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Schwannoma Arising in a Lymph Node
– A Brief Case Report –

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Intranodal schwannomas are extremely rare and only three cases have currently been reported in the English language literature. We report here on a case of a schwannoma that arose in a retroperitoneal lymph node. A 59-year-old male patient had experienced abdominal discomfort for two months. An abdominal CT scan demonstrated a heterogeneous density mass in the retroperitoneum. Histological examination of the mass identified it as a lymph node due to the presence of a peripheral rim of compressed lymphoid tissue that contained a well-demarcated benign spindle cell tumor in its center. The spindle cells were positive for S-100 protein, and they were negative for smooth muscle actin, desmin, and CD 34. Although an intranodal schwannoma is histologically benign, it is important to distinguish this lesion from an intranodal metastasis of a spindle cell tumor and other common benign spindle cell tumors that can arise in a lymph node.

Key Words : Schwannoma; Neurilemmoma; Lymph node

Among the intranodal spindle cell tumors, a primary schwannoma arising in a lymph node is extremely rare.[1-3] Although an intranodal schwannoma is benign, this lesion in a lymph node may be initially alarming and it may suggest a diagnosis of intranodal metastasis from a spindle cell tumor. However, as an intranodal schwannoma can mimic all of the morphological features of schwannomas arising in soft tissue, based on the particular histological and immunohistochemical features, the danger of overdiagnosis can be avoided. We report here on a rare case of a schwannoma that arose in a retroperitoneal lymph node, which is a clinically unexpected finding, and we briefly review the clinical literature on intranodal spindle cell tumors. To the best of our knowledge, this is the first Korean report of a patient with an intranodal schwannoma.

CASE REPORT

A 59-year-old-man was admitted to our hospital because of epigastric and abdominal discomfort he had suffered with for the previous two months. An abdominal CT scan demonstrated a heterogeneous density mass that was located in the retroperitoneum (Fig. 1A). The levels of urinary cortisol, aldosterone, vanillylmandelic acid (VMA), metanephrine, and 17-ketosteroids (17-KS) were within the normal limits, along with the levels of serum cortisol and aldosterone. The tentative preoperative diagnosis was a non-functioning extradrenal pheochromocytoma. We performed a laparoscopic mass excision. On the gross examination, the mass measured 3.6 × 3.2 cm, and it was seen as a well circumscribed, firm, and pale gray nodule surrounded by a thick fibrous capsule. Microscopically, the mass contained lymphoid tissue that was comprised of follicles with germinal centers and, paracortical tissue, and there was a subcapsular sinus in the peripheral area (Fig. 1B). The center of the mass contained proliferating spindle cells. The spindle cells were dispersed in short interlacing fascicles or they were haphazardly arranged in a loose collagenous matrix. Vague nuclear palisading reminiscent of an Antoni A area, was observed, but no well-formed Verocay bodies were identified (Fig. 1C). The Antoni B areas are far less orderly and less cellular (Fig. 1D). The majority of the spindle cells displayed elongated and slightly wavy nuclei with an ill-defined cytoplasmic border. Atypical cells and mitotic activity were not

observed. The spindle cells showed the diffuse expression of vimentin and S-100 protein, based on the use of immunohistochemical staining (Fig. 2A). However, the spindle cells were negative for smooth muscle actin, desmin, cytokeratin, and CD34

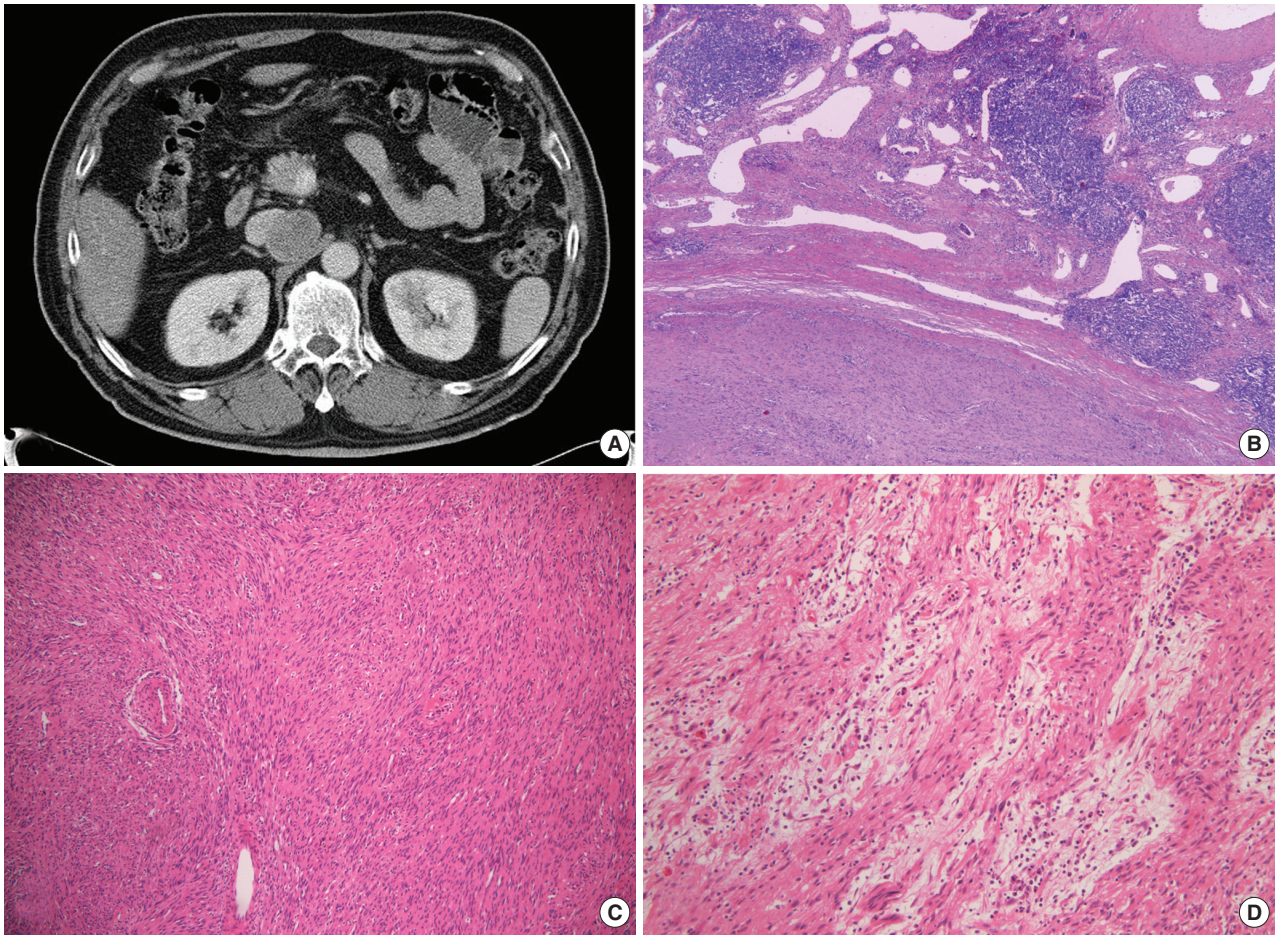

Fig. 1. (A) Computed tomography of the abdomen shows presence of a 3.6×3.2 cm sized, heterogeneous density mass with minimal contrast enhancement. (B) Low-power microscopic finding of the mass shows a peripheral rim of lymphoid tissue with subcapsular sinus surrounding a central well-circumscribed tumor. (C) Spindle cell tumor is arranged in interlacing fascicles mimicking Antoni A areas with focal nuclear palisading. (D) There is a transition between Antoni A areas and loosely textured Antoni B areas (center).

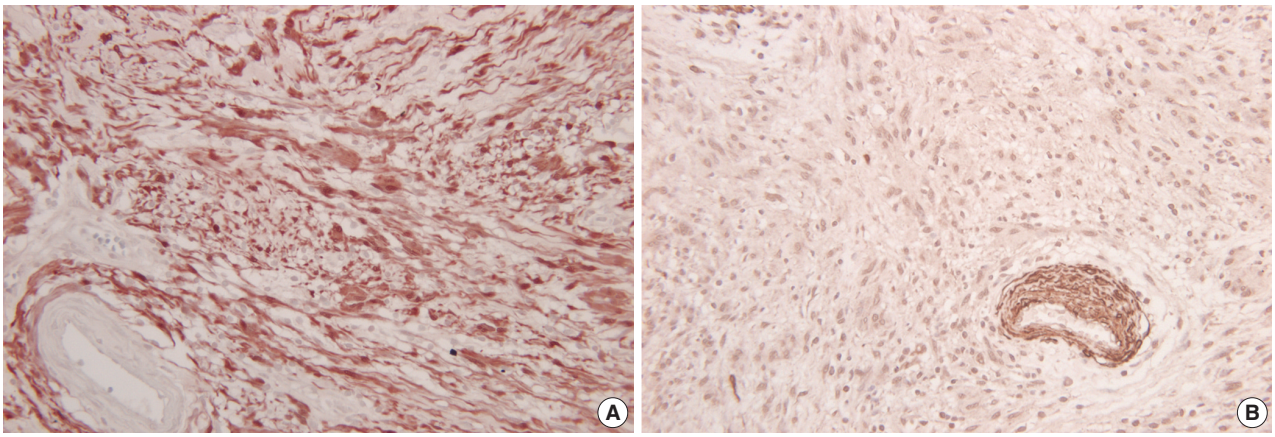

Fig. 2. The spindle cells are strongly immunoreactive to S-100 protein (A) and negative to smooth muscle actin (B).

(Fig. 2B). These findings were consistent with a diagnosis of a primary schwannoma of the lymph node.

DISCUSSION

Schwannomas arising in lymph nodes have been described by Enzinger and Weiss[4] as extremely rare benign lesions, and only three such cases have currently been reported.[1-3] In all of the previously reported cases and also in the present case, a peripheral rim of compressed lymphoid tissue surrounding a central tumor with the typical histological features of a schwannoma in soft tissue was seen. However, identification can be difficult for all cases of primary soft tissue tumors where a prominent rim of lymphoid follicles surrounds the lesion, as these lesions can simulate the appearance of a lymph node. Given that a subcapsular sinus is present in these types of tumors, and chronic inflammation is not usually associated with benign neural tumors, the diagnosis of a true intranodal lesion may be favored.[1,5]

Intranodal schwannomas must be differentiated from other benign and metastatic spindle cell tumors. Among the benign spindle cell intranodal lesions, most are palisaded myofibroblastomas. In 1989, Weiss *et al*.[6] reclassified their collection of 22 cases, including many lesions that were initially diagnosed as intranodal schwannomas, as palisaded myofibroblastomas. These lesions are always characterized by extensive foci of hemorrhage and stellate areas of collagen deposition, the so called "amianthoid fibers" that are not present in intranodal schwannomas.[5,6] Based on immunohistochemical staining, these lesions express smooth muscle actin, but not S-100 protein, which is different from an intranodal schwannoma.[5,6] An uncommon benign leiomyoma that metastasizes to a lymph node may resemble an intranodal schwannoma. The more conventional appearance of smooth muscle cells, along with the immunohistochemical features, is helpful to make a correct diagnosis.[7] In the case of metastatic spindle cell tumors in lymph nodes, the tumor cells exhibit cytological malignant features and mitotically active spindle cell features that are different from the bland-looking spindle cells of a schwannoma.

In summary, we have reported on an extremely rare case of a schwannoma arising in a lymph node. Similar to schwannomas in other sites, intranodal schwannomas appear to behave in a benign fashion. Because of their rarity, a schwannoma that occurs at this particular site can pose problems for its diagnosis. However, an intranodal schwannoma, in most instances, can be easily recognized based on the histological and immunohistochemical findings if an intranodal schwannoma is considered in the differential diagnosis of a primary lesion with a spindle cell appearance in a lymph node.

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