

Refractory Sinonasal Hemangiopericytoma: Rapid Recurrence or Growth of Residual Tumor

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ABSTRACT

Hemangiopericytoma is a malignant vascular tumor that is rarely seen in the nasal cavity and paranasal sinuses. Biological behavior of the tumor is not completely known and its natural history is not predictable. According to the existing literature, local recurrence rate is not common and distant metastasis is exceedingly rare and majority of patients have a favorable clinical course. We have presented female Persian case of sinonasal hemangiopericytoma in which recurrence of the tumor happened very rapidly after initial removal. It also recurred within two months after reoperation. We have discussed about the characteristics of these tumors and known prognostic factors. We have finally proposed that the sinonasal hemangiopericytoma may have significantly more aggressive clinical course than the previous assumption.

Keywords: Hemangiopericytoma, Malignant vascular tumor, Early recurrence.

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INTRODUCTION

Hemangiopericytoma (HPC) is a rare vascular tumor derived from Zimmerman's pericytes, which surround all capillaries. It accounts for 1% of all vascular tumors (1). It is most commonly presented in retroperitoneal space, pelvis and lower extremities and has a malignant behavior (1).

Of all hemangiopericytomas, about 7.5% are found in the head and neck with a significant tendency to occur in nasal cavity and paranasal sinuses (1, 2). Ethmoid and sphenoid sinuses are most commonly involved. According to the majority of reports, sinonasal HPC has more benign behaviour and does not have the similar risk of malignancy and metastasis as HPCs elsewhere (1). It seems to be due to a variation in histological and clinical features of sinonasal type hemangiopericytomas compared with its other soft tissue counterparts. Effective treatment requires wide surgical excision with clear resection margins. These tumors are relatively radioresistant (3).

CASE REPORT

A 40 year old Persian woman presented to the otolaryngology clinic of Loghman Hakim general hospital with a 6 months complains of nasal obstruction. She had occasional episodes of epistaxis and headache. There was no significant past medical and familial history. On physical examination, there was a fleshy polypoid mass filling the entire left nasal cavity and displacing the septum to the right. There was slight bleeding on touch. A paranasal sinus CT scan was performed which revealed a soft tissue density lesion occupying the entire left nasal cavity as well as maxillary and ethmoid sinuses, projecting through the posterior choanae into the nasopharynx and deviating the septum to the right. In gadolinium enhanced MRI, the tumor did not seemed to be extended beyond the cribriform plate. Histopathological examination of the tumor biopsy specimen revealed tumoral cells, which were arranged in interlacing fascicles, separated by dilated and irregularly shaped branching blood vessels. Cells had oval vesicular and elongated hyperchromatic nuclei with indistinct cytoplasmic borders. Mitosis was 5/10 in high power field

(HPF). Immunohistochemistry (IHC) markers investigated for confirmation of the diagnosis. Smooth muscle actin (SMA) was strongly positive and vimentin was positive while CD34, S100 and CD99 were negative. Diagnosis of Hemangiopericytoma was confirmed.

posterior part of the hard palate were involved by the tumor. A complete removal including the resection of hemipalate was done. The histopathology of the tumor did not show any difference comparing with the initial one regarding the pleomorphism of the tumoral cells.

Figure 1.Initial CT scan of the patient.



Figure 2. Recurrence of the tumor.



A grossly complete removal of the tumor was performed using midface degloving approach. A complete sphenothomideotomy along with maxillectomy and resection of orbital floor was done. In histopathology report, surgical margins were free of tumor. Seven weeks after surgery, the patient developed nasal obstruction again. The imaging study showed a huge recurrence in which the tumor had extended from palate up to cribriform plate with involvement of the hard plate. The intracranial structures seemed to be spared. Reoperation performed with the same approach. The second and third molar teeth and

The resected margins were reported to be free of tumor. Six weeks after the second surgery, the patient developed proptosis and further investigations showed a significant recurrence. The patient refused more work up and preferred to continue her treatment by non-surgical means. Hemangiopericytoma is a rare vascular tumor that may present anywhere in the body. It more commonly involves the soft tissues of the trunk and lower extremities. Approximately 17% of all hemangiopericytomas occurs in the head and neck region, mainly in the nose and paranasal sinuses (5). Hemangiopericytoma is present in all ages but

they are more common in the third to fifth decades of life. The tumor has no known risk factors such as sex, race or hereditary. The pathogenesis of the tumors is not well known. Sinonasal HPC is commonly presented with nasal blockage and/or epistaxis. In advanced stages, HPC presents with local pain, headache and visual disturbances (6).

Diagnosis is depended on endoscopic examination and imaging modalities including CT scan and MRI. The characteristic findings is a soft tissue mass, which enhances after administration of intravenous contrast. The role of routine angiography in the management of the tumor is not clearly defined. However it is accepted for pre-operative planning and embolization, which in turn significantly reduces the risk of intraoperative hemorrhage (7, 8). The confirmation of diagnosis is based on histopathological findings, which reveals the tumor as tightly packed proliferated spindle cells surrounded by an intact reticulin sheaths. The cells have oval to elongated nuclei. The cytoplasm are sparse, and the nuclei are vesicular. The arrangement of tumor cells produces interlacing bundles with many vessels lining between them occasionally interconnected with stag horn pattern (6, 8, and 9). In IHC, the HPC cells are invariably positive for vimentin and SMA. They are very rarely positive for other smooth muscle markers such as desmin. A sub-population of HPC cells are immunoreactive for factor XIIIa and histocompatibility antigen HLA-DR. (3, 5).

The treatment of choice is wide surgical resection, which is the only curative modality. Radiotherapy has not been considered as a useful modality because these tumors are radioresistant and there is no confirmed evidence that chemotherapy is beneficial for HPC (6, 10). The recurrence rate of HPC is quite varied, ranging from 7% to 20% (1, 3 and 11). The average time of recurrence is reported to be 6–7 years (4). While a primary incomplete excision has been identified as the initial factor in recurrent disease, severe nuclear pleomorphism, osseous invasion, tumor size larger than 5 cm and a high mitotic rate significantly increase the risk of recurrence (5). A metastatic chance of up to 15% has been reported (1).

We presented a 40 year old female with a large sinonasal HPC measuring more than 5 cm. It also involved several adjacent bony elements. Moreover, these are factors that contribute with higher recurrence rate; Our patient

demonstrated a clinical course that was considerably worse than any scenarios which we could predict using published literature. The histopathological survey of this tumor did not show any predictors of invasive tumoral behavior and the diagnosis of HPC was confirmed by strong immunohistochemical evidence. In either the first and second surgeries, there was no gross residual tumor remained. The surgical approach with a similar technique was performed previously on several similar patients and it resulted in a favorable outcome (11). There is no distinct explanation for the invasive behavior of HPC in the presented case other than large initial size. Nonetheless, as we know, there is no reported case of sinonasal HPC which is as aggressive as the one we were involved with. Even in the case of a residual tumor, based on the published literature, we did not expect a significant regrowth within few weeks. It seems that there are some factors other than the mentioned ones for predicting the course of this disease. We believe that we should expect a significantly more malignant behavior on this previously assumed low grade- slow growing tumor. This is especially important when we talk to patients and their families about the natural course and prognosis of this tumor. Further works are required to find out and explain exact predictors of the tumor behavior and the best treatment modality.

CONCLUSION

The sinonasal hemangiopericytoma may have significantly more aggressive clinical course than the previous assumption. The best clinical predictors of recurrence and treatment modalities are needed to be found by further works.

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