CASE REPORT

UDC: 616-006.48-07
DOI: https://doi.org/10.2298/VSP180819122N

Schwannoma of the abdominal wall – diagnostic challenge

Švanom u abdominalnom zidu – dijagnostički izazov

Bojan Nikolić*, Biserka Vukomanović Djurdjević†‡, Miroslav Mitrović§, Jelena Golubović*, Jasna Pešić*

Military Medical Academy, *Institute of Radiology, †Institute of Pathology, §Clinic for General Surgery, Belgrade, Serbia; ‡University of Defence, Faculty of Medicine of the Military Medical Academy, Belgrade, Serbia

Abstract

Introduction. Schwannoma or neurilemmoma is a benign tumor of the nerve sheath originating from the Schwann cells. Localization in abdominal wall is rare. Schwannomas usually manifest themselves as slow-growing asymptomatic tumors. Symptomatology depends on the location, the involved nerve and the size of the tumor. **Case report.** We present a 43-year-old female patient with a schwannoma localized in the right hypochondriac region. Diagnostic procedures included ultrasound with color Doppler (US), high-resolution magnetic resonance imaging (MRI), and histopathological examinations (HP). US showed a solid, well-circumscribed mass, with a whorled, echogenic internal architecture in the anterior abdominal wall. MRI revealed an oval, well-circumscribed, heterogeneous, 3 × 2.5 × 2.5 centimeters large fusiform mass. T1-weighted imaging presented low signal intensity, while T2-weighted image showed heterogeneously high signal intensity. The lesion was completely removed. After pathohistological analysis with standard and immunohistochemical methods of coloring, the diagnosis of the schwannoma was confirmed. **Conclusion.** Schwannoma as a slow-growing tumor, which is often without clinical manifestations, may cause a delay in diagnosis and treatment. Clinical presentation of a schwannoma is indolent and non-specific. Diagnosis of this tumor requires a multidisciplinary and interdisciplinary approach. MR is a useful and highly specific method for the verification of peripheral nerve sheath tumors. A HP analysis confirmed definitive diagnosis of the lesion.

Key words:
schwannoma; abdominal wall; diagnostic techniques and procedures; surgery; histology.

Apstrakt

Uvod. Švanom ili neurilemom je benigni tumor nervnog omotača koji potiče od Švanovih ćelija. Lokalizacija u abdominalnom zidu je retka. Švanomi se obično manifestuju kao sporo rastući asimptomatski tumori. Simptomatologija zavisi od lokacije, zahvaćenog nerva i veličine tumora. **Prikaz bolesnika.** Predstavljamo 43-godišnju bolesnicu sa švanomom lokalizovanim u desnom hipohondrijskom regionu. Dijagnostika je obuhvatala ultrazvuk sa kolor Doplerom (UZ), magnetnu rezonancu (MR) visoke rezolucije i patohistološka ispitivanja (PH). Ultrazvuk je pokazao čvrstu dobro ograničenu masu, ehogene unutrašnjosti u prednjem abdominalnom zidu. Magnetnom rezonancom ova promena je predstavljena kao ovalna, dobro orijentisana, heterogena, fusiformna masa, dijametra 3 × 2.5 × 2.5 centimetara. Na T1 sekvenci promena je imala nizak intenzitet signala, dok je na T2 sekvenci bila heterogeno visokog intenziteta signala. Lezija je u potpunosti uklonjena. Patohistološkom analizom, standardnim i imunohistohemijskim metodama bojenja, postavljena je dijagnoza švanoma. **Zaključak.** Švanom kao spororastući tumor, koji je često bez kliničkih simptoma, može ostati dugo nedijagnostikovan. Klinička prezentacija švanoma je blaga i nespecifična. Dijagnoza ovog tumora zahteva multidisciplinarni i interdisciplinarni pristup. Magnetna rezonanca je korisna i visokospecifična metoda za verifikaciju tumora perifernih nerva. Patohistološka analiza potvrdila je definitivnu dijagnozu lezije.

Ključne reči:
švanom; abdomen, zid; dijagnostičke tehnike i procedure; hirurgija; histologija.

Introduction

Schwannoma or neurilemmoma is a benign, slow-growing, encapsulated tumor of the nerve sheaths, which arises from the Schwann cells of the peripheral, cranial and autonomic nerves. Schwannoma can push the nerve laterally but without infiltrative potential [1]. Most schwannomas are solitary. Multiple schwannomas are usually associated with

Correspondence to: Bojan Nikolić, Military Medical Academy, Institute of Radiology, Crnotravska 17, 11 000 Belgrade, Serbia.
E-mail: bojan_nikolic@vektor.net

neurofibromatosis type 1 (von Recklinghausen disease) [2]. Depending on the location, involved nerve and the size of the tumor, schwannomas can give different symptomatology. Schwannomas usually manifest themselves as slow-growing tumors, which can exist from months to years without clinical manifestation. Malignant schwannomas are very rare and account for approximately 6% of all sarcomas [3–5].

The aim of this case report was to present the patient with the localization of the schwannoma in the abdominal wall. Discussion about diagnosis and therapeutic approach to these tumors will largely contribute to more efficient clinical management.

Case report

A 43-year-old female patient with a localized, painful mass in the abdominal wall of the right hypochondriac region was admitted for evaluation and surgical treatment. Tumor mass slowly enlarged over a period of 2 years, although the patient complained about symptomatology only two months before examination. There was no history evidence of weight loss, fever, anorexia, stress or trauma, and no family history of the similar symptomatology. On physical examination, in the right hypochondriac region there was present a 2–3 cm solid mass, oval-shaped, painful on the palpation and not fixed to the skin of the abdominal wall. Laboratory tests were in the range of normal values. After clinical examination, the first diagnostic procedure was high-resolution ultrasound performed with Linear Array 3–13 MHz on Siemens Acuson Antares ultrasound machine, which showed the solid heterogeneous mass, well circumscribed, with a whorled, echogenic internal architecture in the anterior abdominal wall. Color Doppler sonography showed no appreciable vascularity.

Magnetic resonance imaging (MRI) was performed on machine GE Signa 3.0T, in T2 SS (T2 Single Shot), T2SSFSE (T2 Single Shot Fast Spin Echo), T1 Dual, DWI 50 (Diffusion-Weighted Imaging), DWI 500 (Diffusion-Weighted Imaging), T2FRFSE (Fast Recovery Fast Spin Echo), FRFSE (Fast Recovery Fast Spin Echo) with fat suppression and dynamic T1 (LAVA) Liver acquisition with Volume acquisition images, in all three projections. A mass was revealed under the muscular layers of the right side and lateral at the height of the 6th segment of the liver. The revealed structure was oval, well-circumscribed, heterogeneous, fusiform 3 × 2.5 × 2.5 centimeters in diameter. Mass compressed the liver, without the signs of infiltration. In the surrounding muscle structures there was no sign of edema. After the application of contrast agent, there was a clear demarcation of the capsule. The central part of the lesion also showed intensive but heterogeneous post-contrast opacification.

On T2-weighted images this lesion was peripherally hyperintense and centrally with heterogeneous low and intermediate signal intensity (Figures 1–3). Unenhanced T1-weighted LAVA sequence showed lower signal intensity compared to the signal intensity of the surrounding muscles (Figure 4) and after contrast administration on T1-weighted LAVA sequence, the mass showed intense enhancement (Figure 5).

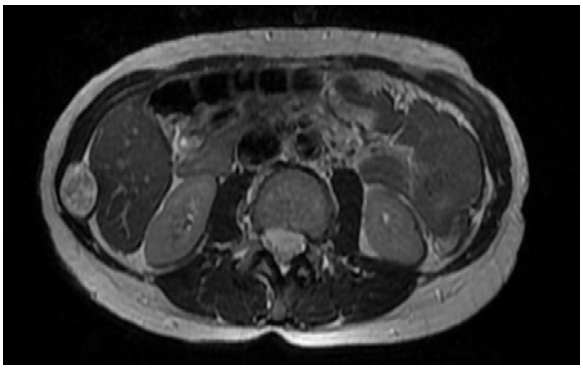

Fig. 1 – T2 axial, clearly demarked incapsulated lesion, peripherally with high signal intensity.

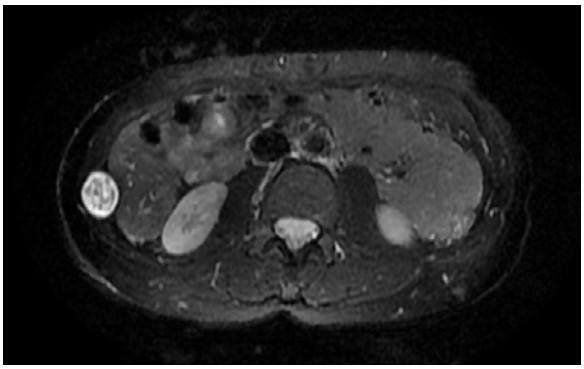

Fig. 2 – T2 fat sat axial lesion, still peripherally hyperintense, probably due to cystic degeneration.

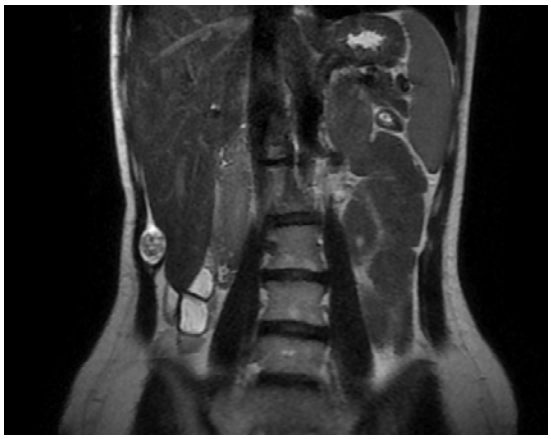

Fig. 3 – T2 core, central part is with "patchy" zones of signal hypointensity.

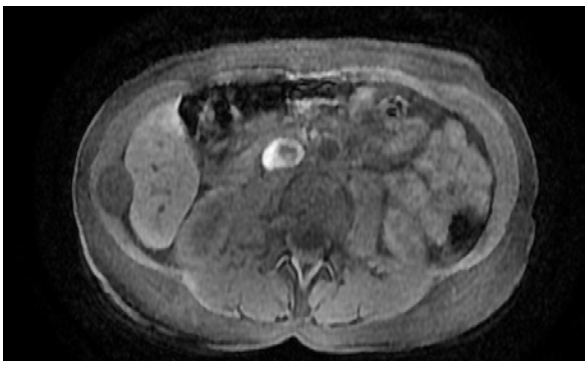

Fig. 4 – T1* LAVA native axial, lesion is hypointense.
*** LAVA – liver acquisition with volume acquisition.**

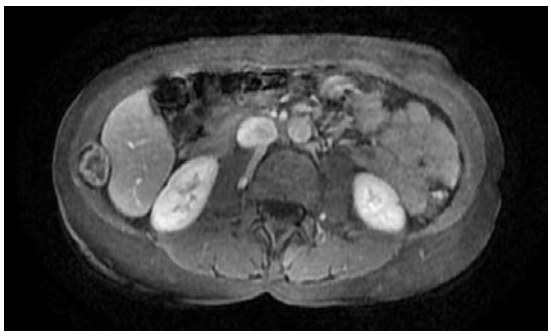

Fig. 5 – T1* LAVA +C axial, there is a smooth rim-like enhancement with central heterogeneous postcontrast enhancement.
*** LAVA – liver acquisition with volume acquisition.**

After all examinations, tumor excision was performed. Abdominal oblique muscles were separated and isolated. The lesion was identified between the right hypochondriac and lumbal region near costal cartilage and intercostal spaces. The lesion was 3 cm large, of ellipsoid shape and solid consistency. It was completely resected together with a capsule and sent for histopathological analysis.

Macroscopically, cystic formation had dimensions 30 × 20 × 10 mm, shiny smooth surfaces, and a narrow pedicel part, dimensions 10 × 1 mm. After cutting, there was a whitish yellow tissue, with partially pseudocystic appearance on the cross-section of the tissue.

Microscopically, the lesion was composed of an area of spindle cells with oval, extruded nuclei and palisading. Histiocytes, mast cells, collagen fibers were also present. Small number of blood vessels had a thickened wall surrounded by a myxomatous extracellular matrix (Figures 6–9).

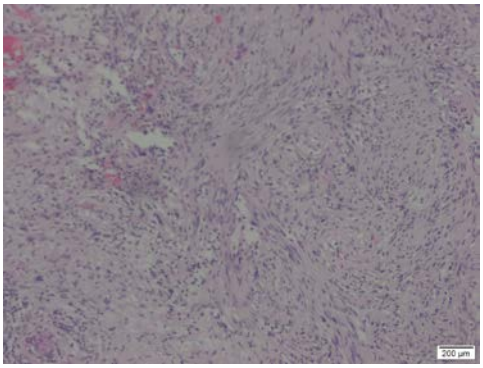

Fig. 6 – Schwannoma (H&E, ×20).

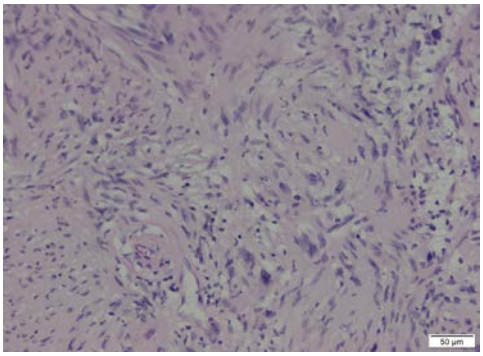

Fig. 7 – Schwannoma (H&E, ×40).

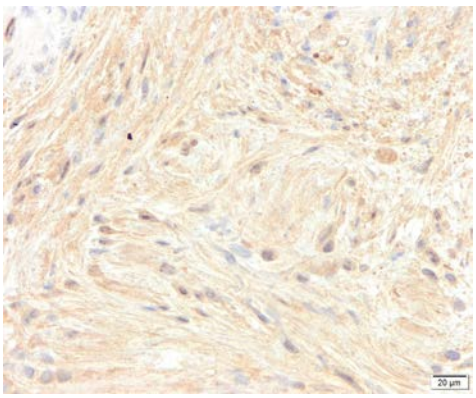

Fig. 8 – Immunohistochemical positivity (S100, ×40).

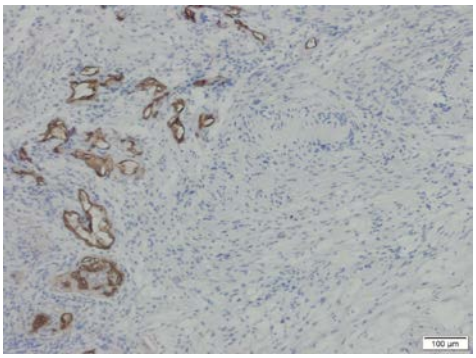

Fig. 9 – Pan Actin positivity only in the blood vessels without immunoreactivity in the Schwann cells (H&E, ×25).

On the histological examination, we found that the tumor cells were positive for S100 protein. The final diagnosis was a schwannoma. The postoperative process was regular.

Discussion

We can conclude the literature review by stating that only few cases of abdominal wall schwannomas were described. Schwannomas are the most common tumors of peripheral nerves with incidence 5% of all benign soft-tissue tumors. Schwannomas affect patients between the ages of 20–50, and with moderate predilection in females. It is commonly associated with neurofibromatosis type 2.

The most common localizations of schwannomas are retroperitoneum (32%), mediastinum (23%), head and neck (18%), and extremities (16%) [5]. There have been reports of their location in the *porta hepatis*, retroperitoneum, pelvis, adrenals, kidneys, vagina and a few in the abdominal wall [4]. Depending on the location, the involved nerve and the size of the tumor, schwannomas can present different symptomatology.

In the majority of cases, schwannomas arise from the nerve sheath of large peripheral nerves, usually asymptomatic and accidentally identified through physical examination or imaging. However, when they grow larger, they usually manifest the symptoms of the compression of the involved nerve [2, 4].

There are two groups of benign peripheral nerve sheath tumors that are usually present as a solitary lesion: schwan-

noma or neurilemmoma and neurofibroma. Schwannomas are encapsulated tumors. They are separated from the nerve with fibrotic capsule, unlike neurofibroma where the nerve is a part of a tumor and must be removed together with the tumor [5, 6]. Ancient schwannomas are a subtype of classic schwannomas and they usually display cellular degenerative changes, including nuclear atypia and pleomorphism, with a tendency to nuclear palisading. Histopathologically, they contain areas of relatively dense cellularity corresponding to Antoni A (AA) regions as well as loose, myxoid Antoni B (AB) regions [7, 8]. Immunohistochemical staining shows that schwannomas strongly react with S100 protein and can be used to differentiate them from malignant peripheral nerve sheath tumors [7–10].

Schvannomas have mostly uniform MR presentation which is T2 high signal intensity with centrally present zones of low and intermediate signal intensity that correspond to central fibrous components and peripheral myxomatous elements seen at pathologic analysis. On spin-echo T1-weighted MR images, the lesion is homogeneous and isointense relative to skeletal muscle [3].

There are certain imaging characteristics that may aid the radiologist in establishing a preoperative diagnosis of a peripheral nerve sheath tumor. These characteristics include association with a peripheral nerve, intermuscular location, and mostly specific MR image [3, 11, 12].

The opposite of schwannomas, malignant peripheral nerve sheath tumors as sarcomas have no specific imaging features, but aggressive biologic behavior may be suggested by indistinct margins, the infiltrative nature of the lesion within the nerve and adjacent structures. In many cases imaging characteristics help identify the neurogenic origin of a mass, but these patterns of signal intensity are neither specific for neural tumors, nor they allow differentiation between benign and malignant nerve sheath tumors. Ideally, at MR imaging we could make difference between a schwannoma and a neurofibroma [3].

The treatment is complete surgical excision, and prognosis is generally good. Incomplete excision can lead to recurrence of the tumor *in situ* or at a distant site after resection [4]. Our patient had good postoperative rehabilitation. We recommended clinical monitoring over a period of one year with shorter intervals between examinations; and during the second year with longer intervals between examinations. It included a review of the surgeon and radiological treatment. During the follow-up there was no evidence of recurrence found on MR images.

Conclusion

Schwannoma as a slow-growing tumor and without clinical manifestations may cause a delay in diagnosis and treatment. Clinical presentation of a schwannoma is indolent and non-specific. Diagnosis of this tumor requires a multidisciplinary approach. MR is a useful method for the verification of peripheral nerve sheath tumors with high sensitivity and specificity. Histopathological analysis confirmed definitive diagnosis of the observed lesion.

REFERENCES

1. *Balzarotti R, Rondelli F, Barizzi J, Cartolari R.* Symptomatic schwannoma of the abdominal wall: A case report and review of the literature. Oncol Lett 2015; 9(3): 1095–8.
2. *Bhatia RK, Banerjea A, Ram M, Lovett BE.* Benign ancient schwannoma of the abdominal wall: an unwanted birthday present. BMC Surg 2010; 10: 1.
3. *Beaman FD, Kransdorf MJ, Menke DM.* Schwannoma: Radiologic-Pathologic Correlation. Radiographics 2004; 24(5): 1477–81.
4. *Liu Y, Chen X, Wang T, Wang Z.* Imaging observations of a schwannoma of low malignant potential in the anterior abdominal wall: A case report. Oncol Lett 2014; 8(3): 1159–62.
5. *Lam R, Hunt B, Owen OA.* Abdominal Wall Schwannoma. Fed Pract 2019; 36(3): 129–33.
6. *Albert P, Patel J, Badawy K, Weissinger W, Brenner M, Bourhill I,* et al. Peripheral Nerve Schwannoma: A Review of Varying Clinical Presentations and Imaging Findings. J Foot Ankle Surg 2017; 56(3): 632–7.
7. *Klijanienko J, Caillaud JM, Lagace R.* Cytohistologic Correlations in Schwannomas (Neurilemmomas), Including "Ancient," Cellular, and Epithelioid Variants. Diagn Cytopathol 2006; 34(8): 517–22.
8. *Mishra A, Hamadto M, Azzabi M, Elfagieh M.* Abdominal wall schwannoma: case report and review of the literature. Case Rep Radiol 2013; 2013: 456863.
9. *Choudhury S, Kumar Das PK, Patro MK.* Ancient schwannoma of neck masquedring as sarcoma. Clin Cancer Investig J 2016; 5(6): 537–9.
10. *Mikami Y, Hidaka T, Akisada T, Takemoto T, Irei I, Manabe T.* Malignant peripheral nerve sheath tumor arising in benign ancient schwannoma: a case report with an immunuhistochemical study. Pathol Int 2000; 50(2): 156–61.
11. *Ginesu GC, Puledda M, Feo CF, Cossu ML, Fancellu A, Addis F,* et al. Abdominal Wall Schwannoma. J Gastrointest Surg 2016; 20(10): 1781–3.
12. *Albert P, Patel J, Badawy K, Weissinger W, Brenner M, Bourhill I,* et al. Peripheral Nerve Schwannoma: A Review of Varying Clinical Presentations and Imaging Findings. J Foot Ankle Surg 2017; 56(3): 632–7.
13. *Crist J, Hodge JR, Frick M, Leung FP, Hsu E, Gi MT,* et al. Magnetic Resonance Imaging Appearance of Schwannomas from Head to Toe: A Pictorial Review. J Clin Imaging Sci 2017; 7: 38.

Received on December 10, 2018
Revised on February 1, 2019
Accepted February 21, 2019
Online First March, 2019