CASE REPORT

Schwannomas of the Left Adrenal Gland and Posterior Mediastinum

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Schwannoma is a rare tumor of neural crest cell origin. Most schwannomas occur in the head, neck, stomach or limbs, with a few cases occurring in the retroperitoneal space. A 30-year-old Taiwanese woman presented with a 1-week history of left anterior chest discomfort and left flank pain. The laboratory findings and endocrine studies were all within normal limits. Chest X-ray revealed masses in the posterior mediastinum. Chest computed tomography and magnetic resonance imaging showed several masses in the left paraspinal region and in the left adrenal region. The patient underwent total excision of the left paraspinal tumors and laparoscopic left adrenalectomy. Pathologic studies showed a picture of benign schwannoma. In conclusion, preoperative differentiation of benign schwannoma from malignant peripheral nerve sheath tumor or other tumors is important for good prognosis. Total excision of benign schwannoma is associated with favorable outcome in patients. [*J Chin Med Assoc* 2009;72(2):83–87]

Key Words: adrenal gland, posterior mediastinum, schwannoma

Introduction

Schwannoma is a rare tumor of neural crest cell origin. Schwann cells, which make the myelin in the peripheral nervous system (PNS), are very important in the regeneration of damaged peripheral nerves.[1] Schwannomas are usually benign slow-growing tumors, and less than 1% become malignant. Most commonly, schwannomas occur as solitary encapsulated subcutaneous tumors. More rarely, they are multiple or arise from points along the PNS, including cranial nerves, spinal roots, the brachial and lumbar-sacral plexus, or major peripheral nerves.[1]

Most schwannomas occur in the head, neck, stomach[2] or limbs, with a few cases occurring in the retroperitoneal space. Schwannomas have been reported to occur over a large age range.[3–9] There appear to be no racial or sex predilection for schwannomas. Histologically, the growth patterns in schwannomas include Antoni type A neurilemoma and type B neurilemoma.[1] Immunohistochemistry is a useful technique for detecting tumors originating from Schwann cells because they stain positive for S-100 antigen, collagen IV and laminin, and show absence of reactivity for keratin, desmin, actin, muscle-related antigens, HMB-45, Melan-A, chromogranin, synaptophysin, and CD34.[3,4,9] Here, we describe a rare case of schwannomas occurring concurrently in the left adrenal gland and posterior mediastinum of a young woman.

Case Report

In May 2006, a 30-year-old Taiwanese woman presented with a 1-week history of left anterior chest discomfort and left flank pain. Physical examination revealed a healthy-looking woman 160 cm tall and 50 kg in weight. Her body mass index was 19.5. No abnormalities were found except for mild left flank knocking pain sensation and anterior chest compression. Chest X-ray revealed left paraspinal lobulated soft tissue tumor (Figure 1A).

Laboratory data revealed normal hematology and biochemistry. Endocrine studies showed normal levels of: serum calcitonin of < 14.0 pg/mL (normal, < 42 pg/mL); cortisol (random) of 10.85 μg/dL (normal, 6–22 μg/dL); corticotropin (ACTH) of 29.4 pg/mL (normal, 6–56.7 pg/mL); aldosterone of 117.2 pg/mL

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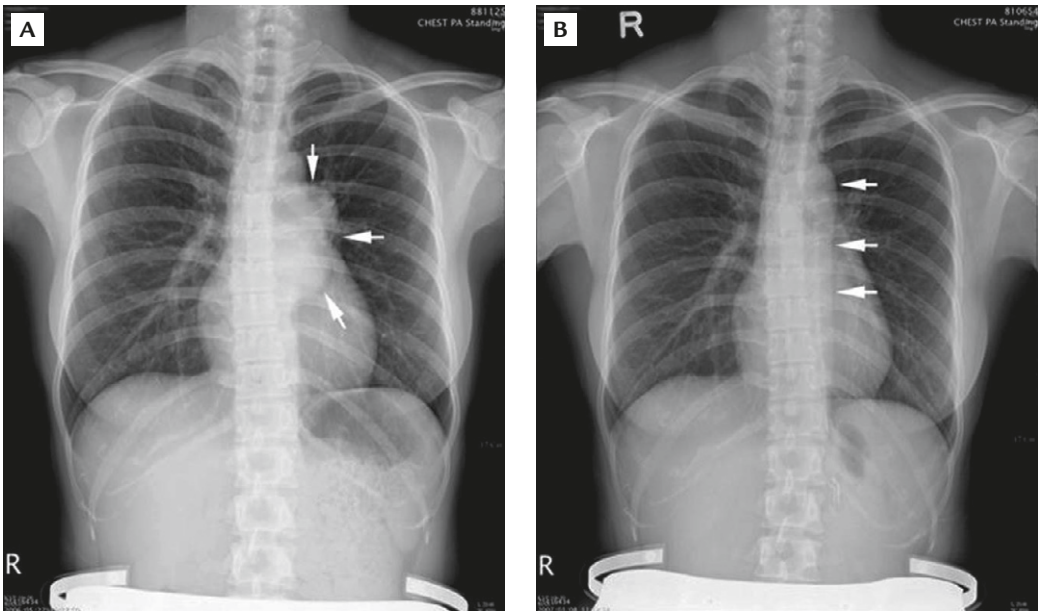

Figure 1. Chest X-ray obtained in the standing position: (A) preoperative X-ray shows left paraspinal lobulated soft tissue tumor (arrows); (B) 1 year after the operation, X-ray shows no recurrent paraspinal tumor over the previous tumor region.

(standing, 70–350 pg/mL); and renin of 1.95 ng/mL (standing, 1.31–3.95 ng/mL). There were also normal levels of 24-hour urine (2,000 mL) free cortisol of 5.1 μg/day (normal, < 60 μg/day), vanillylmandelic acid (VMA) of 3.7 mg/day (normal, 1.0–7.5 mg/day), 17-ketosteroids (17-KS) of 9.12 mg/day (normal, 6–14 mg/day), adrenaline of 4.5 μg/day (normal, < 22.4 μg/day), noradrenaline of 33.3 μg/day (normal, 11.1–85 μg/day), and dopamine of 264 μg/day (normal, 50–450 μg/day), with only mild elevation of 17-hydroxycorticoids (17-OHCS) of 9.68 mg/day (normal, 2–8 mg/day).

Chest computed tomography (CT) revealed several well-marginated heterogeneously enhanced masses with central low density in the left paraspinal region and left adrenal region (Figures 2A–D). Magnetic resonance imaging (MRI) showed tumor masses arising from the left adrenal gland and in the retrocrural and thoracic paraspinal regions (Figure 3).

The patient underwent total excision of the left thoracic paraspinal tumors through a posterolateral thoracotomy and laparoscopic left adrenalectomy. Pathologic finding (Figures 4A and 4B) and immunohistochemical examination (Figures 4C and 4D) of the tumors from both the posterior mediastinum and the left adrenal gland showed a picture of benign schwannoma.

The postoperative course was smooth. The patient was discharged in a good condition after a 1-month hospital stay. At the 18-month follow-up, endocrine studies showed normal serum levels of cortisol (random) (7.32 μg/dL) and ACTH (38.2 pg/mL). Chest X-ray (Figure 1B) and abdominal and chest CT (Figures 2E–H) revealed no evidence of tumor recurrence or metastasis.

Discussion

Clinically, schwannomas usually do not cause symptoms; our patient's symptoms of anterior chest discomfort and left flank pain may be due to tumor compression of intercostal nerves or airways or organs (adrenal gland).[10] Schwannoma can also occur in the mediastinum[11] as in our patient. Schwannoma and neurofibroma represent the most common mediastinal neurogenic tumors; they rarely degenerate into malignant tumors of nerve sheath origin.[11] A malignant peripheral nerve sheath tumor (MPNST) has a high risk of recurrence with incomplete resection; postoperative irradiation and chemotherapy are necessary. We can differentiate benign schwannoma from MPNST in many aspects according to the findings of Lai et al[10] and Hrehorovich et al.[12]

Antoni type A neurilemoma has elongated spindle cells arranged in irregular streams and is compact in nature, and type B tissue has a looser organization, often with cystic spaces intermixed within the tissue. The presence of cystic changes within a retroperitoneal tumor is relatively frequent in schwannomas and may suggest its diagnosis.[13] The tumors of our patient belong to both type tissue patterns. A diagnosis of schwannoma can be supported by immunohistochemical findings

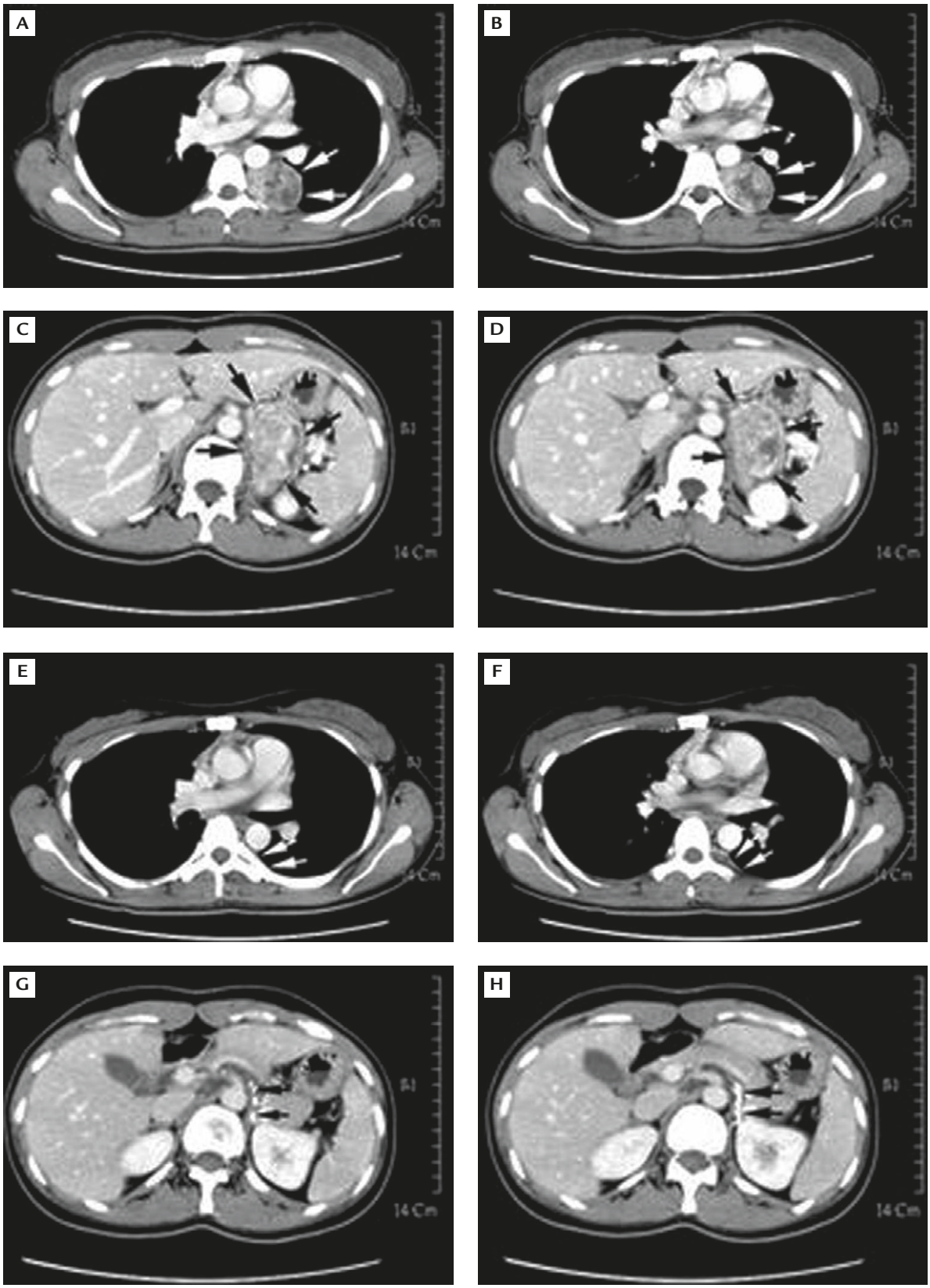

Figure 2. Computed tomography (CT) of the chest and adrenal gland: (A–D) before surgery, CT revealed several well-marginated heterogeneously enhanced masses with central low density in the left paraspinal region and left adrenal region (arrows); (E–H) 18 months after surgery, complete resection of the tumor masses is confirmed, with only surgical clips left over the previous tumor region (arrows), without tumor recurrence or metastasis.

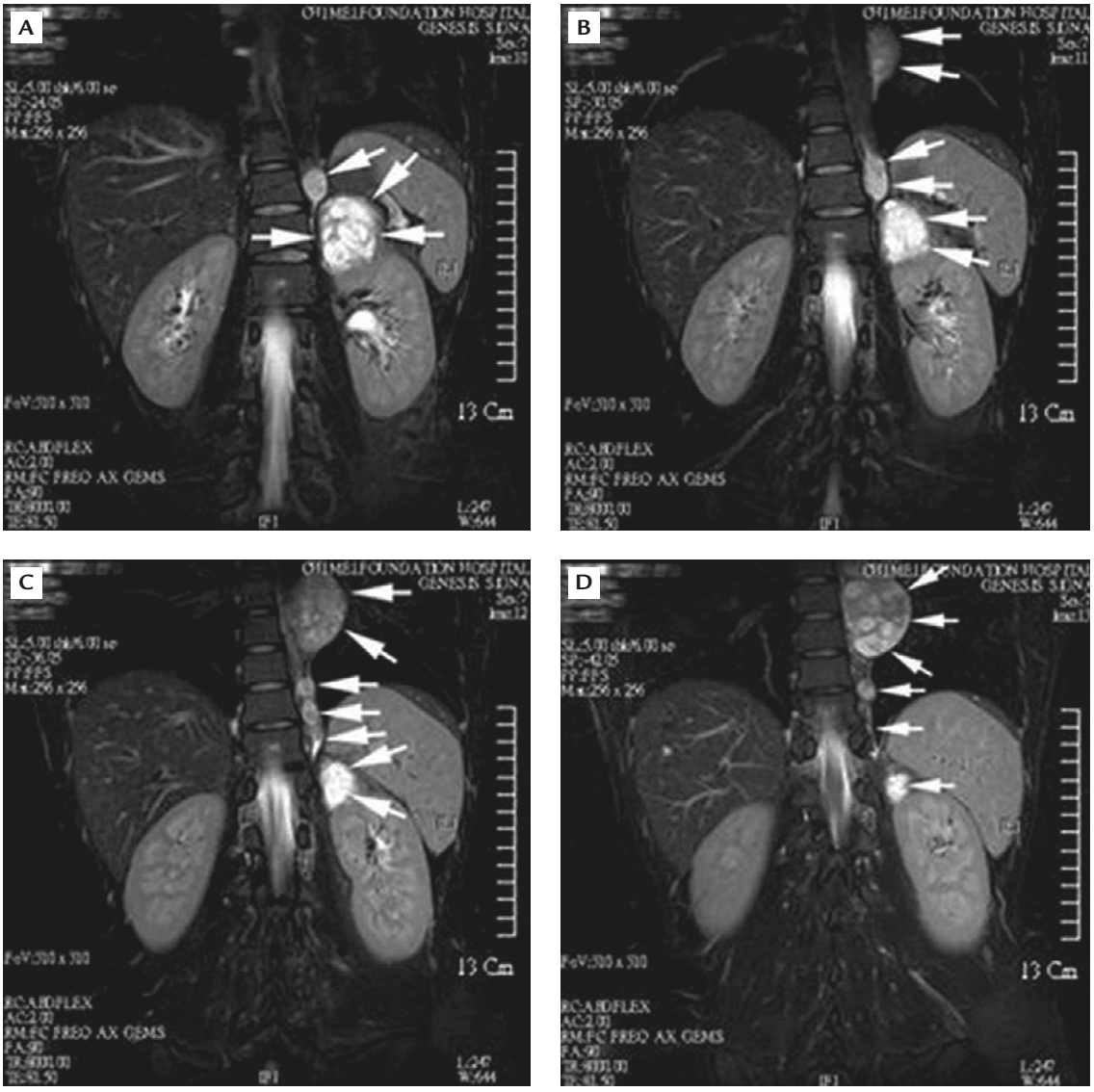

Figure 3. T2-weighted magnetic resonance imaging of the chest and adrenal gland shows high-signal intensity tumor masses on the left adrenal gland and in the retrocrural and thoracic paraspinal regions (arrows).

and electron microscopy.[9] The tumors of our patient stained positive for S-100 protein and negative for CD34, which was the picture of schwannoma.

Our patient's imaging studies showed multiple round, elliptical, lobulated paraspinal masses with mixed attenuation and cystic appearance. The cystic spaces can result in cystic appearance on CT[10,11] and high signal intensity on T2-weighted MRI.[1,10,12,14,15] ^{123}I-meta-iodobenzylguanidine scan has also been shown to be useful in the diagnosis of juxta-adrenal schwannoma.[16] The specific features of schwannoma on imaging studies can differentiate it from other tumors in the posterior mediastinum and adrenal area before operation.

The management of schwannoma requires complete surgical excision. Thoracoscopic resection has been reported as the preferred approach to posterior mediastinal neurogenic tumors.[17] Laparoscopic adrenalectomy is safe and feasible for diagnosis and treatment of benign adrenal or retroperitoneal schwannoma; postoperative recovery is fast.[18]

The prognosis of schwannoma depends on the pathologic features. In central nervous schwannomas, morbidity and mortality result from nerve dysfunction and brainstem compression. Schwannomas arising in other areas are associated with less morbidity and mortality after complete excision.[1] The prognosis is poor in patients with malignant or metastatic tumors.

In conclusion, differentiation of benign schwannoma from MPNST or other tumors before operation is important for good prognosis. Total excision of benign schwannoma is associated with favorable outcome in patients.

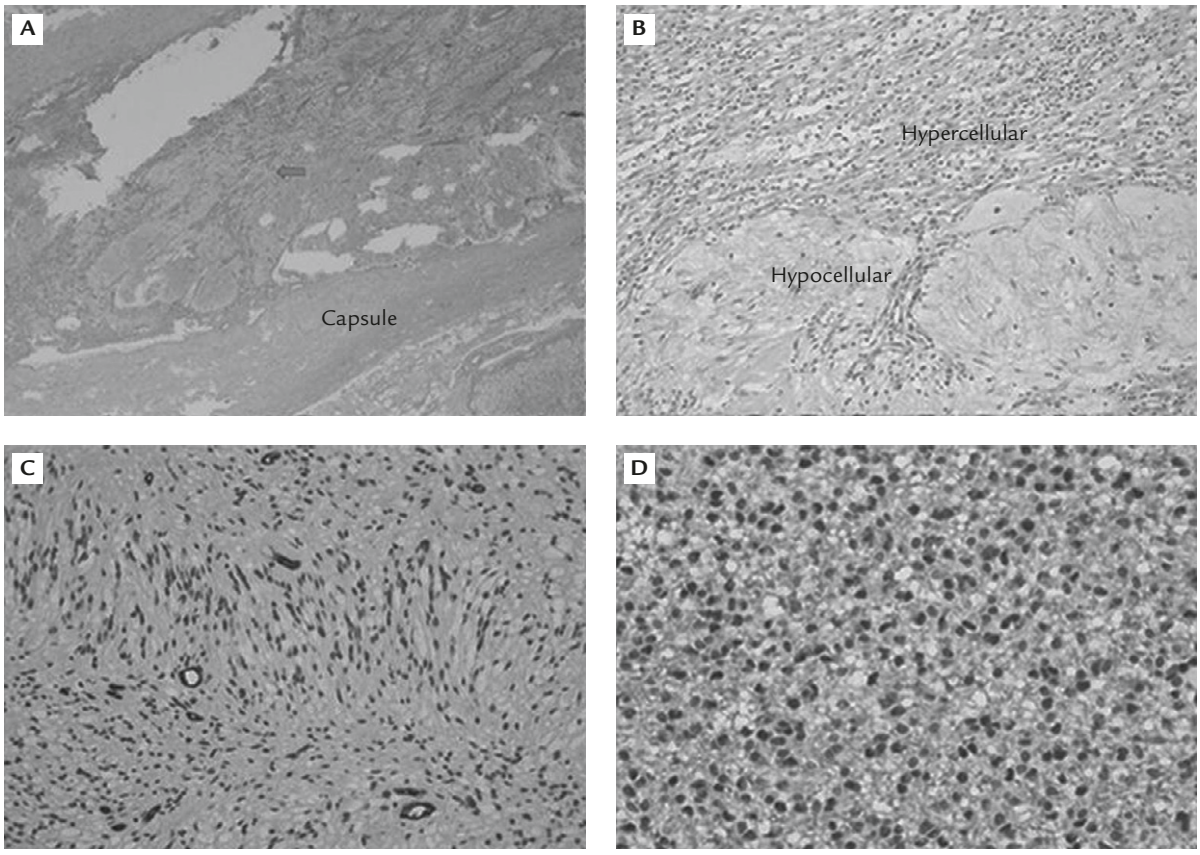

Figure 4. (A, B) Tumor pathology showed ovoid to spindle Schwann cells arranged in fascicles with stromal myxoid changes. (C) Immunohistochemically, CD34 staining highlighted the vascular elements but not the tumor cells. (D) The tumor cells were positive for S-100 in a combined nuclear and cytoplasmic pattern, which is diagnostic for schwannoma.

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