

Case Report

## A case of a cellular schwannoma variant arisen in an intrapelvic space

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### Abstract

Cellular schwannoma is rare neoplasma and often situates in deep structures, so that its diagnosis is quite difficult. We herein report a case of a cellular schwannoma variant arisen in an intrapelvic space. A 62 year-old female had a magnetic resource imaging (MRI) examination of the lumber, which indicated a mass in the intrapelvic space behind the rectum. In T1-weighted image, the mass was slight low in intensity with cystic lesions being low. In T2-weighted image, the majority of the mass was low in intensity, whereas diffuse cystic lesions within it high. In computed tomography (CT), the tumor was well-demarcated round mass with heterogeneous low and intermediate attenuation area and punctual calcification. Because the mass was highly suspected as a pelvic mesenchymal neoplasm such as gastrointestinal stromal tumor or sarcoma, operation was performed. At laparotomy, the tumor had a thin capsule without invasion to the rectum or the mesorectum, and was arisen from the left hypogastric nerve. We cut off the left hypogastric nerve to extirpate the tumor. Macroscopically, it was a solid tumor and the color of the cutting surface was yellow-white with cysts. Pathologically, the tumor consisted of compact spindle cells arranged in short bundles or interlacing fascicles with high density. Nuclear palisading and Verocay body formation that is seen in the typical schwannomas were not spotted. Hyalinization, hemorrhage, xanthic change and lipofuscin precipitation were detected. Mitosis and necrosis were not present. Immunohistochemically, S-100  $\beta$  and glial fibrillary acidic protein (GFAP) was positive in tumor cells. Smooth muscle actin (SMA), CD34, CD117 and neurofilament were negative. MIB-1 index was less than 5%. Final diagnosis was obtained as a cellular schwannoma variant.

**Key Words:** Cellular schwannoma, Antoni A and B.

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### Introduction

Cellular schwannoma is rare neoplasm, accounting for 5-10% of all schwannomas<sup>1-2)</sup>. Cellular schwannoma often situates in deep structures such as the mediastinum, retroperitoneum and intrapelvis, and frequently arises from major nerve. Diagnosis of cellular shwannoma is quite difficult because of the hypercellularity and deep location. We herein report a case of a cellular schwannoma variant arisen in an intrapelvic space.

### Case

A 62 year-old female patient had been followed up by orthopedics for left thigh and left bottom pains for 4 months without any improvement. She had a magnetic resource imaging (MRI) examination of the lumber, which indicated a mass in the intrapelvic space behind the rectum. She had no past history of an abdominal operation. Laboratory examinations, however, identified no

abnormalities in hematological, chemical data or tumor markers. Additionally, plain abdominal radiographs were normal. Computed tomography (CT) depicted a solid mass of 74×71×66 mm in size, located in the presacral midline posterior of the rectum. The tumor was well-demarcated round mass with heterogeneous low and intermediate attenuation area and punctual calcification. The intermediate area showed similar density to that of skeletal muscle with slight enhancement. Neither enlarged lymph nodes nor fluid were seen (Fig. 1). On MRI ex-



Fig. 1 In CT scan, solid mass of 74×71×66 mm in size, is observed in the presacral midline posterior of the rectum.

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amination, a mass with smooth margin of 70×62 mm in size was seen in the presacral. In T1-weighted image, the mass was slight low in intensity with cystic lesions being low. In T2-weighted image, the majority of the mass was low in intensity, whereas diffuse cystic lesions within it high (Fig. 2). A thin hypointense rim was recognized as capsule on T1- and T2-weighted images. Although colonoscopy and barium enema were performed for more information, no mucosal irregularity was found without exclusion of the wall. Because the mass was highly suspected as a pelvic mesenchymal neoplasm such as gastrointestinal stromal tumor or sarcoma, operation was performed.

At laparotomy, the tumor was located at the posterior

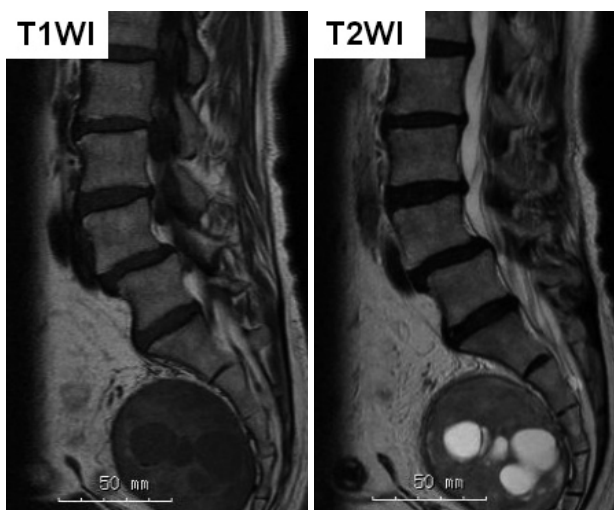


Fig. 2 MRI shows a mass with smooth margin of 70×62 mm in size in the presacral. In T1-weighted image, the mass was slight low in intensity with cystic lesions being low. In T2, the majority of the mass shows low intensity, whereas diffuse cystic lesions within it high.



Fig. 3. Solid tumor is 8.5×7.5×6 cm in size, and the color of the cutting surface is yellow-white with cysts.

region of the rectum. The tumor had a thin capsule without invasion to the rectum or the mesorectum. The tumor was excised from the mesorectum and the sacral. Finally, the tumor was adhered to left sacral because a part of the left hypogastric nerve was embedded in the tumor. Therefore, we cut off the left hypogastric nerve to extirpate the tumor.

Macroscopically, it was a solid tumor 8.5×7.5×6 cm in size, and the color of the cutting surface was yellow-white with cysts (Fig. 3). Pathologically, the tumor consisted of compact spindle cells arranged in short bundles or interlacing fascicles with high density. Nuclear palisading and Verocay body formation were not spotted. Hyalinization, hemorrhage, xanthic change and lipofuscin precipitation were detected. Mitosis and necrosis were not present (Fig. 4). Immunohistochemically, S-100  $\beta$  and glial fibrillary acidic protein (GFAP) were positive in tumor cells (Figs. 5A, B) and CD68 was positive

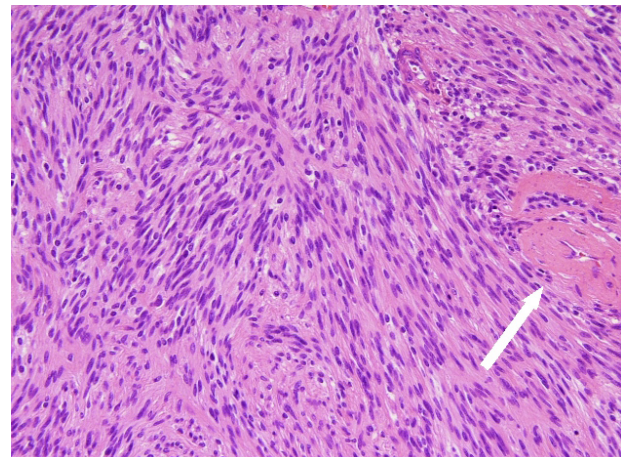


Fig. 4. In HE stain, spindle cells arranged in short bundles or interlacing fascicles with high density. Hyalinization (white arrow) is detected.

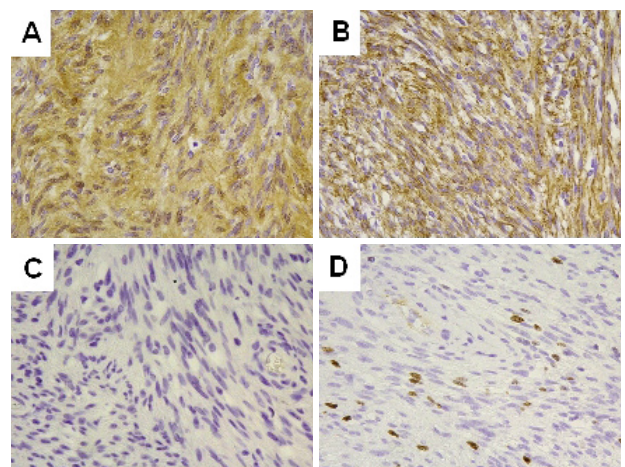


Fig. 5. S-100  $\beta$  (A) and GFAP (B) were positive in tumor cells, whereas SMA (C) was negative. MIB-1 index was less than 5% (D).

in histiocytes. In stromal tissue, Type IV collagen was positive and Leu7 was partially. Smooth muscle actin (SMA) (Fig. 5C), CD34, CD117 and neurofilament were negative. MIB-1 index was less than 5% (Fig. 5D). Final diagnosis was obtained as a cellular schwannoma variant.

## Discussion

Schwannoma, or neurilemoma is a benign neoplasms of schwann cells, which occur at all ages but are most common in females between ages of 20 and 50 years<sup>2)</sup>. It is classified in six types as usual type, cellular, plexiform, antient, epithelioid and neuro-blastoma like<sup>3)</sup>. Schwannomas arise as solitary masses of superficial or deep soft tissues, often association with peripheral nerves of the four legs, trunk, head and neck, cranial nerve roots, cervical nerves, dorsal spinal nerve roots, posterior mediastinum and retroperitoneal regions<sup>2)</sup>. It is a slowly growing tumor that is usually present several years before diagnosis. When the tumor grows and becomes large, symptoms such as sensory disturbance and pain occurs. On image findings, the tumor is quite homogenous, and degeneration can be seen as calcification or cystic lesions.

In a schwannoma, pathologically it consisted of two components: Antoni A and B. Antoni A areas are composed compact spindle cells. They are arranged in short bundles or interlacing fascicles. In highly differentiated Antoni A areas, there may be nuclear palisading and Verocay bodies. Antoni B areas are far less orderly and less cellular<sup>4)</sup>. Immunohistochemically, S-100 protein is strongly expressed by most cells and GFAP is occasionally positive in a schwannoma but not in a neurofibroma. Additionally, it is not immunoreactive for SMA which is positive in smooth muscle-related tumors. So, the immunostaining pattern of our case supports the diagnosis of schwannoma. The relative amounts of these two components vary, and they may blend imperceptibly or change abruptly.

Cellular schwannoma first reported by Woodruff et al<sup>5)</sup> is an uncommon variant of schwannoma, characterized by almost entirely Antoni A pattern growth and the absence of Verocay bodies. In this case, there are almost entirely Antoni A pattern growth and the absence of Verocay bodies, and small amount of Antoni B area represented by the degenerative changes including hya-

linization, hemorrhage and cystic change<sup>2)</sup>.

Although cellular schwannoma is well-recognized, it is important to make sure preoperative diagnosis, because the differential diagnosis is often including malignant tumors such as fusiform sarcoma, leiomyosarcoma and malignant peripheral nerve sheath tumor. Because of its cellularity, mitotic activity and occasional presence of bone destruction, it is diagnosed as malignant in more than one-fourth of cases<sup>2)</sup>. However, malignancy was excluded in our case, because MIB-1 index was less than 5%.

On imaging findings, typical intra-pelvic schwannomas show well defined round, oval or lobulated masses. Capsule formations can occasionally be seen as hypointense rims on T1- and T2-weighted images<sup>6)</sup>. Degeneration changes such as calcification, cystic lesions and hemorrhage can be seen<sup>7)</sup>. In this case, although we found special features such as defined round mass, cystic lesions, hemorrhage and probably capsule formation, still it was hard to get the accurate diagnosis because of common findings which are the part of soft tissue tumors' character. Additionally, when the relative amounts of Antoni A and B areas vary they show the different imaging findings, which make the preoperative diagnosis more difficult. So, we need to pay attention to recognize the variety of differential diagnosis of soft tissue tumors with these findings.

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