Case Report

Schwannoma presenting as an apical thoracic tumour with upper extremity symptoms

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SUMMARY. The case of an apical tumour in the thorax, which presented with upper extremity symptoms, is reported. Surgery was performed and a diagnosis was established of a neurogenic mediastinal tumour, a schwannoma, compressing the T1 nerve root. This case illustrates the need for the thoracic surgeon to consider a brachial plexus or upper thoracic nerve root tumour when evaluating a patient with neurological upper extremity symptoms. Adequate preoperative evaluation is essential in such cases to enable the thoracic surgeon to select the best surgical approach, sometimes aided by a neurosurgical team. Emphasis is placed on the difficulties faced by the surgeon when exposing a tumour complicated by its localization in the superior mediastinum and which is anatomically linked with delicate nerve fascicles. *Pneumon 2010, 23(4):380-383.*

1. INTRODUCTION

Neurogenic tumours frequently appear in the thorax, where they comprise 12-21% of all mediastinal tumours. The predominant histological types are schwannoma, neurofibroma and ganglioneuroma. They are usually located in the posterior apical compartment (95%) and are mostly asymptomatic [1]. The case reported here is of a rather rare manifestation of a neurogenic tumour of the upper posterior mediastinum, which clinically and radiologically mimicked an apical lung tumour.

2. CASE REPORT

A 72 year-old female was referred for surgical assessment after evaluation for pain and numbness of the ring and little fingers of the right hand. The symptoms had appeared 8 months earlier and had been gradually worsening. The patient's history was significant for a left modified radical mastectomy for breast cancer carried out 5 years earlier. During the course of her follow-up, a chest X-ray revealed a smooth, round, mass 3 cm in diameter at the apex of the right lung. The mass was shown on computed tomography (CT) scan to have benign features, and a possible diagnosis

of a schwannoma was made. There were no findings suggestive of breast cancer recurrence. The evaluation proceeded with magnetic resonance imaging (MRI) which showed in more detail a mass in the lower cervical region extending to the apex of the right hemithorax. The mass was cystic and its appearance was consistent with the diagnosis of a schwannoma. The patient was monitored for 3 years with yearly CT scans. The mass was stable at first, but then showed enlargement to 4.5 cm x 4 cm 6 months before referral. Preoperative X-ray and CT scan confirmed the above findings of the apical mass, and surgical clips from the earlier mastectomy (Figure 1, 2). Serum results were normal. Further exploration of the nature of the tumour was attempted with a fine needle aspiration, but the findings were non-diagnostic.

On physical examination the patient was stable. There was no palpable cervical mass but the neurological examination revealed paraesthesia and loss of strength in the right hand, with an ulnar distribution, raising the possibility that the symptoms in the extremity were caused by a compression of the T1 nerve root. It was decided to perform resection of the tumour.

The tumour was approached via a right posterolateral thoracotomy. A brownish, well circumscribed, encapsulated, elastic-soft mass was located in the upper right posterior mediastinum. It appeared to originate near the 1st intervertebral foramen, without evidence of intraspinal extension (Figure 3). The mass was removed by enucleation from its capsule.

On gross examination the mass measured 3.5 cm x 2 cm x 0.5cm and sectioning revealed complete cystic degeneration with micro-haemorrhages in the cyst wall. Microscopically, the lesion corresponded to a mesenchymal neoplasm with spindle-shaped cells and nuclear palisading, arranged in a plexiform pattern (Figure 4), and was diagnosed as a benign schwannoma (neurilemmoma). An enlarged topical lymph node of 0.9 cm in size showed signs of reactive lymphadenitis.

The postoperative course was complicated by bleeding originating from the thoracotomy site, which was managed conservatively with plasma and packed red blood cell transfusion. The patient was eventually discharged on the 8th postoperative day. Twelve months after surgery the patient's symptoms have improved, although there is some residual numbness of the right arm.

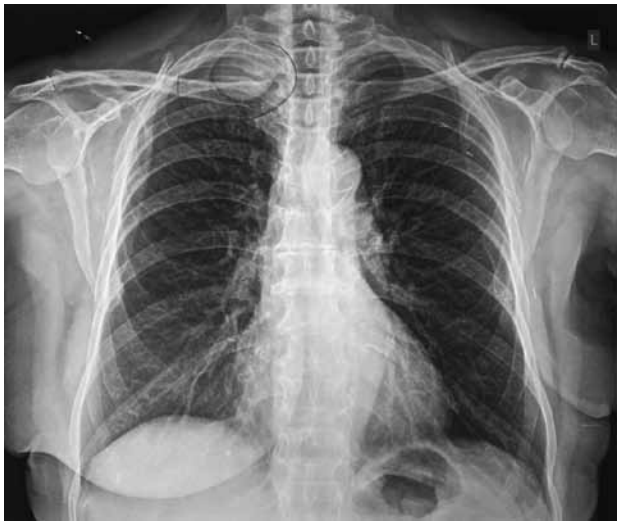

FIGURE 1. Anteroposterior chest X-ray of the patient.

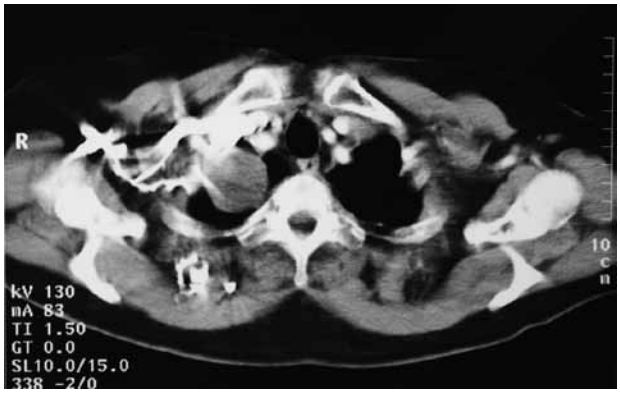

FIGURE 2. Computerized tomography image of the tumour.

3. DISCUSSION

The Fraser classification which divides the mediasti-

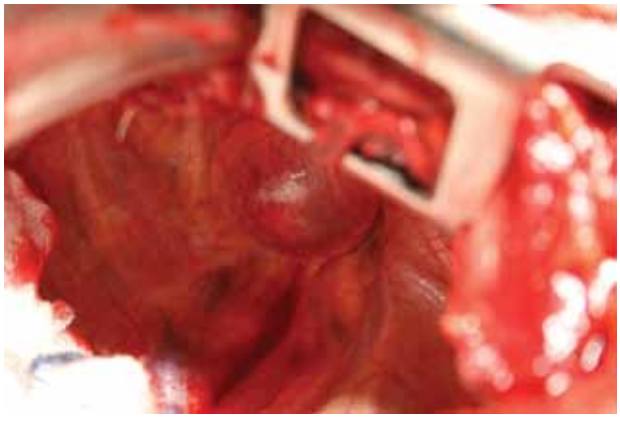

FIGURE 3. Intraoperative view of the tumour.

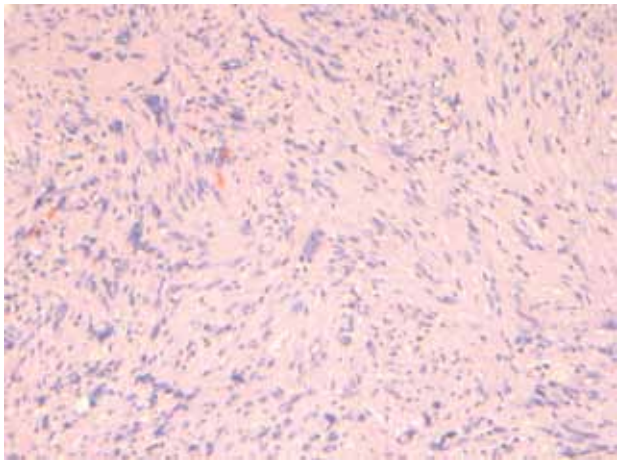

FIGURE 4. Histological slide (H&E counterstain, magnification X100), showing representative areas of the benign neurilemmoma.

num into anterior, middle and posterior compartments is not only easy to use, but it is also important for the diagnosis of mediastinal tumours, which exhibit heterogeneity. Thymomas, germ cell tumours and lymphomas are more frequently located in the anterior compartment, lymphomas and cystic tumours, such as pericardial and bronchogenic cysts, in the middle compartment, and neurogenic tumours in the posterior compartment. The location of a tumour is correlated with the likelihood of malignancy, which has been reported to be 59%, 29% and 16% for the anterior, middle and posterior compartments, respectively. Increasing age and the presence of tumour-related symptoms are also correlated with malignant tumours[2].

Schwannomas constitute the majority of neurogenic tumours of the mediastinum. They arise in the thorax from the nerve sheaths of spinal nerve roots or of any thoracic nerve. They are encapsulated tumours and often show heterogeneity, due to cystic degeneration, haemorrhage, myelin deposition and calcifications. The majority of these tumours are asymptomatic and they are frequently discovered incidentally on chest X-ray, often not before reaching a significant size. They can become symptomatic, however, as they get larger, due to local compression of nerves, bone erosion or distortion of the ipsilateral lung. The symptoms are mainly pulmonary or neurological. When the tumour arises from an intercostal nerve, either at its root or on the thoracic wall, it typically presents with intercostal neuralgia. There have been reports of neurogenic tumours causing Horner's syndrome, cough, dyspnoea, dysphagia, chest pain and, in rare cases, haemoptysis[3-6]. The combination of the specific location of the tumour in this report in the thoracic apex, its radiological appearance and its clinical presentation - unusual for a benign posterior mediastinal tumour - is interesting in its rarity. For a neurogenic tumour localized in the chest to produce upper extremity symptoms, it would have to be either a branchial plexus tumour extending into the chest or a tumour compromising the T1 root, both of which are infrequent[5-7].

Schwannomas appear on chest X-ray as sharply demarcated spherical, sometimes lobulated masses. After identification of the tumour and confirmation of its posterior mediastinal location on posteroanterior and lateral chest X-ray, CT scan is indicated for further evaluation of possible local invasion and determination of the anatomical relationship of the tumour to the various adjacent structures. MRI is of greater value when intraspinal extension is suspected or more detailed imaging of adjacent vascular structures is required[3,8].

Surgical resection is the treatment of choice for these tumours, and the procedure is usually both diagnostic and therapeutic. Currently, video-assisted thoracoscopic surgery techniques (VATS) are used extensively for the excision of such tumours. A tumour located near the thoracic outlet was initially considered a contraindication for VATS excision, although in recent years such excisions have been carried out successfully[1,3-5].

Neurogenic tumours of the thoracic apex and those involving the brachial plexus are particularly interesting since they can be accessed through various surgical approaches. The standard approach is from below, using a posterolateral thoracotomy, but this offers limited exposure of the more superiorly situated tumours. The posterior subscapular approach provides excellent exposure and is particularly suitable for safe dissection of the tumour away from nerve fascicles and for tumours with intraforaminal components[6,9]. An anterior cervical – trans-sternal approach has also been described, and when malignancy is suspected or for especially bulky tumours, a Dartavelle technique can also be used[7]. As noted above, VATS is also an option for these tumours but is subject to the same limitations in exposure as open thoracotomy, although hybrid approaches such as a combined thoracoscopic –supraclavicular approach may prove effective[10].

Schwannomas (in contrast to neurofibromas) are encapsulated tumours which, when benign, as is usually (>95%) the case, can be enucleated so as to avoid nerve injury. In some cases it is technically challenging to dissect the tumour off the nerve fascicles from which it arises.

Intraoperative electrophysiological monitoring may help spare nerve function after tumour resection[5,11].

In conclusion, this case illustrates how a schwannoma of the thoracic apex can be a rare cause of upper extremity symptoms. Surgical resection is, in most cases, necessary to rule out malignancy and to prevent complications. These tumours require thorough preoperative evaluation and meticulous surgical technique for their safe and definitive management.

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REFERENCES

1. Takeda S, Miyoshi S, Minami M, Matsuda H. Intrathoracic neurogenic tumors--50 years' experience in a Japanese institution. Eur J Cardiothorac Surg. 2004 Oct;26(4):807-12.
2. Duwe BV, Sterman DH, Musani AI. Tumors of the mediastinum Chest. 2005 Oct;128(4):2893-909.
3. Zierold D, Halow KD. Thoracoscopic resection as the preferred approach to posterior mediastinal neurogenic tumors. Surg Laparosc Endosc Percutan Tech. 2000 Aug;10(4):222-5.
4. Cardillo G, Carleo F, Khalil MW, Carbone L, Treggiari S, Salvadori L, Petrella L, Martelli M. Surgical treatment of benign neurogenic tumours of the mediastinum: a single institution report. Eur J Cardiothorac Surg. 2008 Dec;34(6):1210-4. Epub 2008 Oct 16.
5. Barrenechea IJ, Fukumoto R, Lesser JB, Ewing DR, Connery CP, Perin NI. Endoscopic resection of thoracic paravertebral and dumbbell tumors. Neurosurgery. 2006 Dec;59(6):1195-201; discussion 1201-2.
6. Bosnjak R, Bacovnik U, Podnar S, Benedicic M. T1-nerve root neuroma presenting with apical mass and Horner's syndrome. J Brachial Plex Peripher Nerve Inj. 2007 Mar 19;2:7.
7. Ladas G, Rhys-Evans PH, Goldstraw P. Anterior cervical-transsternal approach for resection of benign tumors at the thoracic inlet. Ann Thorac Surg. 1999 Mar;67(3):785-9.
8. Yamaguchi M, Yoshino I, Fukuyama S, Osoegawa A, Kameyama T, Tagawa T, Maehara Y. Surgical treatment of neurogenic tumors of the chest. Ann Thorac Cardiovasc Surg. 2004 Jun;10(3):148-51.
9. Kline DG. Posterior approach for plexus tumors in the apical and mediastinal regions. Ann Thorac Surg. 2000 Apr;69(4):1302.
10. Akashi A, Ohashi S, Yoden Y, Kanno H, Tei K, Sasaoka H, Sakamaki Y, Katsura T, Nishino M, Manzurul HS. Thoracoscopic surgery combined with a supraclavicular approach for removing superior mediastinal tumor. Surg Endosc. 1997 Jan;11(1):74-6.
11. Kwok K, Davis B, Kliot M. Resection of a benign brachial plexus nerve sheath tumor using intraoperative electrophysiological monitoring. Neurosurgery. 2007 Apr;60(4 Suppl 2):316-20; discussion 320-1.