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SCHWANNOMA OF THE ABDOMINAL WALL - DIAGNOSTIC CHALLENGE

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Abstract

Introduction: Schwannoma or neurilemmoma is benign tumour of the nerve sheath originating from the Schwann cells. Localization in abdominal wall is rare. Schwannomas are usually manifested as slow-growing tumour and can exist for months to years without producing symptoms. Symptomatology depends of location, involved nerve and the size of the tumour.

Case report: We present a 43-year-old female patient with Schwannoma localized in the right hypochondriac region. Diagnostic procedures included high-resolution ultra sound with color Doppler (US), pathohistological examination (PH) and magnetic resonance imaging (MR). High-resolution ultra sound showed the solid mass, well circumscribed, with a whorled, echogenic internal architecture in the anterior abdominal wall. MR imaging revealed oval, well-circumscribed, heterogeneous, fusiform 3 x 2.5 x 2.5 centimeters large mass. T1-weighted imaging presented low signal intensity and heterogeneously high signal intensity on T2-weighted image. The lesion was completely surgically removed. After histopathological examination with immunobiochemistry, the diagnose of Schwannoma was confirmed.

Conclusion: Schwannoma as slow-growing tumours and of without clinical manifestation may cause a delay in diagnosis and treatment. Clinical presentation of Schwanoma is indolent and non-specific. Diagnosis of this tumor requires multidisciplinaire approach. MR is a useful method for verification peripheral nerve sheath tumors with high sensitivity and specificity. Histopathology analysis confirmed definitive diagnose of observed lesion.

Key words: Schwannoma, Abdominal wall, Abdominal pain, Magnetic resonance imaging, S100.

Apstrakt

Uvod: Švanom ili neurilemom je benigni tumor nervnog omotača koji potiče od Švanovih čelija. 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 slučaja:** Predstavljamo 43-godišnju pacijentkinju sa Švanomom lokalizovanim u desnom hipohondrijskom regionu. Dijagnostika je obuhvatala ultrazvuk sa kolor Doplerom (US), 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 x 2.5 x 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.


**Ključne reči:** Švanom, abdominalni zid, bol u stomaku, magnetna rezonanca, S100.

**Introduction**

Schwannoma or neurilemmoma is benign, slow-growing, encapsulated tumour of the nerve sheaths, that 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 neurofibromatosis 1 (von Recklinghausen disease). (2) Depending on the location, involved nerve and the size of the tumour, Schwannomas can give different symptomatology. Schwannomas are usually manifested as slow-growing tumour which can exist for months to years without clinical manifestation. Malignant Schwannomas are very rare and account for approximately 6% of all sarcomas. (3)
Aim of this study was to present the patient with localization of schwannoma in abdominal wall. Discussion about diagnosis and therapeutic approach of these tumors will largely contribute to more efficient clinical management.

Case report
A 43-year-old female patient with localized, painful mass in the abdominal wall of the right hypochondriac region was admitted for evaluation and surgical treatment. Tumor mass had slowly enlarged over a period of 2 years, although patient complained about symptomatology 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 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, 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.

MR imaging was made on apparatus 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 aquisition) 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 liver. The revealed structure was oval, well-circumscribed, heterogeneous, fusiform 3 x 2.5 x 2.5 centimeters in large. It compressed the liver, without signs of infiltration. In the surrounded muscle structures was no signs of oedema. After application of contrast agent, there is clear demarcation of the capsule. Central part of the lesion also shows intensive but heterogeneous post contrast opacification.

On T2-weighted images this lesion is peripherally hyperintense and centrally is with heterogeneous low and intermediate signal intensity (Figure 1, 2, 3). Unenhanced T1-weighted LAVA sequence shows lower signal intensity comparing to the signal intensity of
surrounding muscles (Figure 4) and after contrast administration on T1-weighted LAVA sequence, the mass shows intense enhancement (Figure 5).

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

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

Fig 3. T2 cor, central part is with “patchy” zones of signal hypointensity.
Fig 4. T1 LAVA native axial, lesion is hypointense.

Fig 5. T1 LAVA +C axial, there is smooth rim-like enhancement with central heterogeneous postcontrast enhancement.

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

Macroscopically, cystic formation dimension 30x20x10 mm, shiny smoothly surfaces, with a narrow pedicel part, dimension 10x1 mm. After cutting there was a whitish yellow tissue, partially pseudocystic appearance on the cross-section of the tissue.

Microscopically, the lesion was consisted of an area composed 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 mixomatous extracellular matrix (Figure 6, 7, 8, 9).
On the histological examination we found that the tumour cells were positive for S100 protein. The final diagnosis was Schwannoma. The postoperative process took place in well order.

**Discussion:**
We can conclude the literature review, to date, only few cases of abdominal wall Schwannomas were described. Schwannomas are the most common tumours of peripheral nerves with incidence 5% of all benign soft-tissue tumours. Schwannoma affects patients between the ages of 20-50 years old and with moderate predilection in females. It is commonly associated with Neurofibromatosis Type 2. (1, 2, 3)

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, involved nerve and the size of the tumour, 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, usually manifest symptoms of compression the involved nerve. (2, 4)

There are two groups of benign peripheral nerve sheaths tumors that usually are present as solitary lesion: Schwannoma or neurilemoma and neurofibroma. Shwannomas are encapsulated tumors. They are separated from nerve with fibrotic capsule, unlike neurofibroma where the nerve is the part of the tumour and must be removed together with the tumor. (5, 11) Ancient Shwannomas are a subtype of classic Schwannomas usually displays cellular degenerative changes, including nuclear atypia and pleomorphism, with a tendency to nuclear palisading. Pathohistologaly they contained areas of relatively dense cellularity corresponding to Antoni A (AA) regions as well as loose, myxoid Antoni B (AB) regions. (6, 7) Immunohistochemical staining show that Schwannomas strongly react with S100 protein and can be used to differentiate them from malignant peripheral nerve sheath tumors. (6, 8, 9)

Shwannoma 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, 10, 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 do they allow differentiation between benign and malignant nerve sheath tumors. Ideally, at MR imaging we could make differ between Schwannoma and neurofibroma. (3)

The treatment are complete surgical excision, and prognosis is generally good. Incomplete excision can lead to recurrence of the tumour 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.

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