Giant dumbbell tumor of the posterior mediastinum

Džinovski dumbbell tumor zadnjeg medijastinuma

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Abstract

Background. Giant dumbbell-shaped tumors are very rare and characterized by intra- and extraspinal propagation of different dimensions. In thoracic localization, invasive growth can lead up to rib and vertebra erosion. Case report. We presented a 54-year-old woman with a giant dumbbell schwannoma in the posterior mediastinum. The tumor was removed by the posterior approach with hemilaminectomy and costotransversectomy. By microscopic examination the diagnosis of benign schwannoma was made. A year after the surgery, the patient was without neurological deficiency and without radiological signs of illness relapse. Conclusion. Treatment of dumbbell schwannoma is surgical, dilemmas of the optimal surgical approach. When it is thoracally located the posterior approach with hemilaminectomy and costotransversectomy is safe and effective for its removal.

Key words: neurilemmoma; mediastinal neoplasms; spine; diagnosis; immunohistochemistry; neurosurgical procedures.

Case report

A 54-year old, female patient, admitted because of chest pains, propagating to the spine for the last two years, had more intensive pains in the past two months without neurological deficiency.

Introduction

Neurogenic tumors make even 12–39% of all tumors in mediastinum, and even more than 75% of all tumors in the posterior mediastinum. Most often neurogenic tumors of mediastinum are schwannoma and in 70–80% they represent benign, slow-growing neoplasms.

Schwannomas arise from Schwann cells of nerve sheath. They are solitary in the majority of cases; multiple schwannomas represent a component of von Recklinghausen’s disease. They appear along peripheral and cranial nerves, rarely in the area of spinal nerve roots. Spinal schwannomas often capture sensory nerve roots; most commonly located intradurally in lumbal region. A mass similar to hourglass or dumbbell, forms when interspinal schwannoma spreads through the intervertebral opening. A schwannoma with extradermal extension larger than 2.5 cm that causes rib and vertebra erosion, is called a giant dumbbell tumor. Schwannomas account for about 90% of all dumbbell tumors. Rarely, a dumbbell mass is made by malignant peripheral nerve sheath tumors, hydatid cyst or chondrosarcoma.

A 54-year old, female patient, admitted because of chest pains, propagating to the spine for the last two years, had more intensive pains in the past two months without neurological deficiency.
After chest radiography (RTG) (Figure 1), computerized tomography (CT) (Figure 2) and magnetic resonance imaging (MRI) (Figure 3), the diagnosis of tumor of the posterior mediastinum with infiltration of the chest wall and thoracic vertebra (Th) was made. Bronchoscopy and spirometry results were inside the reference limits. Skeletal scintigraphy showed a physiological distribution of radiopharmaceuticals.

Fig. 1 – Radiography of the chest (a: posteroanterior view, b: lateral view): in projection of the right cardiophrenic angle of half-round, limited, soft-tissue shadow with diameter of about 60 mm, partly in superposition with heart shadow (white arrows)

Fig. 2 – Computerized tomography of the chest: in the posterior mediastinum of saddle-ellipsoid change (white arrow), with expansive growth through intervertebral opening in the spinal canal with destruction of lateral mass and part of the body of the thoracic 11 vertebral (black arrow)

Fig. 3 – Magnetic resonance imaging (a: cross section, b: sagittal section). Soft-tissue tumor formation localized paravertebrally in the posterior mediastinum, with intraspinal spreading and paravertebral propagation through intervertebral foramen of the thoracic 11 vertebral (white arrows)

A precise preoperative localization of the tumor was confirmed by “metal comb” at the levels from Th10 to Th12 and targeted RTG. Posterior hemilaminectomy and costotransversectomy made the thoracic retropleural component of the tumor (Figure 4). The tumor was reduced and removed to the posterior chest wall. Hemilaminectomy of Th10 and Th11 opened spinal canal, so the intraspinal tumor component became visualized at the Th11 level, extradurally condensed medulla spinalis. Transversal endings, joint facet, as well as a part of the body Th11, were removed, ena-

Fig. 4 – Intraoperative picture of the tumor: partially encapsulated, grey-yellowish, hard, size 12 x 10 cm (white arrow) that infiltrates posterior wall of thoracic cavity

bling a complete extirpation of intraspinal and foramina tu-
мор component.

A postoperative period passed without complications,
except for the pain in the back. The patient was dismissed
from the hospital three weeks after the surgery. The patient
had sustained pain three more months. A year after the sur-
gery, CT and MRI showed a stationary result, without local
relapses.

More tissue samples, of irregular shape, diameter of
30–60 mm, total weight of 210 g, grey-yellowish colour,
mostly smooth and glitter surface were pathohistologically
examined. Microscopic analysis showed an image of clearly
limited, encapsulated spindle cell tumor mainly. Multiple
spindle cells were grouped in shorter and longer fascicles,
placed in different directions. Cellularity was mostly uni-
form, and only rarely there were zones of hypercellularity
present, with nucleus that build up palisades and fields of
myxoid degeneration (Antoni A and Antoni B components).
Cytological atypia, mitosis and necrosis were not found. The
tumor was sporadically infiltrated by foamy macrophages,
and focally lymphocytes, and there were also zones of cystic
degeneration. Perivascular hyalinization was also present.
Immunohistochemical examination showed that tumor cells
diffusely exprimate vimentin, S-100 protein and glial fibril-
lary acidic protein (GFAP), sporadically positive for cyto-
keratin, and negative for epithelial membrane antigen,
smooth muscle actin (α-SMA), HMB45, Myo D1, CD 117, CD 68, desmin and CD34. There was a focal positive value
of macrophages on CD 68 and blood vessels on CD 34 and
α-SMA. Application of Ki-67 antibodies showed a vary low
index, lower than 1%. According to microscopic analysis,
the diagnosis of benign schwannoma was made (Figure 5).

Fig. 5 – Histological features of a schwannoma:
a) the tumor is composed of spindle cells grouped in shorter and longer fascicles (hematoxylin and eosin staining technique, $\times 100$); b) tu-
mor's cells express vimentin ($\times 200$); c) S-100 protein ($\times 200$); d) glial fibrillary acidic protein ($\times 200$); e) weakly, focally positive for cyto-
keratin ($\times 200$); f) the tumor cells are negative for smooth muscle actin ($\times 200$); g) the Ki67 proliferation index is very low, less than 1%
($\times 400$); h) macrophages are positive for CD 68 ($\times 400$)
Discussion

Thoracic dumbbell schwannomas are very rare. They mainly possess paravertebral extension, and some are separated with their enormous size, extension in different directions and invasive characteristics. In spinal schwannomas, 70–80% of all tumors are purely intradural tumors, 10–20% have both intra- and extradural components, and 10–20% are only extradurally localized. In the forties of the last century Eden classified neurogenic tumors of posterior mediastinum according to their localization and relation to spinal canal and paravertebral structures in four types: intra- and extradural, intradural and paravertebral, foraminal and paravertebral. Sridhar et al. suggest classification system for spinal schwannomas into five types. Giant tumors involve those that cover two or more than two vertebrae (type II), all whose extraspinal extension is over 2.5 cm (type IV) and those that perform erosion of vertebrae and spread backwards and laterally in myofascial structures (giant invasive schwannoma, type V).

Giant dumbbell invasive tumors represent diagnostic and therapeutic problem. They have been asymptomatic for a long time. Local and segmental reticular pains are initial symptoms. Compression syndrome of spinal cord is a consequence of intraspinal tumor component growth. They can cause intradural hemorrhage. Although very rarely, young people are exposed to an increased risk of malignant transformation.

The diagnosis of these tumors require application of RTG, CT and MRI because of vertebral erosion possibility, spreading in neural foramen or direct extension of tumor into the spinal canal. Currently, MRI is the most reliable method to estimate neuroforaminal tumor spreading. Knowledge of a possible neuroforaminal and intraspinal tumor propagation can define surgical approach, which considerably decrease complications caused by intraoperative spinal cord injury and hemorrhage inside the spinal canal. With posterior mediastinum tumors that are located low, spinal angiography can make reliable diagnosis, identifying Adamkiewicz artery. Its injury can result in the damage to the spinal cord. Spinal angiography represents a procedure followed by enormous risk and its application is often questioned. In the presented case RTG, CT and MRI enabled a precise localization of giant dumbbell tumor of posterior mediastinum that can be extradural and paravertebral according to Eden’s classification, and according to Sridhar et al. classification system with extraspinal extension over 2.5 cm, erosion of vertebra and invasion of the chest.

A complete excision of the giant schwannoma is therapeutic gold standard. Incomplete removal of this tumor is associated with an increased risk of relapse. Relapses is confirmed in 10.2% operated people in 4.3 years on average. Giant dumbbell invasive schwannoma extension represents a surgical problem in the sense of access, resectability and stabilization of the spine, and the most common approaches are thoracal (anterior) and spinal (posterior). Postsurgical complications are a consequence of the spinal cord injury with consequential myelopathy and dura injury with liquefaction in pleural space. A combined thoracal and neurosurgical approach allows most flexibility and certainty in treatment of such tumors. In the presented case, the tumor size caused an application of posterior approach with hemilaminectomy, although Kanemoto suggests hemilaminectomy without costotransversectomy.

Histopathologically, the diagnosis of schwannoma is confirmed on the basis of the present Antoni A and Antoni B zones, that represent special microscopic architectural shapes typical for this type of tumor. Nucleus palisades were present in a couple of patterns, and Varocay bodies were not noticed. Nucleus palisades inside Antoni A region are more often seen in spinal than in intracranial schwannoma, while they are absent in schwannoma of VIII cranial nerve. Cystic degeneration and hyaline degeneration of the blood vessels walls have often been seen, as well as the presence of foamy macrophage and lymphocyte aggregates. Immuno-histochemically, neuronal differentiation has been confirmed with diffuse and strong positivity with S-100 protein and GFAP. Expression of cytokeratin in schwannoma has been described in literature. Ki 67 exprimated only in some tumor cells, which confirms that mitosis in schwannoma is absent or extremely rare.

Conclusion

Giant dumbbell invasive schwannomas are uncommon lesions distinct from other spinal schwannomas. Because of their locally invasive nature and possible extension in all directions, a careful preoperative planning of a surgical approach and pathohistological analysis are very important. Posterior surgical approach with costotransversectomy and hemilaminectomy is safe and effective for resection of dumbbell schwannomas.

REFERENCES


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