Schwannomatosis is a rare form of neurofibromatosis and is characterized by more than one schwannoma without any sign of neurofibromatosis. We report a case of a 60-year-old male patient admitted with progressive chest discomfort who was found to have six encapsulated tumors along the seventh intercostal space with a maximum tumor size of $3 \times 2 \times 1$ cm.

Key Words: schwannomatosis, single intercostal nerve

Schwannomatosis is a rare form of neurofibromatosis and is characterized by more than one schwannoma without any sign of neurofibromatosis. We report a case of a 60-year-old male patient admitted with progressive chest discomfort who was found to have six encapsulated tumors along the seventh intercostal space with a maximum tumor size of $3 \times 2 \times 1$ cm.

**Case Presentation**

A 60-year-old male patient was admitted with worsening chest discomfort over a 1 month period. The patient had a 10-year history of similar vague chest discomfort. Physical examination and laboratory data were negative. A chest X-ray showed multiple oval-shaped masses overlying the left lung field and a sawtooth-like sign on the seventh rib (Figure 1). Computed tomography of the chest showed multiple homogeneous, well-defined, oval-shaped, extrapulmonary tumors along the seventh intercostal space in the posterior chest wall (Figure 2). Thoracoscopic surgical exploration found six encapsulated tumors along the seventh intercostal space with a maximum tumor size of $3 \times 2 \times 1$ cm (Figure 3). All of the tumors were completely resected and the histological diagnosis was schwannoma. The patient is currently symptom-free at 2-year follow-up.

**Discussion**

Schwannoma is a benign nerve sheath tumor that is the most common neurogenic tumor of the thorax, but it is very rare to find multiple schwannomas arising from a single intercostal nerve [1]. Patients can be
definitively diagnosed with schwannomatosis if they have had two or more pathologically confirmed schwannomas without radiographic evidence of a vestibular nerve tumor at greater than 18 years of age. If radiologic examination, such as brain magnetic resonance imaging is not available, then a probable diagnosis can be made if the patient has two or more pathologically confirmed schwannomas and no clinical evidence of eighth nerve dysfunction at greater than 30 years of age, or two or more schwannomas in an anatomically limited distribution without clinical evidence of eighth nerve dysfunction at any age [6]. In our case, multiple schwannomas were present in a 60-year-old man with no stigmata or family history of type 1 or type 2 neurofibromatosis. He had no evidence of vestibular schwannoma or other intracranial tumors. Multiple peripheral tumors were found in the seventh intercostal space.

Schwannomatosis is often symptom-free and is usually an accidental finding. When symptomatic, these tumors typically cause radicular pain that is distributed along the length of the affected nerve [7]. Jeppesen [8] reported an intercostal neurinoma causing recurrent chest pain. All of the other cases reported were asymptomatic [3]. Our patient had progressive chest discomfort that was possibly caused by compression of the nerve root or rib cage. However, it is

Figure 1. Chest X-ray shows multiple oval-shaped masses over the left lung field (arrow). A sawtooth-like sign can be seen on the seventh rib (arrowhead).

Figure 2. Chest computed tomography shows homogeneous, well-defined, oval-shaped, extrapulmonary tumors along the seventh intercostal nerve in the posterior chest wall (arrow).

Figure 3. (A) Thoracoscopic view during surgery revealed multiple tumors along the seventh rib (arrow). Six tumors were resected. (B) The largest tumor is 3 × 2 × 1 cm in size (arrowhead).
interesting that numerous small tumors compressing the rib were detected under thoracoscopic surgical exploration.

The radiologic features of schwannomatosis on contrast-enhanced computed tomography include multiple, iso- or slightly hypodense, homogeneous, well-defined, oval- or round-shaped mass lesions. On magnetic resonance imaging, schwannomatosis demonstrates slightly iso- or hypointense masses on T1-weighted images, and increased signal intensity on T2-weighted images [2]. The lesion may be easily detected by computed tomography or magnetic resonance imaging; however, the lesion may be suspected from plain chest film, as in our case. On chest X-ray, the sawtooth-like sign on the seventh rib was likely induced by tumor compression.

Surgery is indicated for symptomatic lesions in schwannomatosis [9]. Our patient’s chest discomfort resolved postoperatively. Intercostal schwannoma should be considered in the differential diagnosis of intercostal neuralgia, and a chest radiograph is often sufficient to demonstrate this rare but treatable condition [6]. Malignant transformation has been reported in 10% of schwannomatosis cases [10].

Schwannomatosis has been recently recognized as a third form of neurofibromatosis, in addition to type 1 and type 2 neurofibromatosis. Schwannomatosis shares many features with the better-known forms of neurofibromatosis. Our case had no characteristics suggestive of type 2 neurofibromatosis, but we will still need to follow-up this case closely, because recurrence, although rare, has been reported.

**REFERENCES**

單一肋間神經許旺氏細胞瘤 — 病例報告

李瑞文 1 許正義 2
國軍高雄總醫院 1耳鼻喉科 2胸腔外科

單一肋間神經的許旺氏細胞瘤非常少見。至今全世界的病例報告文獻不到 10 例，一位 60 歲台灣男性因漸歇性的胸痛來求診，胸部 X 光片可見左側肺野有多顆腫瘤以及腫瘤壓迫第 7 肋骨的明顯跡跡，電腦斷層顯示多顆均質性、邊緣規則、卵圓形的肺外腫瘤。胸腔內視鏡手術發現沿著第 7 肋間有 6 顆腫瘤，予以全數切除，病理報告為許旺細胞瘤。追蹤病患迄今 2 年，並無復發跡象。

關鍵詞：許旺氏細胞瘤，單一肋間神經
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