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Case Report / Olgu Sunumu

CELLULAR VARIANT CEREBELLAR HEMANGIOBLASTOMA: A CASE REPORT

SELÜLER VARYANT SEREBELLAR HEMANJİYOBLASTOM: BİR OLGU SUNUMU

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ABSTRACT

Hemangioblastoma is a highly vascularized, have a slow-growing rate grade 1 tumor which are containing neoplastic stromal cells with clear, vacuolated cytoplasm and diagnosed with specific immunohistochemical staining features. Hemangioblastomas can occur in the brain, spinal cord or retina and they account for about 1-2.5% of all intracranial tumors. We presented a 36-year-old male patient presented with a headache and glaucoma. In the radiological images, a cerebellar mass was observed in the localization of the right middle cerebellar pedincle. After surgical resection pathological evaluation was done and as a result, the patient was diagnosed as cellular hemangioblastoma. He was followed up in the intensive care unit for about two months and unfortunately died due to the cardiopulmonary arrest associated with non-tumoral reasons. This case report was a descriptive example for a rare variant of a rare cerebellar tumors.

Keywords: hemangioblastoma, cerebellum, vascular neoplasm

ÖZET

Hemanjioblastom oldukça vaskülarize, yavaş büyüyen, berrak, vakuollü sitoplazma ve spesifik immünohistokimyasal boyama özellikleri ile tanı koyulan, neoplastik stromal hücreler içeren derece 1 tümörlerdir. Hemanjiyoblastomlar beyinde, omurilikte veya retinada ortaya çıkabilir ve tüm intrakranyal tümörlerin yaklaşık %1-2,5' unu oluşturur. Baş ağrısı ve glokom şikayetiyle hastanemize başvuran 36 yaşında erkek hastayı sunduk. Hastanın radyolojik görüntülerinde sağ posterior fossada lokalize beyin kitlesi görüldü. Cerrahi rezeksiyon sonrası patolojik değerlendirme yapıldı ve hastaya selüler hemanjiyoblastom tanısı konuldu. Yaklaşık iki ay kadar yoğun bakımda takip edilen hasta maalesef tümör dışı nedenlere bağlı kalp-akciğer durması nedeniyle hayatını kaybetti.

Anahtar Kelimeler: Hemanjiyoblastom, serebellum, vasküler neoplazm

INTRODUCTION

Hemangioblastoma is a highly vascular grade 1 tumor containing neoplastic stromal cells with clear and vacuolated cytoplasm characterized by immunohistochemical features (1) and accounts for 1–2% of all intracranial tumors (2).

The patients are mostly dominated by Asia, Europe, and North America. It is most often located in the cerebellum but can occur in the brainstem, fourth ventricle, cerebellopontine angle and the craniocervical junction. Complete tumor resection has been stated as the most effective treatment (2,3). The 1, 3, and 5-year survival estimates of the patient were 95.3%, 92%, and 88.8% (4). The annual incidence of intracranial hemangioblastoma is thought to be 0.15 cases per 100,000. Multiple hemangioblastomas were associated with VHL syndrome. Histologic variants of hemangioblastoma are reticular and cellular variants. Reticular type was composed more capillaries than stromal cells and more common but stromal cells predominant in cellular type and it is less common (1,5).

Differential diagnosis between hemangioblastoma and glial tumors is very important. Because a wrong diagnosis may lead to the use of unnecessary complex therapies with potentially harmful complications.

In this report we aimed to present a 36-year-old male patient diagnosed as cerebellar hemangioblastoma.

CASE REPORT

A 36-year-old male patient who had glaucoma was admitted to the neurosurgery department with progressive headaches. He was a heavy smoker. His general condition was fine on neurological examination, but MRI images shown an interaxial mass lesion extending from right superior cerebellar peduncle to the middle cerebellar peduncle, measuring 33 x 29 mm at its widest point, containing cystic components, creating a significant compression effect from the right posterolateral to the pons, and obliterating almost

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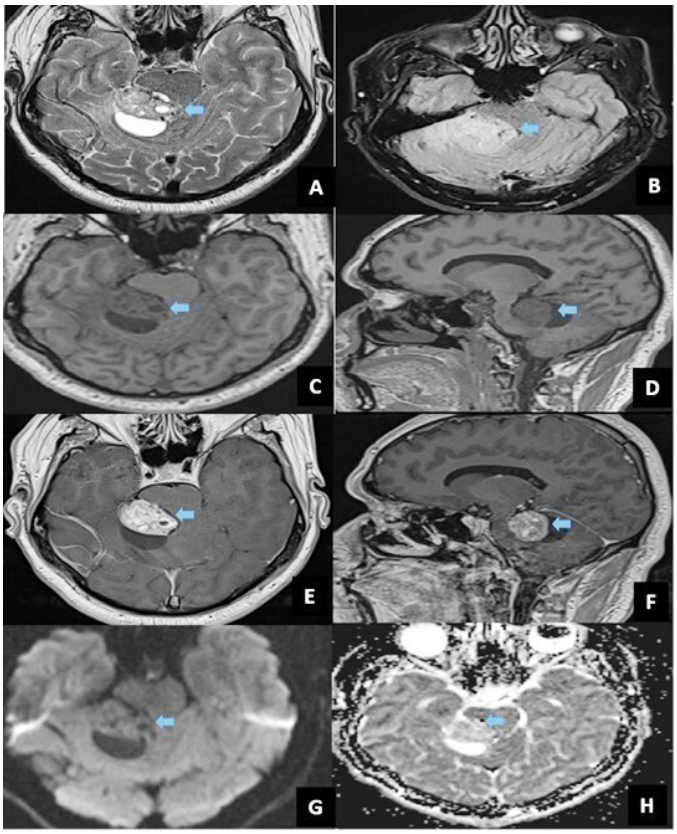

Figure 1: A. In the axial plane, T2-weighted sequence has cystic components in iso-hyperintense heterogeneous, creating a significant compression effect from the right posterolateral, obliterating the 4th ventricle close to total. B. Signal changes consistent with hyperintense edema in the hyperintense heterogeneous surrounding parenchyma in the FLAIR sequence in the axial plane C and D. T1-weighted sequence iso-hypointense heterogeneous in axial and sagittal planes, respectively. E and F. There is an intraaxial mass lesion with heterogeneous intense contrast enhancement in T1-weighted sequence with contrast in the axial and sagittal planes, respectively. G. No diffusion restriction in Diffusion-weighted and ADC sequences, respectively.

the entire 4th ventricle (Figure 1). With these findings, he was ministered to the neurosurgery service and operated but mass cannot resected completely. The invased surrounded structures were not excitable (Figure 2). The tumoral tissue was submitted to pathology laboratory for microscopic and immunohistochemical examination. Tumor was composed of neoplastic stromal cells arranged between numerous small vessels with compact non infiltrative growth with variable lobularity. Neoplastic stromal cells were predominant component. Tumoral stromal cells were positive with inhibin, CD56 and S100; vessels were positive with CD34 on immunohistochemical staining procedures. Patient was diagnosed as Cellular Hemangioblastoma with these findings.

Due to postoperative intracranial bleeding, the patient was admitted to the intensive care unit. Unfortunately after two months of care the patient was lost due to cardiopulmonary arrest. Informed verbal consent was obtained from the patient's relatives for this study.

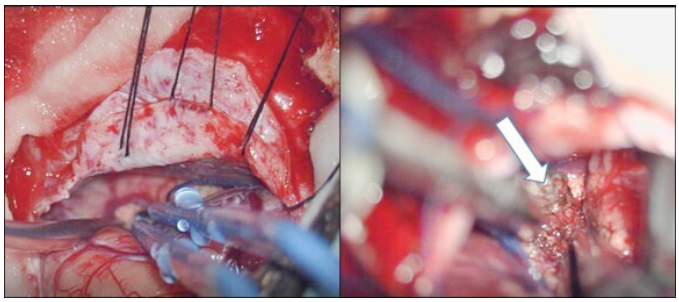

Figure 2: Perioperative images (arrow: tumoral mass)

DISCUSSION

Cerebellar hemangioblastomas are rare tumors that composed of vascular and stromal components and comprise of 1-2.5% of primary intracranial tumors.

Ataxia, dizziness, headache, and intracranial hypertension are dominant symptoms of Hemangioblastoma's. It is usually located in the posterior cerebellar fossa and is the most common primary intra-axial and infratentorial tumor seen in adults (6). Neuroimaging can be used to view nodules associated with cystic structures. Angiography can also be used to demonstrate arteriovenous malformations (1). Our patient was admitted to the hospital with a headache complaint and MRI shows us the tumor whose dimensions were 33 x 29 mm located in the widest part of the right middle cerebellar peduncle localization (Figure 1,2.).

While hemangioblastoma can be seen sporadically more commonly, it can also occur in association with VHL syndrome (5). It usually occurs in adults however VHL-associated tumors occur at an earlier age than sporadic cases (1).

Morphologically different types of hemangioblastomas have been defined. The most common was solid

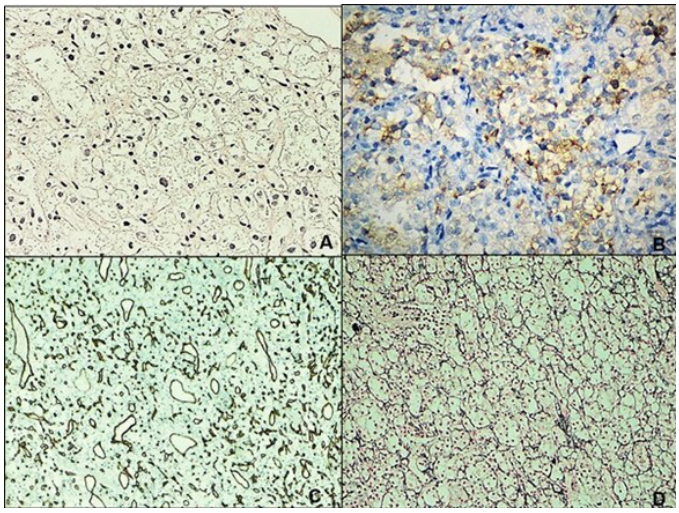

Figure 3: A:Neoplastic stromal cells placed among many thin-walled small vessels. Clear, foamy cytoplasm due to mild nuclear pleomorphism, degenerative atypia and lipid contents in stromal cells (hematoxylin & eosin stain, x400). B: positivity on stromal elements (Inhibin x200), C: Staining in an increased number of vessels surrounding the stromal elements (CD34 x200), D:Staining surrounding tumor islands (Reticulin x200)

hemangioblastomas, followed by cystic and cystic with a mural nodule, whereas tumors were described as being both solid and cystic (3).

Hemangioblastoma is characterized by two main components: large and vacuolated stromal cells which can show cytological diversity and abundantly vascularized cells (1). For this reason, it should be kept in mind that intratumoral hemorrhage may occur, albeit rare, and clinicians should consider this possibility (7).

Microscopically two variants, reticular and cellular, have been described. The predominance of cellular clusters in the cellular variant, and the dominance of capillaries in the reticular variant are noticeable. The cellular variant is characterized by fine granular eosinophilic cytoplasms, endothelial hyperplasia and the formation of glomeruloid architecture. In the reticular variant, stromal cells with high vacuolated clear cytoplasm and abundant capillaries are observed (8). Lobules of foamy cells with large clear cytoplasm and small nuclei; capillaries around the lobules were seen on our microscopic examination. As a result of this examination, the tumor was compatible with the cellular type (Figure 3.).

Appropriate immunohistochemical staining helps to establish the correct diagnosis. The positivity of inhibin, CD56, neuron-specific enolase, S100, vimentin and the negativity of epithelial membrane antigen and glial fibrillary acidic protein (GFAP) are important findings in differential diagnoses of hemangioblastomas (9).

Excision of the tumor is the most reliable way of treatment but its association with other parts of the cerebellar structure may make it difficult. Furthermore, if the excision is insufficient it may lead to a recurrence. Due to the risk of recurrence, it is highly recommended for patients to undergo regular inspection throughout their lifetime after treatment (1).

As a matter of fact, our results were compatible with cellular hemangioblastoma. The presented case is rare variant of hemangioblastoma and notable in that although this tumor is benign, it shows similar features to many malignant tumors in MRI and hematoxylin-eosin staining.

CONCLUSION

In conclusion cerebellar hemangioblastomas are uncommon intracranial tumors with good prognosis due to its slow rate of growth and benign nature. Cellular hemangioblastomas are extremely rare. Recognition of this rare tumor on radiologic, pathologic examination and considering it in the differential diagnosis is important to protect patients from overtreatment with unnecessary complex therapies and potentially harmful complications.

Informed Consent: Since the patient died after clinical follow-up, consent for the case presentation was obtained from his family.

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

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