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Recurrent Hemangiopericytoma With Multiple Extracranial Metastates: A Case Report

Multiple Ekstrakranial Metastazı olan Rekürren Hemanjioperisitoma: Olgu Sunumu

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Hemangiopericytoma is an uncommon, highly vascular tumor of pericyte origin that may occur anywhere capillaries are found. These tumors are usually located in the musculoskeletal system, retroperitoneum and the skin. Their intracranial localization is rare. Hemangiopericytomas have a predilection for both local and distant central nervous system recurrences and exhibit an increased tendency to metastasize compared with nonmalignant meningiomas. The biological behavior of hemangiopericytoma is sometimes malignant although it is considered grade II-III. Local tumor recurrence after many years is not uncommon, and late and widespread extracranial metastases may occur although this is rare in other intracranial tumors. A 46-year old male who had a recurrent intracranial hemangiopericytoma with multiple metastases such as cervical vertebrae, paravertebral soft tissue, lung, kidney and shoulder has been reported.

Key Words: ***Hemangiopericytoma, Meningioma, Extracranial metas, Recurrence***

Hemanjiyoperisitomalar nadir görülen, oldukça vasküler, kapiller damarların bulunduğu her yerde görülebilen perisit orijinli tümörlerdir. Bu tümörler genelde kas iskelet sisteminde yerleşirler. Intrakranial yarleşimleri oldukça nadirdir. Hem local hem de santral sinir sisteminin uzak bölgelerinde rekürrens yapmaya meyillidirler ve malign olmayan menenjiyomlara kıyasla uzak metastaz yapma ihtimalleri daha fazladır. Hemanjiyoperisitomların biyolojik davranışları bazen grade II-III olarak sınıflandırılmalarına neden olacak kadar malign olabilmektedir. Uzun yıllar sonra lokal tümor rekürrensi çok sık değildir ancak yaygın ekstrakranial metastaza rastlanabilmektedir. Bu yazıda, servikal vertebra, paravertebral yumuşak doku, akciğer, böbrek ve kas-iskelet sistemi metastazı olan, aynı zamanda da intracranial rekürrensi olan bir hemanjiyoperisitom olgusu sunulmuştur.

Anahtar Kelimeler: ***Hemanjioperisitom, Menenjiom, Ekstrakranial metastaz, Rekürrens***

Hemangiopericytoma (HPC) is an uncommon, highly vascular tumor of pericyte origin that may occur anywhere capillaries are found and it accounts for only 1.6-2.4 % of meningeal tumors and less than 1 % of all intracranial tumors (1-5). HPCs are usually located in the musculoskeletal system (lower extremities, pelvis, vertebrae), retroperitoneum and the skin, but they have been reported in the larynx, spleen, lung, kidney, pancreas, mediastinium, orbita, adrenal gland, lymph nodes, liver, bone, breast, and pleura. Intracranial and to the other region metastasis of HPC is seen very rare together (6-8).

In this report, we describe a patient with hemangiopericytoma to the other side of brain and a large metastasis to the cervicothoracic spine and humerus which occurred with a delay of 4 years after the removal of right frontal meningeal HPC.

Case Report

The patient, a 46-year old male, had undergone surgery for the removal of right sided anterior cranial fossa tumor with invasion of right cavernous sinus (Figure 1A, 1B). At that time, he had diplopia, head-

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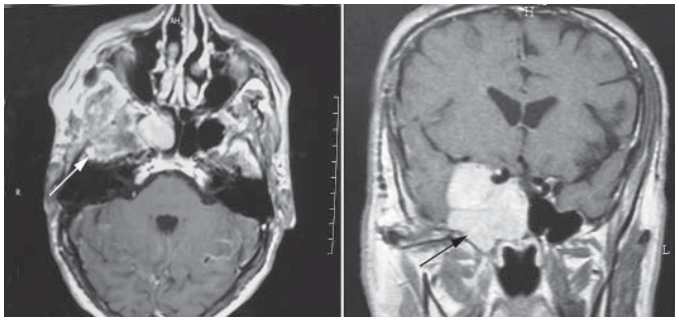

Figure 1A - 1B. Tumor, involing the anterior skull bose and right covernous sinus car be in A and B.

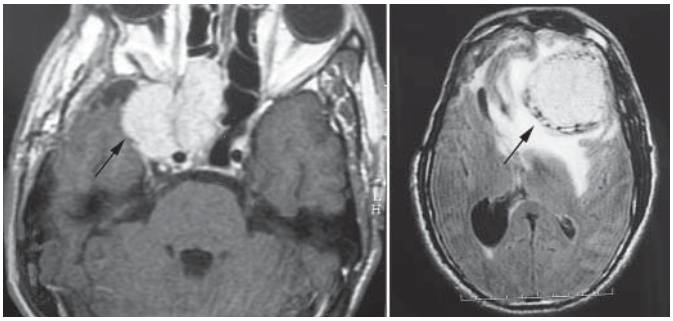

Figure 2A - 2B. Tumor recurrence can be seen at the same localization (A). And a new mass detected in his control WRI (B) at the left frontal lobe.

ache, visual deficit in his right eye and hypoesthesis on the right side of his face. Hemangiopericytoma was detected in his first histopathologic investigation and cranial radiotherapy was performed.

One year later, the patient started to present clinical symptoms, such as headache and diplopia. MRI revealed a tumor recurrence at the same region of the first localization (Figure 2A). He underwent a second neurosurgical operation, with subtotal removal of the lesion which was located in the frontal lobe, anterior cranial fossa, and the right cavernous sinus.

Three years later, he was referred to our department because of confusion and headache on left side, nubness and intractable pain at the left side of his body, with gait disability and dizziness. In his neurological examination; hypoesthesia under the level of C5 , increased the deep tendon reflexes on the left, clonus and babinski signs were positive, truncal ataxia, and also disability on pressure and heat discrimination on the left were found. Cranial MRI revealed a new mass in the left frontal lobe, but no recurrence on the right side (Figure 2B). For the purpose of explaining this clinical situation, servical MRI was performed and indicating a mass lesion (Figure 3A, 3B) wich was invading fourth cervical vertebrae corpus completely. The lesion was extending to the left lamina vertebrae via left foramen intervertebrale and also invading left vertebral pedicle as same as other classical metastatic lesions of spinal column. Left vertebral artery was encased by the mas but its lumen was still patent. Craniocervical angiograpy also showed a highly vascular lesion in his cervicothoracic region (Figure 3C). Left shoulder MRI was performed because of his left-sided arm and shoulder pain and detected a bone metastasis to his left humerus (Figure 3D). Thorax CT revealed left-sided parenchymal multiple metastatic nodules in the lungs, while abdominal CT showed multiple cystic lesions in the left kidney.

The patient underwent a third surgical operation with subtotal removal of his cranial lesion, but not for the others. The lesion was diagnosed as hemangiopericytoma as expected. The patient did not accept any adjuvant or surgical therapy; however, radiotherapy was offered as a choice. Currently, the patient is followed-up with 6-month intervals of craniocervical MRI and whole body CT scan for recurrences, metastases, and the lesion progression.

Discussion

HPC is commonly affect adults in the fifth or sixth decade of life. The biological behavior of HPC is sometimes malignant although it is considered grade II-III. Local tu-

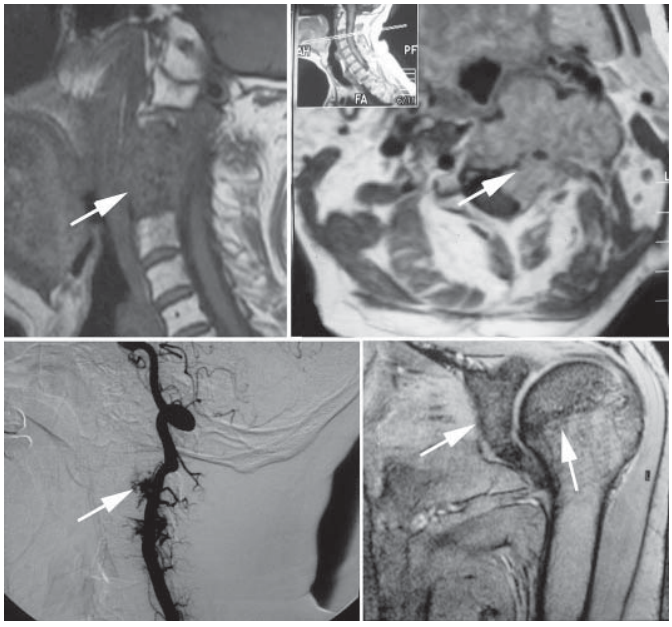

Figure 3A - 3B - 3C - 3D. Cervical metastasis can be seen in A and B. Cervical angiography revealed a highly vascular lesion at servical region (C). The left sholder MRI revealed a metastatic lesion in bony structures (D).

mor recurrence after many years is not uncommon, and late and widespread extracranial metastases may occur (7). Metastatic HPC to the spine is rare. Review of the literature identified only eight reported cases (9).

The preoperative differential diagnosis between meningiomas and HPCs confers therapeutic advantages. May any radiologic investigation help surgeons? In a recent study, Akiyama et al. reported some radiological features that may help preoperative differential diagnosis (3). Their analysis indicated that HPCs have multilobular or irregular margins, strong contrast enhancement, mainly pial-cortical blood supply, unlike meningiomas, which do not show hyperostosis or calcification. Compatible with their aggressive behavior, HPCs tend to show features such as irregular or lobulated borders, apparent parenchymal invasion (mushrooming), and more heterogeneous contrast enhancement than meningiomas. A recent study suggested that in vivo magnetic resonance spectroscopy could distinguish between meningiomas and HPCs due to the higher levels of myo-inositol in the HPCs. Recurrent solitary fibrous tumor, schwannoma, meningioma should also be considered in differential diagnosis (10).

Microneurosurgery and adjuvant radiotherapy constitute the main treatment. The tumor should be surgically removed completely, including the excision of involved dura mater and bone; when this is not possible, aggressive cauterization of the dura and bone is mandatory. Radiation therapy for treatment of this tumor has been reported to be ineffective, but the literature has increasingly documented tumor response to greater than 450 cGy of local therapy. The response rates of HPCs to currently available chemotherapy are poor (2,5). However, complete resection of the tumor is sometimes impossible because of the risk of hemorrhage during the operation (1,4,5). Hemorrhage represents the most frequent cause of surgery related death, as well as greatest hindrance to tumor removal. Early reports noted operative mortality rates that varied from 9 to 24 % (1,4). To reduce the risk, some authors prefer to use radiotherapy as neoadjuvant treatment (2).

In our case; medulla spinalis was under compression at the lesion level and this situation was able to explain why the patient's complains and the neurological findings were on the same side. The nubness and intractable pain , motor and sensorial deficitis on his left side were thought to be due to compression of fasciculus anterolateralis and tractus corticospinalis. On the other hand, gait disability and dizziness, truncal ataxia, and also disability on pressure and heat discrimination on the left were thought to be due to compression of fasciculus cuneatus and gracilis.

Conclusion

There is a general consensus on the aggressive behavior of hemangiopericytomas. Surgical resection is usually the initial treatment modality. However, even after the resection, their high recurrence rate and tendency to metastasize make HPCs very challenging to manage as our case. Radical surgery is the treatment of choice but must be supplemented with postoperative radiotherapy, which has proved to be the therapy most strongly related to the final prognosis. On the other side, in the case of discrepancy between the clinical signs and neurological findings, we decided that a hole spinal MRI should be performed for the aim of searching vertebral column metastasis and medulla spinalis compression.

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