Hemangiopericytoma of the adult male breast

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ABSTRACT

It is well known that most of the breast neoplasms are epithelial lesions with only less than 5% as non-epithelial. Vascular breast malignancy accounts for only 0.04% of all breast malignant neoplasms. Hemangiopericytoma (HPC) was first described by Stout and Murray in 1942 as unusual vascular tumors that originated from the Zimmermann's pericytes which proliferate in the vessel wall. Hemangiopericytoma has been reported to develop throughout the human body, such as the lower extremities, meninges, pelvis, retroperitoneum, trunk, upper extremities, uterus, liver, orbit and oral cavity. Hemangiopericytoma is uncommon in the breast, only few cases of HPC in the breast have been reported in the literature including 2 cases of HPC in the male breast. In this report, we presented a 24-year-old man with a mass in his right breast, and reviewed the clinical, pathologic, immunophenotype characteristics of this case and our treatment protocols for this rare tumor.

Case Report. On March 2010, a 24-year-old man with a mass in his right breast was admitted to our hospital. The mass was 7 cm in diameter, no pain, no nipple discharge or hemorrhage. He had no remarkable family diseases or breast cancer history. The patient had detected the mass in right breast one month earlier before he visited us. Physical examination revealed that an approximately 7 cm in diameter, irregular, firm mass was located in the upper lateral quadrant of his right breast, and no nipple discharge or lymph nodes were touched in the right axillary fossa. Ultrasound examination showed that the mass was approximately 9.0 cm x 6.7 cm x 4.2 cm, well-circumscribed, solid, predominantly hypoechoic with heterogeneous internal echoes, and without calcification. For the accurate diagnosis, we performed the complete excision of the neoplasm under the local anesthesia. The fast frozen pathological examination showed that the mass was a malignant tumor with a fibrous capsule and multiple rounded or irregularly shaped areas with abundant fibrous tissue and scattered round and oval tumor cells with eccentric nuclei and eosinophilic cytoplasm. Immunohistochemical staining revealed that the tumor cells were positive for CD34 and vimentin, and negative for S100, Ulex europaeus lectin, and melan-A. The tumor cells also showed a high expression of CD99. These features are consistent with the diagnosis of HPC. The patient tolerated the operation well and was discharged on the second postoperative day. The patient has been followed up for one year and has remained disease-free.

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examination revealed that it was the potential malignant tumor of the breast, perhaps originated from the vessels of the breast. Then, the modified radical mastectomy was operated. After operation, no adjuvant therapy was administered. There were no evidences of tumor recurrence or metastasis as detected by ultrasound and computer tomography methods during the 9 months of postoperative follow-up.

**Pathology examination.** The resected specimen was approximately 8 cm x 5 cm x 3 cm, grey and yellow in cross-section, without skin infiltration. The lesion was composed of relatively monomorphic oval to spindle shaped cells, and the tumor cells had eosinophilic or amphophilic cytoplasm and distinct cell borders, round nuclei. The tumor cells grew concentrically lamellated pattern around the blood vessels. The blood vessels of the lesion were close-up and variable in size, most of them were capillary. In some area, the spindle tumor cells spread as that of the myofibromatosis (Figure 1). Neither necrosis nor calcification was observed. The mitoses were no more than 1/10 high power field. Four axillary lymph nodes were found, which were soft without any evidence of metastasis. All of the lymph nodes showed symptoms of reactive hyperplasia.

**Immunohistochemical examination.** Immunohistochemical examination showed that the tumor cells were positive for CD31, CD34 (Figure 2), CD99 (Figure 3), F-VIII, vimentin and SMA, and were negative for actin, S-100, CD10, CD117, CD68, EMA, desmin, bcl-2 and AE1/AE3. Staining with MIB-1, an antibody against Ki-67, demonstrated that the positive proliferating cells were less than 3%. Based on the morphological aspects and immunohistochemical examination, the tumor was diagnosed as HPC.

**Discussion.** Hemangiopericytoma in the male breast is very rare and only 2 cases of HPC in the adult male breast have been reported until now. Hemangiopericytoma can occur anywhere in the body, and also develop as a primary malignancy or a secondary tumor following treatment of an epithelial breast cancer. In addition, HPC can metastasize to the breast.

**Clinical characteristics.** Due to lack of specific clinical aspects, HPC could not be diagnosed by routine examinations such as ultrasound, mammography, blood or serum detection. Notably, most of HPC patients only present a mass at one of the breast.

The accurate diagnosis of HPC is based on the complete excision of the neoplasm or the punch biopsy, followed by morphological characteristics and immunohistochemical examination. Buecker et al reported a case of 66-year-old female with a mass in her left breast, a punch biopsy was obtained and diagnosed as hemangiolipoma. She did not receive any treatment since no malignant tissue was found. Two years later, another mass was detected in the same breast and grew quickly. Based on ultrasound-guided core biopsy and immunohistochemical examination, this tumor was
diagnosed as the malignant HPC. This case demonstrated that traditional pathological examination may lead to the wrong diagnosis. Another report revealed that sampling by fine-needle aspiration (FNA) is a useful and accurate tool to confirm recurrent or metastatic HPC in combination with immunohistochemistry method.

To avoid the metastasis of the same side axillary fossa, it is necessary to examine the patient intimately. The mass of our patient was so big to affect the appearance of the patient that it was necessary to operate. The breast tissue of the male is small, so we performed the modified radical mastectomy and the right axillary fossa tissue was operated and we found 4 little lymph nodes after operation.

**Immunophenotype.** The biological behavior of HPC is varied and difficult to predict. To distinguish it from the solitary fibrous tumors, immunohistochemistry is very helpful for the diagnosis. We employed a range of antibodies to characterize the immuno-phenotype of the tumor cells and found that the tumor cells were positive for CD31, CD34, CD99, F-VIII and vimentin. However, it is important to note that different immunophenotypes of HPC has been observed by different authors. For example, Zalinski et al reported that HPC neoplastic cells were positive for bcl2 and CD99, but not for CD34.

**Therapy.** While no standard therapy has been proposed for HPC, complete tumor resection with negative surgical margins has been employed by most surgeons. In addition, we think it is necessary to dissect the axillary fossa lymph nodes to avoid the potential metastasis, although some researchers think that complete axillary lymph node dissection is not necessary. In our case, the modified radical mastectomy was performed and the lymph nodes from the axillary fossa were resected. As for radiotherapy, only a report demonstrated that it may be useful to treat HPC. Radiotherapy could be administered after the neoplasm was operated completely if the tumor was more than 2 cm in diameter. However, there was no evidence that the radiotherapy was helpful for breast HPC according to the other literature, so we did not administrate the radiotherapy in this case. On the other hand, chemotherapy is controversial for HPC based on current literature.

**Prognosis.** The malignant or benign nature of HPC is still controversial. The key factors of HPC prognosis include the histopathological aspects such as tumor size, tumor cells morphology, mitotic activity, necrosis, and the proliferation index. However, a report supposed that the prediction of the biologic behavior of HPC based on cytological features was not feasible. Adem et al argued that tumor size was a more valuable prognostic factor than tumor grade, while the others described HPC as a clinically low-grade form of sarcoma regardless of tumor size. Spatola and Privitera reported a case of HPC metastasized to the right breast, the patient received the radiotherapy and chemotherapy, and died 12 years later, suggesting that a long survival is possible for HPC patients. The recurrence and metastasis of HPC is rare, perhaps due to poor circumscription of the lesions.

In our case, the tumor size was relatively small compared to previous reported cases. After we resected the mass completely, the patient was followed up without any treatment. A prolonged follow-up to treat liver metastases of HPC was proposed in a recent study. Therefore, we think that the follow-up for HPC is necessary during the clinical practice.

In summary, HPC is a rare tumor in the male breast originated from the vascular pericytes. There were no standard treatment protocol, the key was the accurate diagnosis before the operation. The accurate diagnosis of HPC based on the histo-pathological and immunohistochemical examinations. For the recurrence and metastasis of HPC was rare in the breast, we think it was necessary to follow-up the patient after the operation without any chemotherapy and radiotherapy.

**References**