Metachronous mediastinal and retroperitoneal schwannomas

Schwannomas mediastinal y retroperitoneal metacrónicos

Josefina M. P. Furque, Oscar Brosutti, Ángel L. Pierini, Aída V. Canga, Melisa M. del C. Zuttión

Department of Mixed General Surgery. Hospital J. B. Iturraspe. Santa Fe, Argentina

Conflicts of interest None declared.

Correspondence: Josefina M. P Furque E-mail: josefurque@gmail.com

ABSTRACT

Schwannomas, also known as neurilemoma, are neurogenic tumors that arise from the peripheral nerve sheaths. Forty-five percent of schwannomas occur in the head and neck, 9% in the mediastinum and 0.7-2.7% in the retroperitoneum. Multiple schwannomas are extremely rare. We report the case of a 30-year old male patient with chest pain in the right hemithorax associated with pleural effusion due to schwannoma of the posterior mediastinum that was completely resected with video-assisted thoracoscopy. Three years later, he presented pain on the right lumbar region due to a retroperitoneal schwannoma behind the vena cava that was completely removed with open surgery. We describe the tests used to evaluate this tumor and the histopathological findings confirming its benign nature.

Conclusion: Schwannoma is a rare condition with excellent postoperative outcome after complete surgical resection.

Keywords: multiple schwannoma, neurilemoma, neurinoma, posterior mediastinum, retroperitoneum.

Schwannomas, also known as neurilemomas, are rare tumors that arise from Schwann cells of the peripheral nerves. Retroperitoneal schwannomas are uncommon.

Schwann cells form the myelin sheath on peripheral nerves and play a very important role in regenerating damaged peripheral nerves. Schwannomas are usually benign, slow-growing, and less than 1% becomes malignant. They generally occur as solitary encapsulated subcutaneous tumors. More rarely, they are multiple or arise from points along the peripheral nerves, including cranial nerves, spinal roots, the brachial plexus and lumbosacral plexus, or major peripheral nerves. Multile schwannomas are extremely rare.

We describe an unusual case of a patient with benign posterior mediastinal and retroperitoneal metachronous schwannomas.

A 30-year old male patient sought medical care on April 2012 due to right-sided chest pain associated with exercise-induced dyspnea within the past month. The patient underwent chest X-Ray and computed tomography (CT) scan which showed moderate right pleural effusion and a tumor with a diameter of 10 × 5.5 × 4 cm with well-defined borders in the posterior mediastinum (Figure 1 A and B).

A fine-needle biopsy of the space occupying mass (SOM) was performed. The histopathological and immunohistochemical revealed spindle-shaped cells suggestive of neural tumor. The tumor of the posterior mediastinum was resected by video assisted thoracoscopy (Figure 1, C).

The pathological examination and immunohistochemical tests (staining for vimentin and S-100 protein) confirmed the presence of a benign schwannoma.
The patient was follow-up every 6 months and on November 2015 (3 years later) he made a medical visit due to pain in the right lumbar region within the past month.

A CT scan of the abdomen revealed the presence of a rounded image with increased concentration of the contrast material and a diameter of 33 mm in the right adrenal gland (Figure 2 A and B).

A magnetic resonance imaging (MRI) scan identified that the lesion was below the adrenal gland, displacing it in cranial direction, and could correspond to an adenomegaly; yet, other etiologies could not be ruled out. The inferior vena cava was displaced anteriorly by the mass and was partially collapsed.

The evaluation was completed with a positron emission tomography (PET)/CT scan. A retroperitoneal nodular mass with soft tissue density (SUV 6.7) measuring 33 × 39 cm was detected behind the vena cava in intimal contact with the right adrenal gland. The mass was suggestive of adenomegaly without clear fatty cleavage plane with neighboring structures.

Endocrine tests ruled out a functioning tumor.

The patient underwent resection of the retroperitoneal SOM plus en bloc resection of the right adrenal gland through a Kocher incision in December 2015.

The pathological examination reported the presence of a spindle-shaped cells tumor measuring 4.5 × 4.2 × 2.5 cm suggestive of schwannoma. Mitotic count: 0-1 mitosis per 10 high power fields. Microscopic examination revealed the presence of spindle-shaped cells with elongated nuclei arranged in palisades (Antoni A areas) and other areas with abundant edematous stroma (Antoni B areas). The adrenal gland parenchyma was preserved (Fig. 2 C).

The correlation between the immunophenotype, patient’s history and the morphologic characteristics were consistent with the diagnosis of benign schwannoma, positive for the S-100 protein and negative for CD34, CD117 and smooth muscle actin.
As multiple schwannomas are common in patients with Von Recklinghausen’s disease, this condition was ruled out.

The patient is currently followed-up every 6 months, without evidence of new lesions.

Schwannoma was described by Verocay in 1910, who called it encapsulated neurofibroma. In 1935, Scout coined the term neurilemoma, also known as schwannoma.

Schwannomas commonly occur as slow-growing solitary tumors, well-circumscribed and encapsulated. They most generally develop between 25 and 55 years (as in our case) but can occur at any age, with no predilection for sex.

Forty-five percent of schwannomas occur in the head and neck, with 9% occurring in the mediastinum. Schwannoma is the most common (about 50%) mediastinal neurogenic tumor.

Retroperitoneal localization is very uncommon (0.7% to 2.7% of all schwannomas) and has been associated with Von Recklinghausen’s disease. In this case, tumors are usually malignant with adverse outcome.

Schwannomas most frequently arise and grow in the posterior and posterolateral sites of the spinal cord on the thoracic region. In the retroperitoneum, these tumors develop in the anterolateral site of the spinal cord.

Symptoms are rare in retroperitoneal or mediastinal schwannomas. Diagnosis is often an incidental finding on routine radiography or the tumor presents with symptoms associated of compression of the neighbor structures. In our patient, pain on the chest and right lumbar region can be due to tumor compression of intercostal nerves or airways or organs (adrenal gland), as Chwen-Yi Yang et al. described in their publication.

These tumors are usually large (some series describe sizes between 8 and 20 cm) probably due to the flexibility of the retroperitoneal cavity. Mediastinal schwannomas are smaller. However, large tumors are not aggressive and are not associated with risk of malignancy, which is extremely low.

Ultrasound and CT scan are helpful to determine tumor size, location, invasion or involvement of neighbor organs; yet, none of these methods constitute the gold standard. Recent reports have suggested that magnetic resonance imaging is the best method. Often it is even possible to find the parent nerve.

Recently, with advances in imaging modalities, incidental detection of retroperitoneal tumors has been increasing, especially with increasing medical examination health screening. Small schwannomas appear as a homogenous, encapsulated mass, whereas large schwannomas show cystic degeneration, hemorrhage or central necrosis. The most characteristic finding in schwannoma is cystic change. They usually have a round or oval shape.

The specific features of schwannoma on imaging studies can differentiate it from other tumors in the posterior mediastinum and adrenal area before surgery. However, the diagnosis of schwannoma before surgery is not common.

Given that schwannomas show variable 18F-fluorodeoxyglucose (FDG) uptake, with the SUV ranging from 1.9 to 12.0, PET scan has limited utility in distinguishing between schwannomas and malignant peripheral nerve sheath tumors. In many cases, it is impossible to distinguish between a schwannoma and a malignant tumor before biopsy or surgery precisely because schwannomas can show high FDG uptake (as in our case).

The management of schwannoma requires complete surgical excision, with favorable outcome. Thoracoscopic resection has been reported as the preferred approach to posterior mediastinal neurogenic tumors. Laparoscopic adrenalectomy is safe and feasible for diagnosis and treatment of benign adrenal or retroperitoneal schwannoma. In our case, we decided to remove the retroperitoneal tumor via open surgery because of its retrocaval location on images tests, with stenosis of the vena cava and absence of clear cleavage plane.

When total resection is not achieved, consideration of the Ki-67 index and percentage of p53-positive cells is recommended as an indicator of growth and malignant transformation potential. Nevertheless, regular follow-up is necessary in all cases.

References