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Metastatic Pheochromocytoma of the Thoracic Spinal Extradural Space Case Report and Review of the Literature

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In this case report, a thoracic extradural metastatic pheochromocytoma without bony invasion, is presented. The disease which has been identified with its symptoms, bio-chemical features, radiological appearance, histological diagnosis has been discussed in comparison with malignant pheochromocytoma metastases in the literature. The origin of this tumor is the adrenal glands. Our review of the literature shows that tumor has primary metastasis in bony structures of the spine and then demonstrates secondary intraspinal invasion. This is the first case report of an epidural metastasis from malignant pheochromocytoma without a bony invasion.

The term “pheochromocytoma” has been classically used to describe an adrenal functioning (epinephrine -and norepinephrine- secreting) paraganglioma, but it has also been applied to extra-adrenal functioning paraganglionomas (1,5,7,9,13,21,22). The pheochromocytoma usually is benign, but approximately 10 per cent are malignant (1,5,7,19,22). The incidence of malignant pheochromocytoma varies between 2.5 % and 13 % according to literature (1,5,7,19,22). The malignant diagnosis occurs according to whether metastasis or invasion exists. In 7 % of the cases metastasis occurs in more than one organs (2,26). Malignant tumor usually metastasizes to bone, lung, kidney and lymph nodules, and rarely metastasizes to the brain, skin, prostate, urinary bladder. Metastasis is seen in cervical (10,11,14,17), thoracic (20,21,23,24), lumbar vertebra (11,18,23,25), sacrum (18), bony structures and spine adjacent tissues (sympathetic chain, paraaortic lymph nodules). A case of malignant pheochromocytoma arising from the adrenal glands is presented. This tumor is metastatic pheochromocytoma. This case has not been found in the literature as malignant pheochromocytoma since it is thoracic extradural extramedullary intraspinal metastasis. In our review of the literature, tumor shows primary metastasis in the bony structures of the spine and then shows secondary intraspinal invasion, but our case has an epidural metastasis without a bony invasion.

CASE REPORT

This 47 year old male patient has hypertension for 10 years. He has suffered from headaches, palpitation, flushing, shivers, urinary incontinence, anxiety attacks. He had had pain and numbness in the last five months when paraparesis developed. Findings of neurological examination include paraparesis, hyperactive patella and achille reflexes, positive Babinsky and clonus on the right side, hypoesthesia under the TH8 level. Routine laboratory results showed no abnormality. Elevated level of blood vanillylmandelic acid was discovered. Plasma norepinephrine levels also have been elevated. A 33x 27 mm mass on the

left surrenal and 26x27 mm on the righth surrenal have been found in abdominal computerized tomography (CT). A 3x1.2x1.5 cm on measuring extrameduller extradural mass has been found in the spinal channel in the thoraco-lumbar magnetic resonance (MR) imaging, TH8 level placed posteriorly (Figure 1). The lesion was isointense in T1,

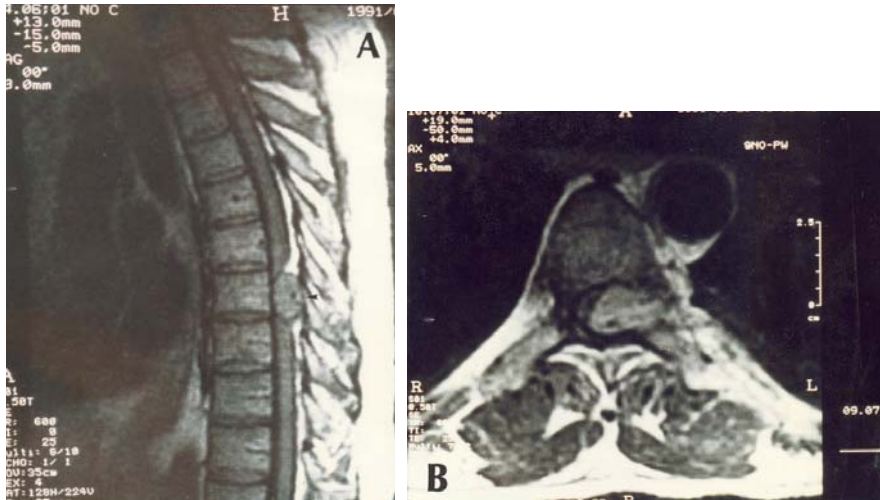


Figure 1. A, sagittal and B, axial MRI; 3x1.2x1.5 cm on measuring extrameduller extradural mass has been in the spinal channel in the thoraco-lumbar MRI, Th8 level placed posteriorly. the lesion was isointense in T1 hyperintense in T2 images .

hyperintense in T2 images. After the administration of contrast material, the limits of the nodular mass lesion was seen in the thoraco-lumbar MRI. After performing a TH8, TH9 total laminectomy, the tumor was totally removed. Histopathological examination with light microscope of lesion revealed a neoplasm with cells arranged in sheets and clusters. Cells were noted to contain pleomorphic nuclei and cell clusters were surrounded by a labyrinth of capillaries. Pathological diagnosis was pheochromocytoma (Figure 2). Paraparesis and I.

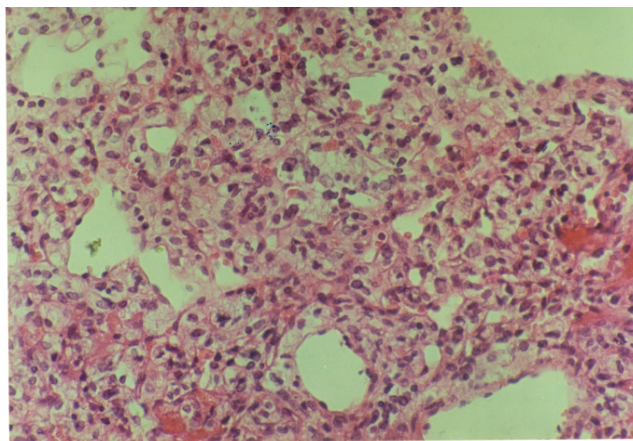


Figure 2. Histopathological examination with light microscope of lesion revealed a neoplasm with cells arranged in sheets and clusters. Cells contained pleomorphic nuclei, and cell clusters were surrounded by a labyrinth of capillaries.

METASTATIC PHEOCHROMOCYTOMA OF THE THORACIC SPINE

motor neuron findings were improved postoperatively. Urology and general surgery consultations were given in postoperative days. In the abdominal CT, bilateral surrenal mass diagnosis and peripheral sympathetic findings continued. Whole body scintigraphy was requested. There were no significant pathological incidents. Hypertension and vasomotor indications continued. He was operated by general surgery. Laparotomy was realized by bilateral subcoastal incision and then the restricted capsulated mass in the bilateral surrenal were totally removed. There was no other masses than surrenal in the abdomen. The patient's signs and symptoms improved. MR image obtained 2 years postoperatively was normal (Figure 3). There were no recurrences and no abnormality which revolved in 2 years of follow- up.

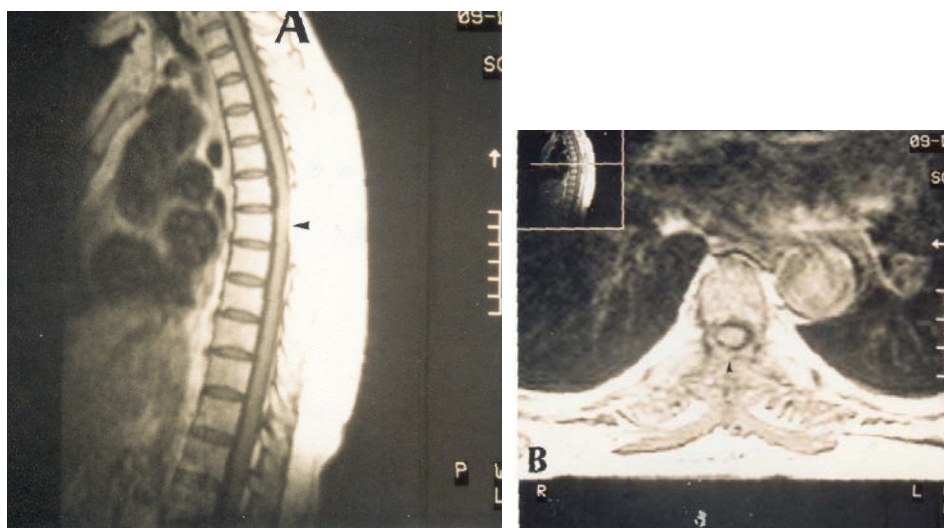


Figure 3. Postoperative sagittal (A) and axial (B) MRI scans.

DISCUSSION

The incidence of pheochromocytoma is 0.001 in the general population (18). The incidence of malignant pheochromocytoma varies between 2.5% and 13% according to literature (1,5,7,19,22). The malignant diagnosis is noted according to whether metastasis or invasion exists. In 7 % of the cases metastasis occurred in more than one organ (2,26). It is seen less frequent in distant metastasis cases (6). In our case, no distant metastasis was shown with abdominal CT, MRI. Whole body scintigraphy, except thoracic epidural mass without a "bony invasion". 72 pheochromocytoma cases have been described by Modlin et al (15). In the Scott et al . series only two patients were observed to have skull metastasis (22). James et al. have monitored 16 patients within 33 years by radiological examination (8) . According to the frequency of metastasis lungs, liver, lymph nodes and bones (8). Mornex et al. have observed metastasis in 14 malignant pheochromocytomas (5 lymph nodes, 5 bone, liver 4, lungs 2, brain 1) between 1966 and 1990 (16). In the literature it is seen that neurological deficits, such as paraplegia connected to vertebra collapse due to pheochromocytoma metastasis on the spine bone structures had occurred (18). Compression of the spinal cord secondary to the incident of vertebra bone structures have been observed (13,16,17,23). Metastasis has been observed in the lower sacral spinal channel (4). In our case, unique example of distant metastasis of malignant pheochromocytoma to the thoracic

epidural space without a bony invasion in the literature was realized. This case is metastatic pheochromocytoma. So this is the first case report of an epidural metastasis from pheochromocytoma. The origin of this tumor is the adrenal glands.

Though cognizant of the fact that metastatic pheochromocytoma is relatively radioresistant, as compared with lymphoma or breast carcinoma (15), the relatively benign neurologic symptoms were treated with radiation therapy. Bone lesions from these tumors tend to respond well to radiation therapy (10). Radiotherapy has not been used in our case.

Other reports have suggested that metastases of pheochromocytoma have considerable potential to be hormonally active (2,28). Thus, surgical manipulations of these are as likely to result in hemodynamic instability as their primary counterparts (2,4). Preoperative alpha-adrenergic blockade with agents such as phenoxybenzamine, prazosin, or phentolamine has been credited with reducing the morbidity and mortality from hypertensive episodes during anesthetic inductions and surgical manipulations (2,5,8,27,28). In this case, the hemodynamic instability was minimal during resections of the original tumors. Because of this it was elected to not administer an alpha-adrenergic blocking agent preoperatively. As mentioned above, the intraoperative blood pressure fluctuations were adequately managed with transient sodium nitroprusside infusion.

The rate of survival after the appearance of metastasis in malignant pheochromocytoma has usually been less than three years (14). In this case, there were no recurrences and no abnormality which revolved in 2 years of follow-up.

From this experience the authors recommend that following course of action in future cases. If a spinal lesions is discovered and is causing pain alone, or only a radiculopathy, radiation therapy will be used. If the patient progresses to new neurologic deficits during or after radiation therapy, presents with myelopathy or sphincteric dysfunction, or develops hypertension suggesting a hormonally active focus, serious consideration will be given to surgical decompression depending on the patient's overall medical condition. The determination of frequency of metastatic pheochromocytoma shall increase with sensitive activities such as CT and MRI.

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METASTATIC PHEOCHROMOCYTOMA OF THE THORACIC SPINE

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