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The principles of surgical management in dumbbell tumors

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Abstract *Objective.* Tumors located in posterior mediastinum that extends into spinal canal via intervertebral foramen are called as Dumb-bell or Sand-glass tumors. Most of these tumors are neurogenic in origin but sometimes other rare tumors can also form in this shape. Herein three neurogenic tumors of the mediastinum that extended into the spinal canal are presented *Methods.* In all patients that have been operated in our clinic during 1992–1993, we preferred one-stage removal described by Akwari that consists of posterior laminectomy by neurosurgical team to free the tumor within the spinal cord followed by a posterolateral thoracotomy and excision of the tumor by thoracic surgeons in the same setting. *Results.* All three patients are alive and free of symptoms after 23, 16 and 13 months respectively. Accord-

ing to the pathological examinations of the specimens in the three patients, the exact diagnosis were reported as neurofibroma, paraganglioma and pheochromocytoma respectively.

Conclusions. In recent reports, a combined surgical approach is recommended for dumb-bell neurogenic tumors in posterior mediastinum. We also recommend a combined and one stage removal of dumb-bell neurogenic tumors if possible. A teamwork of thoracic and neurosurgeons will minimize the morbidity and mortality after the surgical procedure, as well as giving the opportunity to remove the tumor totally in one session. [Eur J Cardio-thorac Surg (1996) 10:569–573]

Key words Dumbbell neurogenic tumors · Posterior mediastinum · One-stage removal

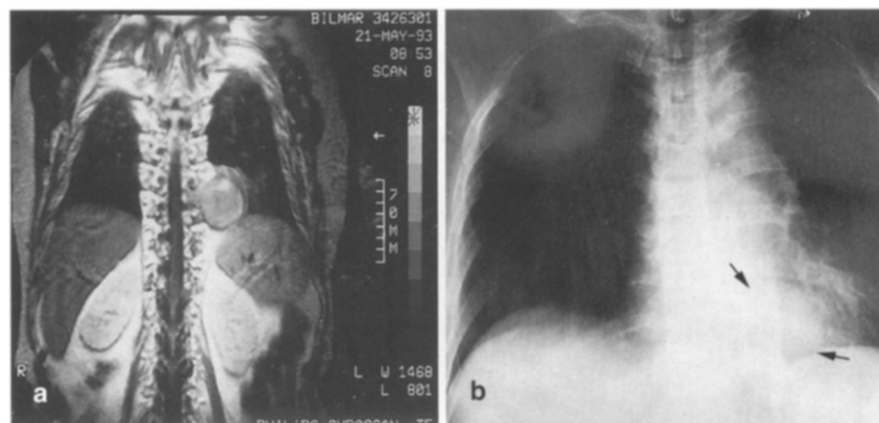
Introduction

Neurogenic tumors are the most common primary mediastinal masses, constituting 20–35% of all lesions in both children and adults [3, 11, 25]. The largest group of patients was reported by Akwari and associates in 1978 [1]. Sixty-nine patients had dumbbell tumors out of 706 mediastinal neurogenic tumors, namely 9.6%. Approximately 10% of all neurogenic tumors located in the posterior mediastinum have intraspinal extensions via intervertebral foramen with a pedicle [1, 8]. In 1929, these tumors were named “hourglass tumors” by Heuer GJ because of their characteristic shape [12]. Although neurogenic tumors are

usually asymptomatic and discovered during a routine chest X-ray, they cause neurological symptoms in 60% of patients with intraspinal extension [7]. They may cause spinal cord damage by compression or overgrowth.

Two main operative techniques have been introduced for dumbbell neurogenic tumors of the posterior mediastinum. In the Akwari method, first the neurosurgical team liberates the spinal extension of the tumor by posterior laminectomy, followed by removal via a posterolateral thoracotomy [1]. This technique is especially favorable for large tumors with vertebral destruction and collapse [18]. In the Grillo method, a midline vertical incision over the spinous processes, extended at the level of the scapular tip, is performed [7]. A small thoracotomy is carried out according

Fig. 1a, b (Patient 1) Magnetic resonance imaging and chest X-ray. **a** The tumor is situated posteriorly over the diaphragm of the left thorax. **b** Anteroposterior view shows the mass near the diaphragm hidden beyond the cardiac shadow (arrows)



to the tumor level. This technique can be used with small tumors involving only one neural foramen [18].

In this report we present three patients with dumbbell neurogenic tumors in the posterior mediastinum. The purpose is to discuss the diagnostic and surgical techniques and the advantages of one-stage removal.

Case reports

Case 1

A 61-year-old woman was admitted to our hospital with back pain of 10 years duration, which had increased in intensity over the previous 4 days. The patient's past history revealed that she had undergone a neurinoma excision from the cervical region 2 years before. During physical examination hypoesthesia below the 8th thoracic vertebra level and 2–3/5 muscle strength loss were noticed on the left half of her body.

A wide 8th–9th left thoracic neural foramen and a bilobed mass extending to the paravertebral region were detected with thoracic magnetic resonance imaging (MRI) (Fig. 1). Cervical MRI was also performed and cervical deformation was detected: angulation at the 4th–6th cervical vertebrae levels, irregular widening of the cord contours, an infarct region in the center of the cord, probable residual mass of the same intensity with the cord at the left contour and a solitary mass (2M×3 cm) of fusiform at the left paravertebral region at the same level. Small airway obstruction was reported after pulmonary function tests, but FEV₁ and FVC were normal. With these findings, the patient was thought to have neurofibromatosis and an operation was planned.

Using the Akwari method, first neurosurgeons performed a posterior laminectomy at the 9th thoracic vertebra level and, to understand the exact level and extent of the tumor, a posterolateral thoracotomy was performed from the 7th left intercostal space. Traction of the mass showed infiltration to the 9th left thoracic spinal nerve. The ninth left thoracic spinal nerve was cut and the mass was freed from the adjacent structures: a 10×10×10 cm mass was resected and totally removed. The pathological result was reported as neurofibroma. She is still alive and healthy 23 months after the operation.

Case 2

A 22-year-old woman had noted weight loss in the last 3 months and her chest X-ray showed a mass. When she was admitted to our hospital, she complained of paraplegia and hypoesthesia of her lower extremities. Her symptoms have begun 15 days before, having started in her right lower extremity and then proceeding to her left lower extremity.

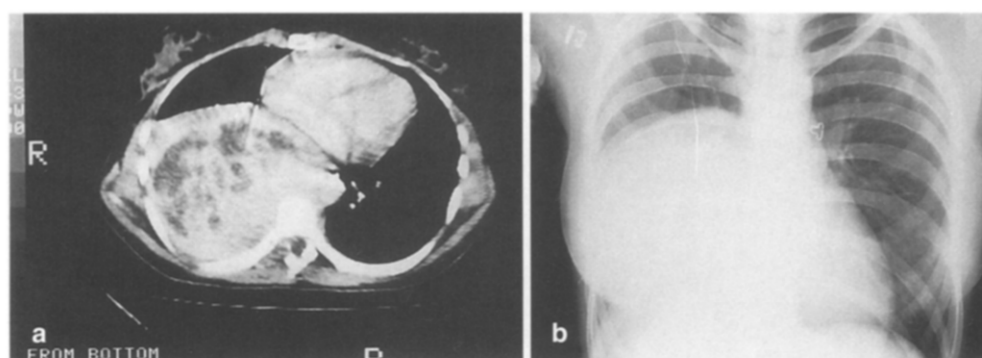
On physical examination, 4/5 and 5/5 muscle strength loss were noted on her left and right lower extremities, respectively. She had bilateral hypoesthesia below the 8th thoracic vertebra level and her deep tendon reflexes were found hyperactive in both of the lower extremities. Magnetic resonance imaging revealed a contrast-keeping mediastinal mass which had infiltrated the vertebrae at the 6th–8th thoracic vertebrae level and obliterated the spinal cord (Fig. 2). A computed tomography (CT) guided fine needle biopsy was reported as malignant. Progression of the hypoesthesia and strength loss led us to consider an emergency intervention. Neurosurgeons performed a 6th–8th thoracic laminectomy and freed the extradural mass from the spinal canal and the vertebrae. Orthopedists performed Cottrel-Debousset (CD) instrumentation and then a right posterolateral thoracotomy was performed. A 14×8×7 cm mass, collapsing the right lower lobe was completely excised. The pathological result showed a paraganglioma. Postoperatively she received adjuvant radiotherapy. She is still alive, healthy and completely mobilized 16 months after the operation.

Case 3

A 17-year-old girl was admitted to our clinic with back pain of 3 months' duration which has intensified recently. On physical examination, her blood pressure was 200/120 mmHg. Doppler ultrasonography of the renal artery and the vanillylmandelic acid level in 24-h urine were reported to be normal. Thoracic CT revealed a soft tissue mass which has infiltrated the medulla spinalis within the spinal canal and destroyed the vertebral bodies and, especially, pedicles at T_{9–10} levels (Fig. 3).

Neurosurgeons freed the mass from the 7th–8th thoracic vertebrae and medulla spinalis and orthopedists inserted a Kirschner pin and cement into the intervertebral space. The 6×6×5 cm mass was totally removed via a right posterolateral thoracotomy. Intraoperatively during tumor removal, her blood pressure rose to 240/120 mmHg but dropped after the administration of nifedipine 10 mg i.v. and furosemide 80 mg i.v. in separate doses. The pathological specimen was reported as a pheochromocytoma. She is still alive and healthy 13 months after surgery.

Fig. 2a, b (Patient 2) Magnetic resonance imaging and chest X-ray. **a** The huge mass is seen just in the right side of the heart. **b** The right hemithorax is totally filled by the tumor



Discussion

Dumbbell neurogenic tumors were identified and named by Heuer [12] in 1929. He named them hourglass tumors of the spine, as did Naffziger and Brown [15] in 1933. The term “dumbbell” was first used by Love and Dodge [14] in 1952, describing the spinal canal involvement. Neurogenic mediastinal tumors originate from the nerve sheath (68%), sympathetic chain (30%) or paraganglionic cells (2%). The recent trend for a combined and total resection has caused an increase in the number of neurogenic tumors resected [17].

The major steps to be followed in preoperative evaluation are [8]:

1. The intervertebral foramen near the tumor location should be scanned for widening and expansion with thin slice CT.
2. If widening or expansion is detected, MRI or myelography and CT should be performed.
3. If extension to the foramen is proved, a combined-team surgical approach should be preferred.
4. If a major spinal artery is at risk from the surgery, its course should be identified with angiography.

Preoperative diagnosis of intraspinal extension is essential for determining the surgical approach. Computed tomography and myelography have been used together for the detection of neural canal involvement [1, 2, 7, 8], but myelography causes discomfort to the patient and CT is limited to the axial plane. Magnetic resonance imaging, because of its accurate description of the existence and longitudinal extension of the spinal component of the tumor, is the preferred diagnostic tool nowadays [10, 18]. The Adamkiewicz artery is the major blood supply of the spinal cord. Its anatomy shows variations, but usually it originates from the left T₉–L₂ intercostal artery. In 15% of the population it originates between T₅–T₈ [4]. If possible involvement is suspected, therefore, an arteriography should be carried out. In two cases the intraspinal extension was scanned by MRI and in one patient with thin slice CT. The pathological examinations revealed neurofibroma, paraganglioma and pheochromocytoma in our patients (Table 1).

Spinal nerve involvement in neurofibromatosis may result in dumbbell-shaped tumors in nearly half of the patients [24]. Family history and typical neurological signs may be lacking. Paraganglioma of the posterior mediastinum is a rare neurogenic tumor arising from the aortico-

Fig. 3a, b (Patient 3) Magnetic resonance imaging and chest X-ray. **a** The mass is seen near the vertebral corpus extending into the neural canal. **b** Anteroposterior view shows the mass hidden in the right side beyond the cardiac shadow (arrows)

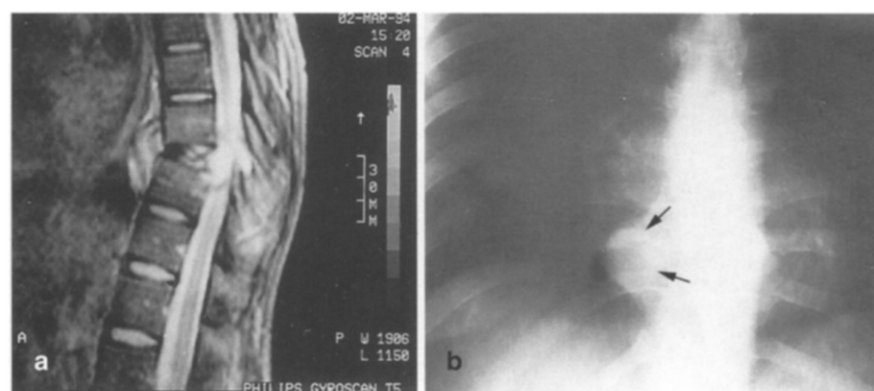


Table 1 Patient data

| Patient no. | Operation type | Tumor dimension (cm) | Laminectomy level | Histology | Follow-up period (months) |
|-------------|---|----------------------|-------------------|------------------|---------------------------|
| 1 | Laminectomy, left posterolateral thoracotomy | 10×10×10 | T ₈₋₁₀ | Neurofibroma | 23 |
| 2 | Laminectomy, CD instrumentation, right posterolateral thoracotomy | 14×8×7 | T ₆₋₈ | Paraganglioma | 16 |
| 3 | Laminectomy, CD instrumentation, right posterolateral thoracotomy | 6×6×5 | T ₇₋₈ | Pheochromocytoma | 13 |

sympathetic ganglia [6]. Paragangliomas are vascularized masses so they keep contrast under CT scanning. I-metaiodobenzylguanidine (¹³¹I-MIBG) has proven to be very useful in localizing catecholamine-producing tumors such as pheochromocytomas and paragangliomas [20]. Magnetic resonance imaging is also an excellent tool for the detection of mediastinal paragangliomas [23].

Definite diagnosis of most mediastinal neurogenic tumors is achieved on surgical excision [5, 11, 22]. Complete resection offering a long survival should be the aim, because 50% of tumors in the posterior mediastinum are malignant and only 3% have tumor metastases. Almost all authors suggest a one-stage operation and complete resection of the tumor; but the techniques advised differ [7, 13, 17–19]. According to the first technique, which was described by Akwari [1], a vertical paravertebral incision and a posterior laminectomy is carried out by neurosurgeons and, after the tumor has been freed from the spinal canal, a posterolateral thoracotomy is performed and the tumor removed. We used Akwari's technique in all patients (Table 1). It is appropriate if the tumor is large and involves more than one foramen. In 1983, Grillo introduced an easier and faster technique [7]. A vertical paravertebral incision extending horizontally to the tip of the scapula is performed. Through this incision a thoracotomy from the

4th or 5th intercostal space is carried out. This technique has been found to be effective in small tumors that involve one foramen. Osada et al. recommended a vertical midvertebral incision and removal of the articulation parts of the ribs of the involved level [17]. They suggested that sufficient exposure can be achieved if parts of the ribs are removed. But the tumor sizes of the cases reported are small.

Incomplete resection of dumbbell neurogenic tumors may result in serious complications. During surgery care should be taken of the spinal cord, as it can be easily injured by excessive traction or bleeding [1, 5]. These complications may result if laminectomy alone is performed as a first stage procedure [1]. If vertebral involvement is evident, an orthopedic consultation is essential, as removal of vertebral bodies may cause kyphoscoliosis afterwards.

In conclusion, our limited experience has revealed that the total removal of dumbbell tumors should be attempted. Preoperative evaluation of the tumor is extremely important. The surgical technique is dependent on the patient's clinical status and size of the tumor. In cases of large tumors with single or multiple spinal canal involvement Akwari's method seems to be appropriate, because removal of the tumor from the thorax can be difficult if the incision is too small. It is certain that one-stage removal yields excellent results and offers minimal morbidity and mortality

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