BALKAN MEDICAL JOURNAL
THE OFFICIAL JOURNAL OF TRAKYA UNIVERSITY FACULTY OF MEDICINE

Balkan Med J 2013; 30: 126-8 • DOI: 10.5152/balkanmedj.2012.099
Available at www.balkanmedicaljournal.org

Case Report

A Case of Posterior Mediastinal Ganglioneuroma: The Importance of Preoperative Multiplanar Radiological Imaging

Betül Kızıldağ[1], Timuçin Alar[2], Ozan Karatağ[1], Şule Koşar[1], Tarık Akman[3], Murat Coşar[3]

[1]*Department of Radiology, Faculty of Medicine, Çanakkale Onsekiz Mart University, Çanakkale, Turkey*
[2]*Department of Thoracic Surgery, Faculty of Medicine, Çanakkale Onsekiz Mart University, Çanakkale, Turkey*
[3]*Department of Neurosurgery, Faculty of Medicine, Çanakkale Onsekiz Mart University, Çanakkale, Turkey*

ABSTRACT

Ganglioneuromas are mostly seen in adolescents and young adults and they are neurogenic tumors originating from sympathetic ganglions with a benign histology. Although ganglioneuromas are benign, the treatment is surgical as they can cause pain or compression symptoms, can be locally aggressive and can lead to cord compression. We present a young adult female with a ganglioneuroma of the right posterior mediastinum who presented with lower back pain, together with the clinical features, Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) findings, differential diagnosis tips and their contribution to surgical planning.

Key Word: Ganglioneuroma, neurogenic tumor, posterior mediastinum

Received: *06.06.2012* ***Accepted:*** *15.10.2012*

Introduction

Ganglioneuromas are mostly seen in adolescents and young adults and they are neurogenic tumors originating from sympathetic ganglions with a benign histology (1). They can originate from any location in the paravertebral sympathetic plexus or more rarely from the adrenal medulla. They are usually asymptomatic but can lead to compression symptoms according to tumor size and rarely to systemic symptoms (2). The treatment is complete surgical resection (3). It is essential to characterize and fully localize the tumor and to evaluate its intraspinal extension preoperatively with computed tomography (CT) and magnetic resonance imaging (MRI) to direct the surgical approach (4). We present a young adult female with a ganglioneuroma of the right posterior mediastinum who presented with lower back pain, together with the clinical features, CT and MRI findings, differential diagnosis tips and their contribution to surgical planning.

Case Report

An 19-year-old female patient presented at the pulmonary disease outpatients department with back pain and shortness of breath for the last year. There were no additional systemic findings or any history of smoking and the physical examination revealed nothing of significance except for the pectus excavatum deformity for which she had undergone surgery when 4 years old. The hematological and biochemical laboratory tests were within normal limits and the serological evaluation results for Echinococcus granulosus were negative. The PA chest X-ray, the first diagnostic evaluation, showed an oval, well-delineated homogeneous radioopacity with sharp edges and vertical extension into the T9-L1 vertebral corpus at the paravertebral area of the right hemitorax lower zone (Figure 1A). The differential diagnosis process for the posterior mediastinal solid mass lesion continued with thoracic CT following intravenous (IV) contrast agent administration that showed an oval 9x6x3.5 cm mass in the right paravertebral region at the T9-L1 level with homogenous low density (+18 HU) that did not show marked enhancement, did not contain calcification, had benign borders and extended over the ribs as an extrapleural soft tissue mass (Figure 1B). We obtained sagittal and coronal multiplanar reformatted (MRP) images with a computer from the axial CT images obtained through thin section scanning to eliminate the possibility of retroperitoneal extension and to determine the exact location of the mass. Evaluation of the MPR images showed that the mass was within the thorax with no retroperitoneal extension and that it had pushed the right hemidiaphragm crus caudally. The mass had not caused any erosion in the adjacent vertebrae or the ribs. The patient had no mediastinal lymphadenopathy. The mass was found to extend to the T10-11 right neural foramen. The preliminary diagnosis with these findings was a neurogenic mass and the patient underwent a thoracal-lumbar spinal MR following IV contrast administration to show possible spinal extension. The mass was hypointense on T1-weighted (T1W) images, and contained a hypointense periphery and hyperintense central section on T2-weighted (T2W) sequences with

Address for Correspondence: *Dr. Timuçin Alar, Department of Thoracic Surgery, Faculty of Medicine, Çanakkale Onsekiz Mart University, Çanakkale, Turkey Phone: +90 286 218 00 18 Fax: +90 286 218 38 06 e-mail: timalar@comu.edu.tr*

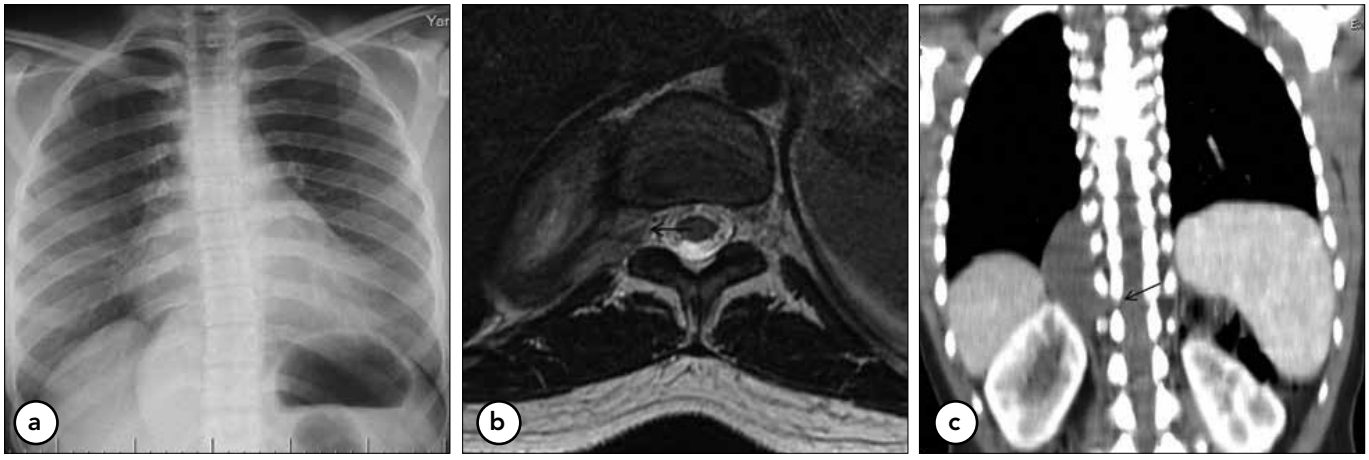

Figure 1. A. PA chest x-ray shows a paravertebral well-delineated radio-opaque mass in the lower zone of the right hemithorax. B. Axial T2-weighted MR images shows extension of the mass to the right T10-11 neural foramen (arrow). C. Coronal reformatted (MPR) enhanced CT section reveals extrapleural low-density soft tissue mass without marked enhancement showing neural foramen extension (arrow)

or without lipid suppression (Figure 1C). Following administration of the contrast agent with IV gadolinium, the T1W sections showed that the mass with diffuse contrast enhancement and regular borders extended to the T10-11 neural foramen on the right (Figure 2). However we did not detect marked extension of the mass within the spinal canal. Transthoracic biopsy was performed under the guidance of ultrasonography (US) using an 18-Gauge semiautomatic cutting needle. Preliminary histopathological evaluation reported that the tumor contained neural elements and fibroadipose tissue. Two-stage surgery was planned by the Thoracic Surgery and Neurosurgery departments. In the first stage, the Thoracic Surgery team found during videothoracoscopic (VATS) exploration that the mass was in the thoracic location and extended inferiorly towards the diaphragm right crus as shown with radiological methods. The mass was grey-white, encapsulated and densely adherent to the vertebral column. Following its dissection from the paravertebral sulcus through a right posterolateral thoracotomy, the Neurosurgery team resected the mass from the T11 nerve root from which it had originated. After one month, the same team performed a complete excision from the T11 neural radix from which the tumor had originated with a T10-11 right hemilaminectomy and facetectomy approach in the second stage. The patient's severe pains disappeared and she was discharged without complications. The histopathological evaluation of the obtained material was reported as ''S100 positive lesions consisting of cells with spindle-length extensions between striated muscle bundles and mature fat cells", typical for ganglioneuroma.

Discussion

Neurogenic tumors make up 75% of all posterior mediastinal tumors. The intercostal nerves or ganglion cells are the origin of 95% of neurogenic tumors (5). Ganglineuromas are somewhat histologically different from schwannomas that originate from sympathetic ganglion cells, neurofibromas and neurofibrosarcomas that originate from peripheral nerves and paragangliomas, pheochromocytomas and other neurogenic tumors that are of paraganglionic origin, together with ganglioneuroblastomas and neuroblastomas. Ganglioneuromas histologically consist of more benign and well-differentiated cells compared to ganglioneuroblastomas and neuroblastomas. These tumors contain spindle cells and a collagen stroma besides mature ganglion cells. They are typically well-delineated tumors with a fibrous capsule and can be seen anywhere that sympathetic tissue is present, most commonly in the posterior mediastinum and retroperitoneum and rarely in the neck, adrenal gland and pelvis (6-8). Ganglioneuromas are the most common posterior mediastinal mass in adolescents and young adults and are usually asymptomatic (1). They can cause nonspecific chest pain or symptoms due to compression of the tracheobronchial tree (9). Spinal canal extension can lead to cord compression and neurological findings. They can extend to the adjacent intervertebral foramen and from here to the spinal canal, giving them a dumbbell or hourglass shape (2). Our case was young, as reported in the literature, and the tumor extended to the neural foramen but did not cause any neurological symptoms. However, the lesion caused severe back pain in our patient and did not respond to treatment with anti-inflammatories and opioids in the preoperative stage.

Ganglineuromas are generally found incidentally on chest X-rays obtained for other reasons (1). The plain X-ray typically shows, as in our case, a well-delineated opacity on the anterolateral edge of the vertebra that continues longitudinally on 3-5 vertebral segments (10).

Kato et al. (11) have defined the imaging features of 14 pathologically proven ganglioneuromas on CT and MRI and reported that ganglineuromas were typically well-delineated lesions that had oval or lobulated contours and showed craniocaudal longitudinal growth with quite low attenuation on CT and seen as a hypointense lesion on T1-weighted and hyperintense lesions on T2-weighted sequences on MRI. The same study reported calcification in 38%, a vortex appearance in 43%, a fat component in 29% and a tail-shaped edge in 14% on CT together with weak enhancement on dynamic CT in 46% and enhancement on MR in 100%. Our case similarly showed a quite well delineated homogenous lesion with low attenuation with a post-contrast density of +18 HU on CT.

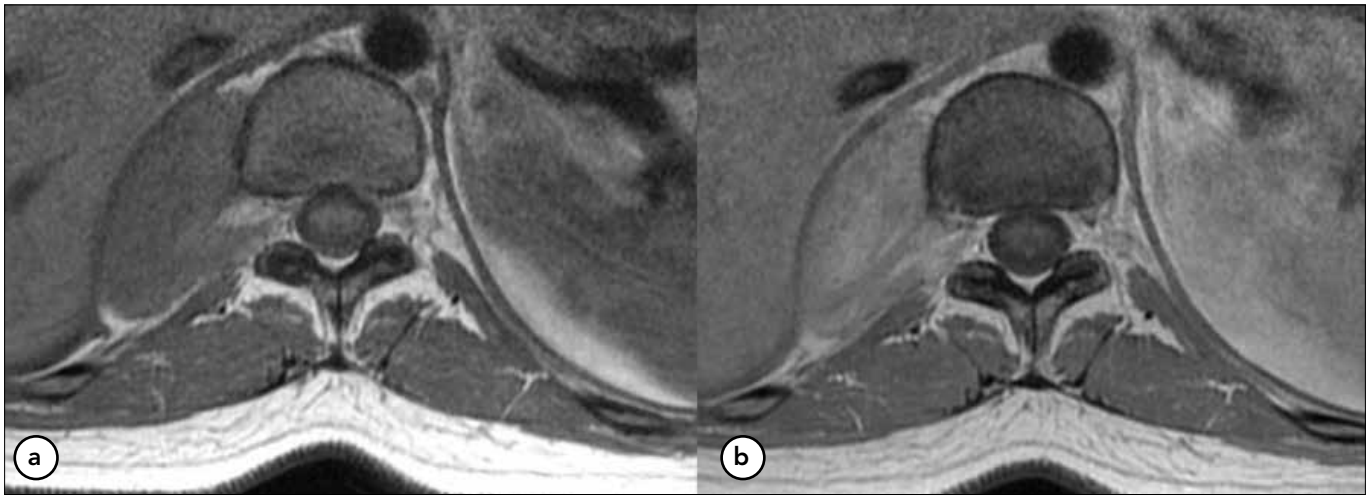

Figure 2. On precontrast (2A) and postcontrast (2B) T1-weighted MR images, prominent contrast enhancement of the mass extending towards the neural foramen is clearly visible

However we did not find calcification, fat content density or marked enhancement on CT. The MR showed a homogenous hypointense signal on T1-weighted and a peripheral hypointense and central hyperintense appearance on T2-weighted images but there was no vortex appearance.

Neural foramen extension limits the differential diagnosis of posterior mediastinal masses to ganglioneuroma and nerve sheath tumors (3). Nerve sheath tumors make up 40-65% of the neurogenic masses of the posterior mediastinum. These tumors are usually of intercostal nerve origin and spherical in shape. They can cause erosion of adjacent bones and can extend into the intervertebral foramen in 10% (12). The CT density is markedly low and the MR appearance is similar to ganglineuroma. Ganglioneuromas are seen at younger ages than nerve sheath tumors (5).

Although ganglioneuromas are benign, the treatment is surgical as they can cause pain or compression symptoms, can be locally aggressive and can lead to cord compression (9). Our case was symptomatic and the mass extended to the neural foramen so a decision was made for surgical excision with a multidisciplinary approach. On CT, the MPR imaging feature was used and on MR the axial, coronal and sagittal images were evaluated enabling a multiplanar evaluation revealing the exact localization of the lesion and eliminating the possibility of retroperitoneal extension. The association of the mass with the diaphragm crus and the neural foramen extension were shown correctly and contributed significantly to planning the surgery.

Ganglioneuromas are benign posterior mediastinal tumors requiring complete surgical excision as they can cause severe pain and compression symptoms. Knowing the patient age and clinical course and typical imaging findings will increase the percentage of correct diagnoses. Being able to show the location and extension of the tumor in all planes (axial, coronal, sagittal) radiologically will also contribute greatly to the multidisciplinary approach to the decision and surgical planning.

Informed Consent: *Written informed consent was obtained from patients who participated in this study.*

Peer-review: *Externally peer-reviewed.*

Author contributions: *Concept - T.A., O.K.; Design - B.K., T.A.; Supervision - M.C., T.A.; Resource - B.K., Ş.K.; Materials - T.A., O.K.; Data Collection&/or Processing - B.K., T.A.; Analysis&/or Interpretation - O.K., T.A.; Literature Search - B.K., T.A., Ş.K.; Writing - B.K., T.A., O.K.; Critical Reviews - M.C., Ş.K., T.A.*

Conflict of Interest: *No conflict of interest was declared by the authors.*

Financial Disclosure: *No financial disclosure was declared by the authors.*

References

1. Lee JY, Lee KS, Han J, Yoon HK, Kim TS, Han BK, et al. Spectrum of neurogenic tumors in the thorax:CT and pathologic findings. J Comput Assist Tomogr 1999;23:399-406. [CrossRef]
2. Geoerger B, Hero B, Harms D, Grebe J, Scheidhauer K, Berthold F. Metabolic activity and clinical features of primary ganglioneuromas. Cancer 2001;91:1905-13. [CrossRef]
3. Strollo DC, Rosado-de-Christenson ML, Jett JR. Primary mediastinal tumors. Part II:Tumors of the middle and posterior mediastinum. Chest 1997;112:1344-57. [CrossRef]
4. Rha SE, Byun JY, Jung SE, Chun HJ, Lee HG, Lee JM. Neurogenic tumors in the abdomen:tumor types and imaging characteristics. Radiographics 2003;23:29-43. [CrossRef]
5. Kumar A, Kumar S, Aggarwal S, Khilnani GC. Thoracoscopy:the preferred approach for the resection of selected posterior mediastinal tumors. J Laparoendosc Adv Surg Tech A 2002;12:345-53. [CrossRef]
6. Shimada H, Ambros IM, Dehner LP, Hata J, Joshi VV, Roald B, et al. The international neuroblastoma pathology classification (the Shimada system). Cancer 1999;86:364-72. [CrossRef]
7. Shields TW, Reynolds M. Neurogenic tumors of the thorax. Surg Clin North Am 1988;68:645-68.
8. Enzinger FM, Weiss SW. Pirimitive neuroectodermal tumors and related lesions. In:Enzinger FM, Weiss SW, editors. Soft tissue tumors, 3rd edn. St. Louis, MO:Mosby, 1995;929-64.
9. Forsythe A, Volpe J, Muller R. Posterior mediastinal ganglioneuroma. Radiographics 2004;24:594-7. [CrossRef]
10. Ko SM, Keum DY, Kang YN. Posterior mediastinal dumbbell ganglioneuroma with fatty replacement. Br J Radiol 2007;80:e238-40. [CrossRef]
11. Kato M, Hara M, Ozawa Y, Shimizu S, Shibamato Y. Computed tomography and magnetic resonance imaging features of posterior mediastinal ganglioneuroma. J Thorac Imaging 2012;27:100-6. [CrossRef]
12. Aughenbaugh GL. Thoracic manifestations of neurocutaneous diseases. Radiol Clin North Am 1984;22:741-56.