

Sinonasal tract glomangiopericytoma (hemangiopericytoma)

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A glomangiopericytoma (sinonasal-type hemangiopericytoma) is a tumor believed to derive from perivascular modified smooth-muscle cells. Its origin is similar to that of a glomus tumor (not to be confused with glomus jugulare, which is a different neoplasm) but distinctly different from soft-tissue hemangiopericytoma. There is a very slight female preponderance, and the tumor's peak incidence occurs during the seventh decade of life. Most affected patients experience nasal obstruction and epistaxis along with a wide array of other nonspecific findings that are generally present for less than 1 year. Glomangiopericytomas have a predilection for the nasal cavity and paranasal sinuses, where they grow as polypoid masses. Their average size is approximately 3 cm, and they are often mistaken clinically for inflammatory polyps.

Histologically, these tumors are submucosal, usually covered by an intact respiratory epithelium. There is a diffuse growth of closely packed cells that appear in short interlacing fascicles (storiform, whorled, and palisaded patterns can be seen) that are richly vascularized (figure, A). The vascular channels range from capillary-size to large patulous spaces that may have a ramifying "staghorn" or "antler-like" configuration. A prominent peritheliomatous hyalinization or fibrosis is characteristic (figure, B). The neoplastic cells form a closely packed syncytium of uniform, monotonous, oval to slightly spindle-shaped cells with indistinct cell borders that contain vesicular to hyperchromatic, round to oval to spindle-shaped nuclei.

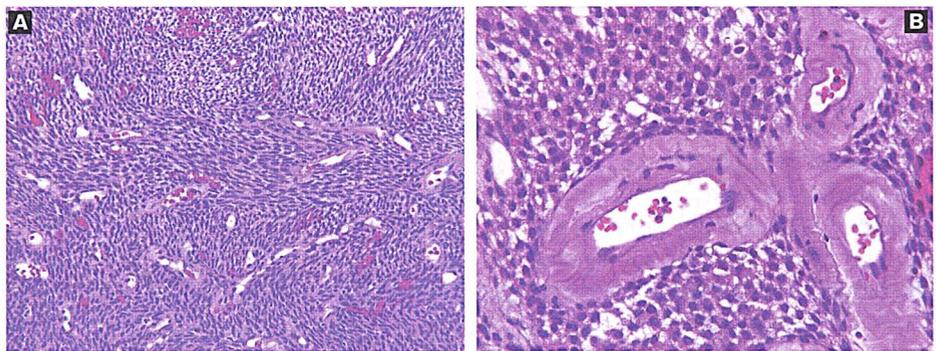


Figure. A: An interlacing spindle-cell population contains elongated to oval nuclei with coarse nuclear chromatin distribution. Many vascular spaces are identified, along with areas of erythrocyte extravasation (small red cell aggregates) and rare mast cells. B: The very dense hyalinization around small to medium-sized vessels is characteristic of a glomangiopericytoma. The cells are nondescript, although the nuclei are hyperchromatic.

Extravasated erythrocytes, mast cells, and eosinophils are almost always present. The tumor cells are immunoreactive with actins and vimentin but not with CD34, CD31, or FVIII-RAg. A "hemangiopericytoma-like" pattern can be found in a wide array of neoplasms of divergent differentiation (e.g., lobular capillary hemangiomas, angiofibromas, meningiomas, and leiomyomas), but the characteristic histologic and immunophenotypic features allow for separation.

Glomangiopericytomas are indolent tumors; the overall survival rate achieved with complete surgical excision is excellent (>95% at 5 yr). Recurrences have been reported to develop in as many as 30% of cases; they can be managed by additional surgery and/or adjunctive therapy.

Suggested reading

Thompson LD, Miettinen M, Wenig BM. Sinonasal-type hemangiopericytoma: A clinicopathologic and immunophenotypic analysis of 104 cases showing perivascular myoid differentiation. *Am J Surg Pathol* 2003;27:737-49.