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A rare case of pulmonary neurofibroma: clinical and diagnostic evaluation and surgical treatment

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Abstract. – **The authors report a rare case of a pulmonary neurofibroma treated by surgical excision. The case report is accompanied by a review of the literature and the discussion of the diagnostic problems posed by neurogenic tumors of the thorax.**

Key Words:

Pulmonary neurofibroma, Thoracic neoplasms.

Introduction

The thoracic cavity is an anatomical site containing numerous nervous structures that may be subject to malignant transformation.

Neurogenic tumors often originate from the nervous structures located in the costovertebral recesses of the posterior mediastinum (ganglioma, ganglioneuroblastoma and neuroblastoma) while neoplastic degeneration of the vague or the phrenic nerves and of the pleuro-pulmonary nerves (neurofibroma, neurilemmoma, neurosarcoma) is less frequently observed[1] (Table I).

Pathological features of these neurogenic tumors are various, and malignancy grading is highly different among the various tumor forms. Tumor grade is the single best prognostic indicator for the development of metastases and eventual outcome.

Neurinomas, ganglioneuromas and neurofibromas can be considered scarcely invasive neoplasms; they remain mostly asymptomatic and are often an occasional finding (low-grade lesions).

Neuroblastomas are, instead, highly malignant and show an early and rapid local and distant diffusion (high-grade lesions)[2].

Pulmonary location of soft tissue tumors is rare (Table II) and symptomatology often confusing (Table III).

We herein report a case of pulmonary neurofibroma recently observed and treated in the Fourth Department of Surgery, University "La Sapienza", Rome.

Case Report

A 58 year-old male was admitted to the hospital for a dull gravative pain of the postero-lateral region of the left hemithorax, accompanied by symptomatology of dyspnea and cough. The pain increased during inspiration. On physical examination, the patient presented hypomobility and dullness of the left pulmonary base in absence of other auscultatory findings.

A postero-anterior thoracic radiography showed an ovalar opacity located postero-laterally in the inferior lobe of the left lung, surrounded by an apparently, non infiltrated lung. There were no signs of ilar or mediastinic lymph node enlargement.

A CT-scan of the thorax confirmed the presence of a mass confined by a capsule and with a policyclic contour.

On surgery the tumor resulted located on the postero-lateral surface of the inferior left

Table I.

Original tissue	Mediastinal location	Tumor type
Sympathetic ganglia	Posterior mediastinum	Ganglioma Ganglioneuroblastoma Neuroblastoma
Intercostal nerves	Thorax-trunk	Neurofibroma
Phrenic nerves		Neurilemmoma
Vagus nerve		Neurosarcoma
Paraganglia cells	Thorax-trunk	Paraganglioma

From: Sabiston DC: The Surgical Basis of Modern Surgical Practice. 14th Ed, WB Saunder, Philadelphia 1991.

pulmonary lobe; the mass was of grayish-white color, fibrous-hard consistency, and measured approximately 9 by 5 by 4 centimeters. The tumor was entirely capsulated and could easily be enucleated from the surrounding parenchyma. Pathological exam showed a neurofibroma.

Discussion

Neurogenic tumors of the thorax originating from the nervous structures of the costo-vertebral recesses are reported in literature relatively often. Less frequently these tumors originate from the pleuro-pulmonary nerves[3-13].

Symptomatology, when not absent, is often aspecific. However, according to the location of the mass, the patient may refer irritative and/or deficitary symptoms due to compression or infiltration of nervous structures as the brachial plexus, the cervical ganglium, the recurrent nerve, etc. Symptoms usually appear when the dimensions of the tumour determine distortion of mediastinic anatomycal relationships, that may lead to dyspnea, dysphagia or, less frequently, a classic mediastinic syndrome.

Neurofibromas of the thorax are often associated with analogous cutaneous lesions that consent the definition of Von Recklinghausen disease[10,14].

Diagnostic procedures in the work-up of these lesions are conventional radiology and CT-scan. This last exam is also useful for the detection of distant metastases in the malignant forms. Diagnosis must be confirmed by histological examination of the surgical sample, even though differentiation between neurinoma, neurofibroma and other morphologically similar tumors of the soft tissues may result difficult. In order to differentiate neurinomas and neurofibromas from other histotypes, S 100 protein identification may be useful[15].

Moreover, while neurinomas are usually capsulated and do not contain nervous fibres, these fibres can be found within neurofibromas confirming the policlonal origin of these tu-

Table II. Sarcomas of the soft tissues.

Site	
Lower extremity	38.9%
Retroperitoneal/intrabdominal	15.2%
Trunk	12.9%
Upper extremity	10.9%
Genitourinary	7.2%
Visceral	5.4%
Head and neck	4.8%
Other	4.6%

From: Posner ML, Brennam MF: Soft tissue sarcomas. In Holleb A, Fink DJ, Murphy GP: Clinical oncology. American Cancer Society 1991.

Table III. Presenting symptoms in patients with mediastinal mass.

Symptoms	Percentage of patients (N = 441)
Chest pain	29%
Dyspnea	22%
Cough	18%
Fever	13%
Weight loss	9%
Superior vena caval syndrome	8%

From: Sabiston DC: The Surgical Basis of Modern Surgical Practice. 14th Ed, WB Saunder, Philadelphia 1991.

mors, as demonstrated by the studies on the heterozygosis of the G6PD[16].

An accurate and precise diagnosis is often useful also for surgical purposes since neurinomas can be resected preserving the nervous structure from which they originate while this is often impossible in the case of neurofibromas.

Conclusion

Pleuro-pulmonary tumors originating from nervous structures cause considerable diagnostic problems. Differential diagnosis between benign and malignant masses is often difficult, considering the rarity of this localization (0.2 to 4%)[10]. Diagnosis must be confirmed by histologic examination in all cases.

As for surgical treatment of neurofibroma, it should be as radical as possible in consideration of the high incidence of recurrence and of the possibility of malignant degeneration particularly in cases of Von Recklinghausesn neurofibromatosis[17,18].

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