Retroperitoneal ancient schwannoma – case presentation

Cristian Mesină1), Stelian Ștefaniță Mogoantă1), Daniel Alin Cristian2), Theodor Viorel Dumitrescu1), Petru Octavian Drăgoescu3), Mihaela-Justina Mesină-Botoran4), Marius Eugen Ciurea3), Mirela-Corina Ghiuș5), Daniela Cioabanu7)

1) Department of Surgery, Emergency County Hospital, Craiova, Romania; University of Medicine and Pharmacy of Craiova, Romania
2) Department of Surgery, “Colțea” Clinical Hospital, Bucharest, Romania; “Carol Davila” University of Medicine and Pharmacy, Bucharest, Romania
3) Department of Urology, Emergency County Hospital, Craiova, Romania; University of Medicine and Pharmacy of Craiova, Romania
4) Department of Human Anatomy, University of Medicine and Pharmacy of Craiova, Romania
5) Department of Plastic Surgery and Reconstructive Surgery, Emergency County Hospital, Craiova, Romania; University of Medicine and Pharmacy of Craiova, Romania
6) Laboratory of Pathology, Emergency County Hospital, Craiova, Romania; University of Medicine and Pharmacy of Craiova, Romania
7) Department of Gastroenterology, Emergency County Hospital, Craiova, Romania; University of Medicine and Pharmacy of Craiova, Romania

Abstract

Retroperitoneal ancient schwannomas are rare tumors, more usually found in the head, neck and flexor surfaces of the extremities. Ancient schwannomas are a subtype of classic schwannomas with a predominance of degenerative changes, calcifications, hemosiderin deposition, interstitial fibrosis and vascular hyaline degeneration. A 33-year-old male was referred on our hospital with a painful mass in left iliac fossa. The patient underwent surgery and intra-operatively the cystic encapsulated mass was found to be retroperitoneal, between the left psoas major muscle and left iliac muscle. On microscopic examination, we found the presence of Schwann cells in regions with high and low cellularity (Antoni A and B areas) and S100 protein immunohistochemical examination was intensely positive, being consistent with the diagnosis of schwannoma. Complete excision is the only method of the surgical treatment; schwannomas are not sensitive to radiotherapy and chemotherapy. Some authors consider that a complete excision of the tumor, while others believe that enucleated or partial excision of the tumor is sufficient. The prognosis is good, and the most common complication is recurrence, possibly by incomplete excision of it being reported in 5–10% of cases. In conclusion, retroperitoneal schwannomas is usually identified incidentally on tomographic images. Diagnosis is based on histopathological examination after surgery and immunohistochemical examination.

Keywords: retroperitoneal ancient schwannoma, Schwann cells, Antoni A and B areas.

Introduction

Schwannomas are rare variant of peripheral nerve sheath tumors and usually occur as solitary encapsulated mass with demarcated margins with neighbor organs. Retroperitoneal schwannoma are rare tumors comprising only 0.5% to 12% of all retroperitoneal tumors [1]. In the absence of von Recklinghausen’s disease, schwannomas rarely occur in the retroperitoneal space comprising 0.5% to 5% of all schwannomas [2, 3]. Retroperitoneal schwannomas are usually found in the presacral region or paravertebral space [4, 5]. Schwannomas infrequently undergo malignant transformation, this tumors being usually benign [4]. In this article, we present a case of retroperitoneal ancient schwannoma and review the literature on this problem.

Objective

We describe a 33-year-old man with retroperitoneal ancient schwannoma. Our patient is unique in its presentation of retroperitoneal ancient schwannoma discovered on the left genito-femoral nerve path and completely excised from paravertebral region by neurolysis of the left genito-femoral nerve sheath.

Case report

B.M., a 33-year-old male patient was admitted in the 2nd Surgical Clinic, Emergency County Hospital, Craiova, Romania, in March 2014, with paresthesia on the left thigh, left iliac fossa pain. Physical examination showed non-tender mass, measuring 4/5 cm in left iliac fossa slightly sensitive to palpation, regular surface. Patellar reflex, Achilles reflex and cutaneous reflexes were present bilaterally. Laboratory investigations showed: hemoglobin 15.4 g/dL, white blood cell count 6000/mm³, platelets 168 000/mm³, urea 43 mg%, creatinine 0.72 mg%, SGOT (serum glutamic oxalacetic transaminase) 13 IU/L, ALAT (alanine aminotransferase) 12 IU/L, glycemia 94 mg%. A chest-X-ray was normal. The ultrasound (US) examination revealed: in the left iliac fossa a solid tumor upon the iliac crest, with 5 cm diameter, homogenous and regular shaped, approximately 10 mm away from the skin plan. The tumor was poorly vascularized.
Computed tomography (CT) examination of the abdomen and pelvis performed native and post-contrast reveals in the left iliac fossa a tumor mass of 5.2–4.6 cm, which is round-oval, well-defined but is disappearing in some CT sequences. The tumor is tangent to the psoas muscle but has a cleavage space (Figure 1) towards it and shows a small extension not exceeding inguino-femoral region.

**Figure 1** – A CT image showing a heterogeneous cystic mass, which is localized between of the left psoas muscle and the left iliac muscle, beginning from the left L4–L5 intervertebral foramina.

Based on these, the patient underwent surgery. Intraoperatively, the cystic encapsulated mass measuring 5/6 cm was found to be retroperitoneal in location (Figure 2), between the left psoas major muscle and left iliac muscle, over the left front of the psoas muscle. The tumor was discovered on the left genito-femoral nerve path (Figure 3). The mass was dissected from all adjacent tissue and completely excised from paravertebral region by neurolysis of the left genito-femoral nerve sheath.

**Figure 2** – Intra-operative imagine of the retroperitoneal cystic encapsulated mass.

**Figure 3** – The mass was completely excised by neurolysis of the left genito-femoral nerve sheath.

Tumor gross examination reveals a solitary encapsulated mass of 56/42 mm diameter. It has roseate appearance, hemorrhage areas and areas of cystic degeneration on tumor section (Figure 4).

**Figure 4** – Macroscopic aspect of the encapsulated mass measuring 56/42 mm with hemorrhagic and cystic areas on the section.

Surgical specimen was fixed in 10% formalin solution for three weeks. After fixation, thick sections of 1 cm from different lesions parts of postoperative specimen were performed. These sections were included in 12 paraffin blocks, which were therefore examined using standard histological techniques. Five μm sections were stained with Hematoxylin–Eosin (HE). Microscopic examination was performed using Nikon microscope.

Immunohistochemical examination was performed on paraffin blocks containing tumor formation eradicated of the retroperitoneal space. It was used the standard immunohistochemical technique. On microscopy, the tumor was composed of Schwann cells with regions of high and low cellularity (Antoni A and B areas, respectively). Immunohistochemistry showed that spindle cells were strongly positive for S100 protein and final diagnosis of benign ancient schwannoma was made.

The postoperative course was uneventful. During the follow-up, there was no evidence of recurrence.

Gross examination of the resected tumor showed a solitary tumor, encapsulated with 56/42 mm diameter. Macroscopically, hemorrhagic areas and areas of cystic degeneration in the tumor mass were observed.

From the microscopic point of view, the tumor was delimited by fibrous capsule and was composed of hypocellular and hypercellular areas. They show spindle cells with focal nuclear palisading patterns arranged in distinctive dense (Antoni A) and loose (Antoni B) areas (Figure 5). Antoni B areas overcome Antoni B areas (Figure 5). Antoni A areas were composed of fusiform cells arranged in short bundles or network with imprecise cytoplasm and nuclei twist boundaries (Figure 6). On well-differentiated Antoni A areas, we were able to observe focal nuclear palisading patterns and connective tissue fragments (Verocay bodies) and intranuclear vacuoles (consisting of twp compact rows of aligned nuclei separated by fibrillar cell processes) (Figure 7). Antoni B areas (hypocellular areas) were edematous and myxoid containing chronic inflammatory cells and spindle cells, which were haphazardly arranged, aggregating in areas to form small focal palisades, thin collagen fibers, microcystic spaces and rich vascularity (Figure 8).

The spindle cells showed hyperchromatic and irregular nuclei although no mitotic figures. There were several foci of stromal hyalinization and sclerosis with numerous ectatic blood vessels. Vascularization was made up of thin-walled vessels and vessels with thickened walls in a dense fibrosis, with a uniform distribution or with the issue angiomatoid forming area (Figure 9). There were observed large vessels,
irregular spaced tortuous lumen empty or filled with thrombi in various stages of organization (Figure 10).

Emphasized hemorrhage was found to be the primary cause (caused by rupture of intratumoral vessels) and no secondary cause (surgical trauma), hemorrhagic areas near being observed siderophage or hemosiderin pigment (Figure 11).

The presence of inflammatory cells (lymphocytes, mast cells, macrophages) with the diffuse or focal was observed in tumor stroma (Figures 12 and 13).

Also, were observed the areas of cystic degeneration, simple cysts were present (without distinction like glandular, the lack organization of the wall and maintain the characteristics of the surrounding tissue) (Figures 14 and 15).

Immunohistochemical profile of the lesion revealed an intense and diffuse positive immunoreactivity for antibodies anti-vimentin (Figure 16) and anti-S100 protein (Figure 17) and negative immunoreactivity for α-smooth muscle actin (α-SMA) antibodies (Figure 18). Ki67 proliferative index was reduced in the nuclei of tumor cells (Figure 19).
Figure 11 – Schwannomas, with the presence of siderophages and thrombosed vessel. HE staining, ×100.

Figure 12 – Schwannomas, inflammatory cells (lymphocytes, xanthomatous deposits). HE staining, ×200.

Figure 13 – Schwannoma, moderate inflammatory infiltrate. HE staining, ×200.

Figure 14 – Schwannoma with cystic spaces and chronic inflammatory reaction, rich xanthomatous deposits. HE staining, ×100.

Figure 15 – Schwannoma with cystic spaces lined by rounded Schwann cells. HE staining, ×100.

Figure 16 – Schwannoma: vimentin positive immunostaining in tumor cells. Anti-vimentin immunostaining, ×200.
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Discussion

Retroperitoneal schwannomas are rarely encountered, especially to the young adults; incidence is two times higher in women. These are benign tumors arise from Schwann cells of the peripheral nerve sheath [6]. Only a few cases of retroperitoneal schwannomas have been reported in the literature. The common sites of occurrence include head, neck, flexor surfaces of extremities, posterior mediastinum [7]. Rare sites of occurrence have been reported such as: retroperitoneal space, pancreas, small bowel, extrahepatic biliary tree, pelvis and sacrum [8]. Retroperitoneal schwannomas may be asymptomatic and incidentally discovered during medical examination or imagistic techniques. However, occasionally they produce pressure effects on surrounding large nerves [9]. Neurological symptoms are rare. In our case, the patient had paresthesia on the left thigh, left iliac fossa pain. A few cases also present with abdominal complaints or lower back pain [10]. Many theories attempt to explain the degeneration cystic schwannoma. One theory involves the degeneration of Antoni B areas lead to cyst formation, while progressing in size; another theory holds that with increasing tumor central ischemic necrosis occurs that causes cysts within the tumor [10].

Ancient schwannoma is a rare subtype of typical schwannoma with histological predominance of degenerative findings [11]. This type of tumors presents spindle cells arranged in distinctive dense (Antoni A) and loose (Antoni B) areas [11, 12]. In our case, the diagnosis was of benign retroperitoneal ancient schwannoma.

On gross-appearance, schwannomas are encapsulated, solitary and well-circumscribed tumors [4]. On microscopy, schwannomas are composed of two components: a highly ordered cellular component (Antoni A area) and a looser myxoid component (Antoni B area). The encapsulated and the presence of these two components distinguish schwannoma from neurofibroma [13–15]. Retroperitoneal schwannoma show cystic degeneration in up to 60% of cases, while calcification is seen in 23% of cases [16]. In absence of signs, diagnosing of retroperitoneal schwannoma is quite difficult on cross-sectional imaging alone. CT and ultrasonography should be used to obtain tissue specimen (biopsy) and then morphopathology should identify the aggressive features of malignant schwannoma [8, 17, 18].

Although fascicular target and fascicular signs are characteristic radiological features of schwannoma, these are not usually seen in retroperitoneal schwannomas. The target sign is seen on T2-weighted CT image as a hypointense signal in centre and hyper-intense signal in the periphery. This is caused by fibrous components in the center surrounded by myxomatous elements in periphery [19, 20].

Ultrasonography and CT are helpful in differentiating retroperitoneal tumors. In our case, a CT image showed a heterogeneous cystic mass, which was localized between of the of left psoas muscle and of the left iliac muscle, beginning from left L4–L5 intervertebral foramina. Magnetic resonance imagines are a useful alternative and allow a better view of the origin of tumor vascular architecture and the relations with other organs. Complete surgical excision is the only treatment method; schwannomas are not sensitive to radiotherapy and chemotherapy [21].

The need for negative tissue margins of resection piece is controversial especially when adjacent viscera and neighborhood tissues are excised [22]. Some authors consider that a complete excision of the tumor, while
others believe that enucleated or partial excision of the tumor is sufficient [4]. The prognosis is good, and the most common complication is recurrence, possibly by incomplete excision of it being reported in 5–10% of cases [5].

Conclusions

Retroperitoneal schwannomas are usually identified incidentally on tomographic images. Diagnosis is based on histopathological examination after surgery and immunohistochemical examination. Total excision of the tumor had the best prognosis.

Conflict of interests

The authors declare that they have no conflict of interests.

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References


Corresponding author

Cristian Meşină, Lecturer, MD, PhD, Department of Surgery, University of Medicine and Pharmacy of Craiova, 2 Petru Rareş Street, 200349 Craiova, Romania; Phone +40724–513 221, e-mail: mesina.cristian@doctor.com

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