CASE REPORT
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Is Ganglioneuroma Provoked by Scoliosis? Concordant of Mediastinal Ganglioneuroma and Scoliosis in a Case with Advanced Age

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Abstract
Mediastinal ganglioneuromas are usually diagnosed in early ages. In advanced ages, its presentation is very rare, especially in the ages older than 40 years. We present a 51-year- old woman case with mediastinal ganglioneuroma associated with scoliosis. Ganglioneuroma may provoke scoliosis as it occurred in childhood, or scoliosis may provoke ganglioneuroma development. However, it is possible to claim none of them in our case. At least, co-existence of neurogenic tumors such as ganglioneuroma should be considered in the presence of progressive and persistence back pain in a case with scoliosis.

Keywords: Ganglioneuroma, scoliosis, advanced age, surgery.

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INTRODUCTION

Ganglioneuromas originate from ganglion cell of the sympathetic nervous system or, infrequently, from sympathetic nerves or other peripheral nerves. They are slow growing tumors. They are usually located anterior-posterior mediastinum and retroperitoneal tissues but may also be located neck, adrenal glands and guts [1, 2]. Mediastinal ganglioneuromas, which are usually encountered in childhood, are mostly diagnosed benign tumors in the children after 3-4 years old of childhood [3]. Bulmer, reported on three children with ganglion tumors and concordant scoliosis resulting from a neuroblastoma, ganglioneuroma and ganglioblastoma. Consequently, a scoliosis may be provoked by the tumor [4]. In addition, these tumors are considered to be responsible in etiology of neuromuscular scoliosis of childhood [5]. Although it is mostly diagnosed in childhood, we herein present a 51-year-old female case, who had known scoliosis for many years, was admitted with a history of progressive back pain for a year; and diagnosed as mediastinal ganglioneuroma which has extradural, intraforaminal and paraspinal localization. The tumor was excised surgically and no problem was found in two years' follow up period.

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CASE REPORT

A 51-year-old female was admitted with a history of progressive back pain for the last year. She had a scoliosis history for 20 years. Her back pain complaint has been attributed to her scoliosis. In physical examination, no abnormal finding was found except for elevated right shoulder and right scapulae. Neurological examination was also normal. There were thoracic scoliosis and right paraspinal mass in chest X-ray (Figure-1a). Computerized tomography (CT) and magnetic resonance imaging (MRI) of thorax showed a mass lesion on the convex right side of scoliosis between T4-T7 (Figure-1b, c, d). Her scoliosis was dextro-scoliotic and Cobb angle of scoliosis was measured as 37 degree. In the laboratory exam, no abnormality was found.

In the evaluation of thorax CT and MRI findings, the mass was well-circumscribed encapsulated and in benign nature. Surgery was planned because it was considered as neurogenic tumor. Right postero-lateral thoracotomy was performed. A yellowish elastic mass lesion originating from fourth foramina intervertebralis and protruding into the thorax and spinal channel, which has no capsule and 8x7x3 in diameter, was meticulously excised at the level of duramater (Figure-2a). Lesion could easily be dissected from adjacent tissues and no destruction observed on the adjacent rib and vertebra.

In the histopathological examination of the specimen, spindle cell and mature ganglion cell proliferation was seen (Figure-2b). No findings of mitosis and neuroblastomatosis were detected. It was reported as ganglioneuroma. No complication was occurred in the post-operative period. She was discharged on day 9 after operation. The patient had no complaint or finding of recurrence in the two years follow-up period.

DISCUSSION

Neurogenic tumors are relatively uncommon, though they comprise 15-25 % of all primary mediastinal tumors.

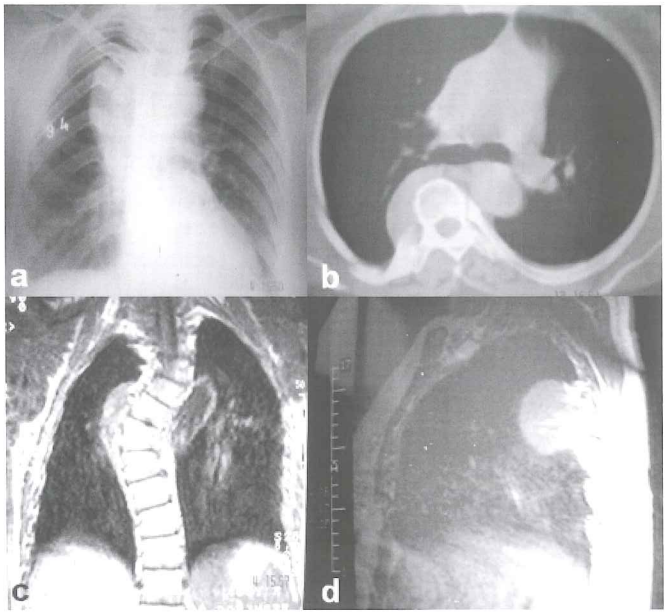

Figure 1: (a) P-A Chest X-ray, (b) axial computed tomography section, (c) coronal MRI section and (d) sagittal MRI section demonstrated a mass lesion at right upper mediastinum

Intrathoracic neurogenic tumors are found almost exclusively in the posterior mediastinum around the paravertebral area from the intercostal and sympathetic nerves, and only rarely are represented as an intrapulmonary mass. Ganglioneuromas forms of 35% of neurogenic tumors located intrathoracically. Of them, 65 % are diagnosed in childhood [6].

Most of the ganglioneuroma cases are asymptomatic (98%). However, some patients who had increased diameter of the tumor may be presented with cough, neck swelling, dyspnea, chest pain, Horner's syndrome and back pain [1, 2, 6]. Approximately 10% of neurogenic tumors located posterior mediastinum may extend to spinal channel and cause neurologic symptoms due to compression. These tumors are called as dumb-bell or hour-glass tumors. Dumb-bell tumors cause symptoms due to compression of spinal chord or/and nerve roots [7].

Macroscopically, ganglioneuromas are well-circumscribed, uncapsulated, benign tumors with a firm gray to yellow surface. Microscopically, the tumor is composed of mature ganglion cells admixed with spindle cells of Schwannian origin and nerve fibers. Mature ganglion cells consist of a large nucleus with nucleoli and a granular, basophilic cytoplasm, containing basophilic and rarely, eosinophilic Nissl granules. Malignant form of ganglioneuroma is ganglioneuroblastoma. One fourth of the tumors demonstrate calcification and rib or vertebral erosion that is subtle but often apparent. Erosion is more severe in ganglioneuroblastoma [8].

The cross-sectional and multiplanar capacity of CT and magnetic resonance imaging makes them the choice modalities for obtaining images of the ganglioneuroma. CT demonstrates tumors of homogenous or heterogeneous attenuation. Magnetic resonance imaging demonstrates homogenously intermediate signal intensity on T1-weighted images and heterogenously high signal intensity on T2-weighted images. T1- and T2-weighted images have a whorled appearance caused by curvilinear or nodular bands of low intensity [9].

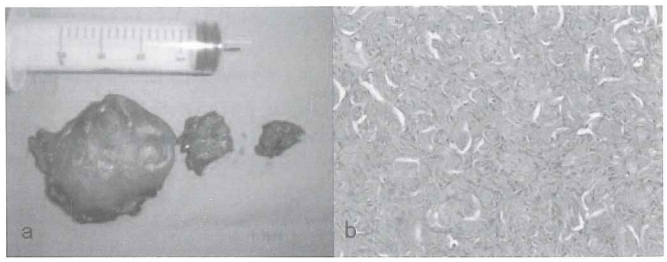

Figure 2: (a) The surgical specimen after removal from the right thoracic spine, (b) The basic pattern of spindle cell proliferation in this tumor and intralesional presence of ganglion cells. (H&Ex100)

If a ganglioneuroma is confirmed by pathology, surgical excision alone is curative. Postoperative adjuvant radiotherapy is controversial. In the tumors of "hourglass" that has spinal extension, it should be done bilateral laminectomy and fusion with bone graft in the first intervention and then excision via thoracotomy [2].

As it was occurred in our case, the presence of persistent and progressive back pain in a patient with scoliosis should remind co-existence of neurogenic tumors such as ganglioneuroma and should be taken into consideration in differential diagnosis of back pain. When scoliosis and ganglioneuroma co-existence diagnosed in childhood, it is considered that ganglioneuroma provokes scoliosis development. However, in adult ages, in contrast to childhood, such relationship may not be completely true. To answer the question that "does scoliosis may provoke ganglioneuroma development?" needs further larger studies investigating ganglioneuroma presence in the population with scoliosis.

Scoliosis may provoke ganglioneuroma or ganglioneuroma may provoke scoliosis in adult age as occurred in childhood. Both of them may be possible, but we can not claim such relationship with a case. However, at least, we may conclude that in case of presence of progressive and persistence of back pain in a patient with scoliosis should remind coincidence of neurogenic tumors.

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