Hemangioblastoma originating from the right cerebellopontine angle

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ABSTRACT

يعد الورم الأرومي الوعائي من الأورام القليلة الظهور في منطقة فوق الخيمة، كما ويندر حدوثه في الزاوية المخيخية الجسرية ولذلك فإن الخطأ في تشخيص هذا المرض غالباً ما يحدث قبل إجراء العملية الجراحية. نستعرض في هذا المقال حالة نادرة لمريضة تبلغ من العمر 42 عاماً وقد كانت تعاني من الورم الأرومي الوعائي في الزاوية اليمنى من المنطقة المخيخية الجسرية.

The morbidity of hemangioblastoma in the supratentorial region is very low, and is seldom found in the area of the cerebellopontine angle, so it is easily misdiagnosis before surgery. We report and discuss a case of hemangioblastoma originating at the right cerebellopontine angle in a 42-year-old female patient.

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Hemangioblastoma is a well-differentiated benign tumor that originates from the residual cells in the mesoderm. It consists of a variable mixture of mesenchymal cells and numerous capillaries. It is developed most commonly in the cerebellar hemispheres.[1] As imaging techniques such as CT, and MRI have become more widely performed, some characteristics of the imaging features have been summarized.[2] However, its morbidity in the supratentorial region is very low, as well as seldom found in the area of cerebellopontine angle. It is easily misdiagnosis before surgery because its position and imaging demonstration are not typical. Here, we investigate the features of one hemangioblastoma originating at the right cerebellopontine angle to help in improving the diagnostic accuracy.

Case Report. A 42-year-old female patient was admitted for a tumor mass found by CT scan following headaches for around 2 months. Her family history was not contributory. Physical examination revealed no abnormalities. An MRI scan showed an abnormal signal in the right cerebellopontine angle (**Figure 1**). At operation, the tumor mass was located in the right middle cranial fossa under the cerebral dura, fish meat like, medium texture, abundant blood supply, and invading the cavernous sinus, temporal lobe, and cerebellopontine angle of the same side. On pathological examination, there were abundant capillary and mesenchymal cells on Hematoxylin and Eosin stain. The results of immunohistochemical stain were CD34 (+), GFAP (-), Vimentin (+) (**Figure 2**). The pathological diagnosis was of hemangioblastoma. During 12-months follow-up, no significant symptoms of this tumor were observed and she was apparently healthy.

Discussion. Hemangioblastomas originate from the residual cells in the mesoderm, which are also called the angioreticuloma of brain, and comprise around 2.3% of intracranial tumors.[1] It is a capillary rich hemangioma with a proliferation of vascular cells. The clinical manifestations are headache, vomiting, mental confusion, blurred vision, neck pain, and ataxia. It is more common in adults aged 20-40, and males account for twice the number of females in the incidence rate. Around 15% of patients have a family history with a genetic background. Around 10-13% of patients also have polycythemia and high hemoglobin disorders. The latter being near normal level after operation, but aggravated on relapse.

In 1926, Lindau reported the first case,[3] after that, doctors found it could be combined with retinal

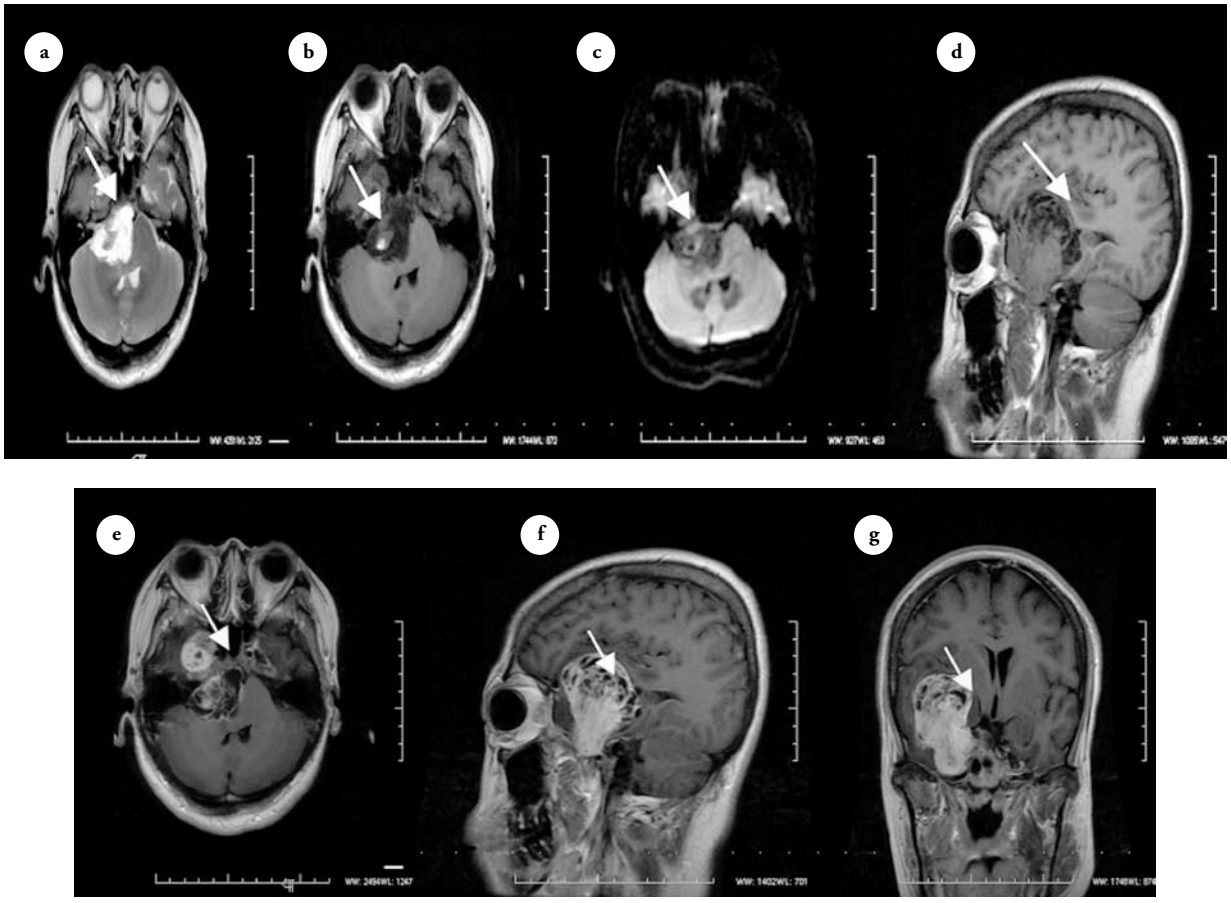

Figure 1 - Plain MRI scan showing **a, b, c, d**) 3.1 x 2.8 cm mass, mixed T1 and mixed T2 abnormal signals in the right cerebellopontine angle, round like, and peripheral edema, the neighboring tissues were compressed, distorted, and displaced. Diffusion weighted imaging demonstrated that the mass was composed of mixed signals (mainly low signal). **e, f, g**) Enhanced MRI scan showing the mass was unevenly enhanced (arrows).

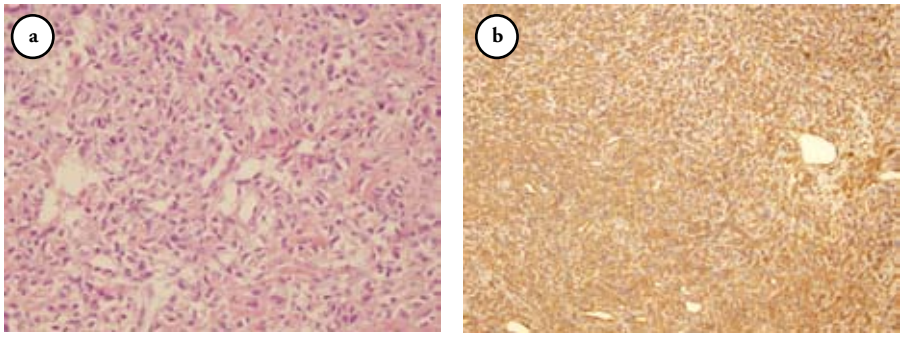

Figure 2 - Light microscopy showed **a**) the mass composed of abundant capillary and mesenchymal cells (Hematoxylin and Eosin stain x 400). **b**) Immunohistochemical stain showed vimentin expression, cytoplasmic positive (S-100P method x 400).

hemangioma, kidney, pancreas, liver and epididymal tumors, or pheochromocytoma, and so forth, which formed a special kind of syndrome, called Von Hippel-Lindau syndrome.[4] This disease is often seen in the cerebellar hemispheres (80%), secondly in the cerebellar vermis and fourth ventricle, and occasionally in the supratentorial region.[5] The MRI is the most valuable imaging technique in the diagnosis of hemangioblastoma. It has high soft tissue resolution, can produce images from multiple angles and multiple aspects, and can

demonstrate neighboring structures more clearly. According to pathology and imaging demonstration, hemangioblastoma can be divided into cystic and solid subtypes. Cystic tumors comprise around 60-90%. The cyst is under the cerebellar cortex, round or oval, high tension, with clear boundaries, the cyst fluid is straw yellow, can be self-curing, tens to hundreds of milliliters, 60% of the cyst wall has one or more wall nodes, which composed the main part, the diameter is around 1-2 cm. Most are large cysts with small mural nodules; vascular signal void around the mass can be seen in some cases. The tumor appears as hypointensity on T1WI and hyperintensity on T2WI, the wall nodes are enhanced clearly on Gd-DTPA contrasted MRI. The solid subtype does not have typical MRI characteristics.[6] Therefore, the preoperative diagnosis of this subtype is usually difficult as its position and imaging demonstration are not typical, especially in the supratentorial region. Thus, these untypical tumors need to be distinguished from cholesteatoma, cystic glioma, and cystic metastatic tumor, and so forth. However, surgeons should at least be reminded of the possibility of a hemangioblastoma.

Histopathologically, this tumor has 2 main components: mesenchymal cells and numerous capillaries. Immunohistochemically, the endotheliocytes were positive for CD34 and FVIII Rag, but most of the mesenchymal cells were positive for s-100 and part of the cells were also positive for neuronspecific enolase. The endotheliocyte and the mesenchymal cells were all positive for vimentin, but negative for GFAP, epithelial membrane antigen, and p53.The expression of Ki-67 is reported as very low.[7]

Surgical excision is the treatment of choice to confirm the diagnosis and prevent further growth and compression of neighboring structures.[8] By far, with the rapid development of microsurgery, most patients have a good effect after surgery, except for a small number of patients with recurrence in recent years.[9] So far, there have been supratentorial cases reported from different countries, however, there is limited data on cases involving the cerebellopontine angle. As the hemangioblastoma can be combined with abdominal tumors, it is necessary to undertake a careful examination of the whole body when the location and imaging results are not typical.

In conclusion, mesenchymal cells and numerous capillaries characterize hemangioblastoma. The clinicopathologic and image features have some characteristics. Clinical doctors should consider the possibility of hemangioblastoma when the location or imaging features are not typical before surgery.

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