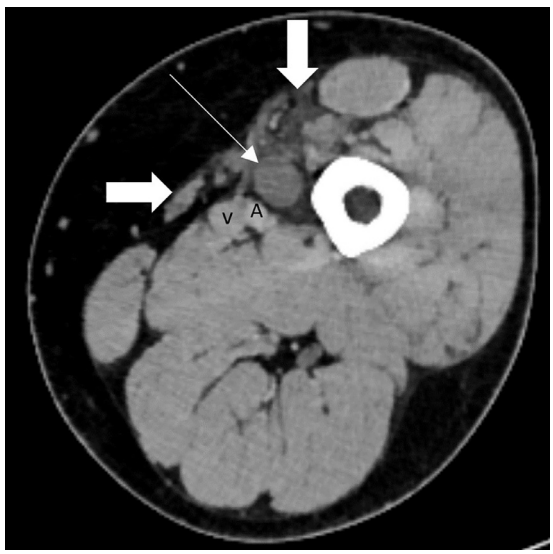

Fig. 3 – Axial contrast-enhanced CT (level of the middle third of the left thigh) showing significant hypotrophy of the sartorius muscle (white thick right arrow) with medial vastus muscle atrophy (white thick downwards arrow); (white thin arrow- schwanomma, V- femoral vein, A- superficial femoral artery).

and clarified morphological findings of the tumor (Fig. 4A-E). It was sharply marginated from adjoint structures, multiseptated and filled with fluid or slightly cellular matrix. Within the tumor, few nodular thickenings were found, showing mild vascularity and mild diffusion restriction (Fig. 4D,E). MRI also clarified very mild contrast enhancement of multiple intratumorally septa (Fig. 4A-C).

No trial biopsy was performed due to the presumed watery content of the tumor.

Initially, the Multidisciplinary Team (MDT) for urogenital tumors at UHC Zagreb recommended surgical intervention. However, this course of action was abandoned due to the patient's overall good general condition, and a decision was made to proceed with regular follow-up instead. Subsequently, the patient expressed a strong desire to have the tumor removed, and in collaboration with the abdominal surgeon, the surgical procedure was carried out.

While waiting for the operation, the patient started feeling left leg pain with increasingly visible superficial veins. The operation lasted 4 h, the femoral nerve was removed 1 cm from its origin in the spinal cord up to the distal third of the femur. The intraoperative diagnosis using the frozen section was performed and the tumor was mesenchymal, probably originating from peripheral nerve. The soft and white tumor 31 cm in length was removed completely. In the wider part, which was 15 cm long, the tumor was up to 8 cm in diameter and in the narrower part that was 16 cm long the diameter was 2 to 3 cm. Histologically, the tumor consisted of variously large, predominantly smaller nodules composed of spindle-shaped cells without polymorphia and mitosis (Fig. 5A). There were more cellular areas (Antoni A) with nuclear palisading (Verocay bodies) and a hypocellular component (Antoni B) (Fig. 5B). Between the described nodules, there was a loose connective stroma with blood vessels and individual tumor cells (Fig. 5C). Tumor cells were immunohistochemically intensely positive for S-100 (Fig. 5D). Perineurium (immunohistochemically EMA+) was preserved and there were parts of a peripheral nerve in some of the slides (Fig. 5E). The resection margins were negative.

The final diagnosis was plexiform schwannoma.

After the operation, intensive physical therapy was carried out. Two years after the procedure, the patient is a healthy and cheerful married young man, with an almost fully functional left leg.

Discussion

Patient presented in this case had no symptoms at the time the tumor was discovered, but after 2 months he started having certain difficulties. As previously described, preoperative diagnosis of schwannoma is challenging due to lack of specific radiological features on CT and MRI [7]. Positron emission tomography scanning can be a useful tool but it cannot reliably differentiate malignant from benign schwannomas [8].

Even a percutaneous biopsy has been reported to be inaccurate for the characterization of these tumors [9]. In most other published case reports of retroperitoneal schwannoma, patients had symptoms, unlike this patient, regardless of size

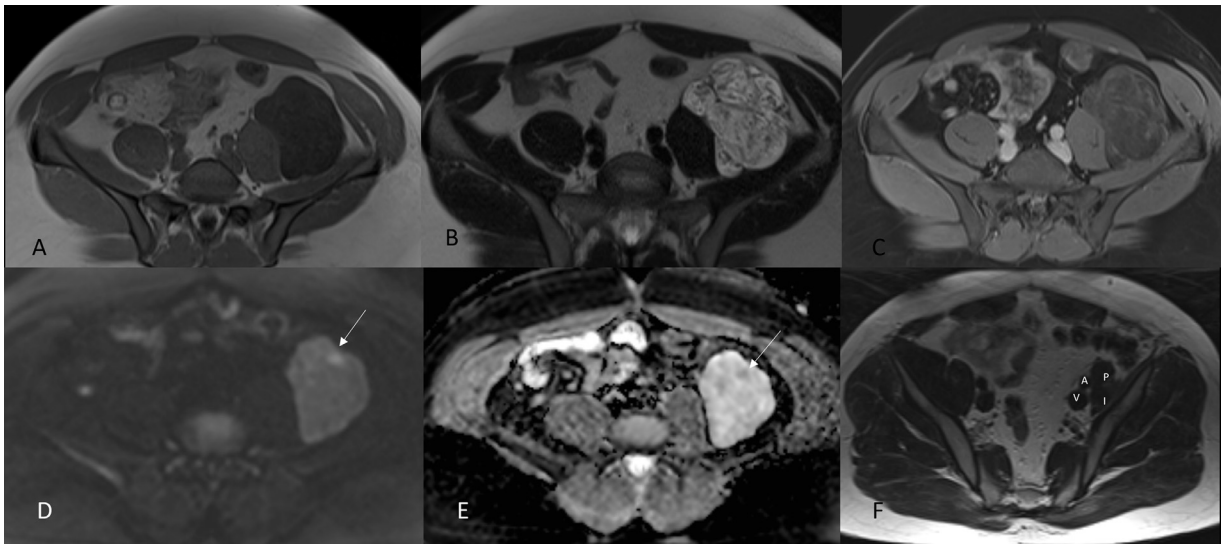

Fig. 4 – MRI imaging clarified morphological tumor characteristics depicking multiple intratumorous septa (B) with mild contrast enhancement (C), and fluid or slightly cellular matrix among septa. Few nodular septa thickenings with mild diffusion restriction and mild contrast enhancement (D and E) were also seen. A 12-month-follow-up MRI showed no residual od recurrent disease (F; A- external iliac artery; V- external iliac vein; P-psoas muscle; I- iliac muscle).
4A. T1 axial non-enhanced. 4B. T2 axial 4C. T1 fs axial contrast enhanced. 4D. DWI, b800 4E. ADC map. 4F. T2 axial (12-month-follow-up).

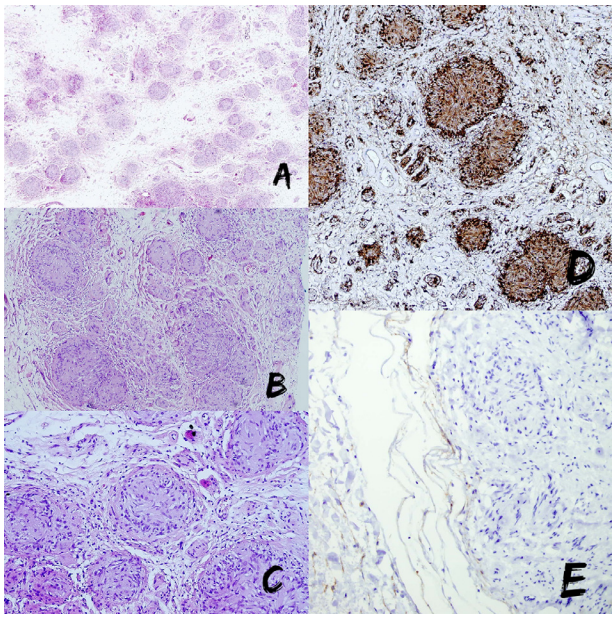

Fig. 5 – Digital photographs of the pathology slides from the tumor specimen.
Findings: Image A shows a tumor consisting of variously large nodules composed of spindle cells and a loose connective stroma between the nodules. Image B demonstrates more cellular areas (Antoni A) with nuclear palisading - Verocay bodies and hypocellular areas (Antoni B). Image C shows monomorphic spindle cells without any signs of polymorphia or mitoses. Image D demonstrates positivity of spindle cells for S100. Image E shows preserved perineurium which is EMA positive.

[1,2,5–8]. In 2 case reports, extremely large schwannomas were found to affect both the pelvis and thigh, respectively the abdomen and thigh. The patients in these cases had been symptomatic for 13 and 15 years, respectively [10,11]. It can be assumed that our patient would have sought help soon because of pain in his leg, if the workup of the tumor mass in the retroperitoneum had not been started before. Schwannoma of the femoral nerve is one of the differential diagnoses of leg pain [12].

Diagnostic imaging, like in many other retroperitoneal tumors plays a crucial role in both identification and preliminary lesion characterization, even though imaging findings are nonspecific [13]. Thanks to a preventive ultrasound a benign and non-infiltrative but extensive tumor was discovered. Thanks to the dedicated work and multidisciplinary cooperation of hematology, radiology, nuclear medicine, oncology, pathology and surgery specialists, the tumor was completely excised, and the patient cured. A 12- month-follow-up MRI of pelvis and left thigh revealed no residual or recurrent disease (Fig. 4F).

Conclusion

Retroperitoneal schwannomas are rare, slowly growing and mostly benign although they can be extensive, and in young and otherwise healthy patients can manifest with minimal, if any symptoms. Detailed and careful radiological workup is very important in the assumption of diagnosis, but only surgical removal and histopathological analysis can confirm a benign form of the tumor.

Author contributions

Concept and design: JN; Acquisition, analysis and interpretation of data: JN, DM and IJ; Drafting the article: JN, LS and DM; Revising it critically for important intellectual content: DJ, IJ; Approved final version of the manuscript: JN, LS, DJ, DM and IJ.

Patient consent

Author obtained written informed consent from the patient for submission and publication of case report "A Rare Case of Asymptomatic Retroperitoneal and Thigh Femoral Nerve Schwannoma" for publication.

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