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Emilio Q. Villanueva III, MD[1]
Jose M. Carnate, Jr., MD[2]

[1]*Department of Laboratories,*
Philippine General Hospital
University of the Philippines Manila

[2]*Department of Pathology,*
College of Medicine
University of the Philippines Manila

Glomangiopericytoma: The Sinonasal Mimic of Soft Tissue Hemangiopericytoma

A 42-year-old Filipino male with a 10-month history of progressive left nasal obstruction and rhinorrhea and a clinical impression of nasal polyposis underwent endoscopic sinus surgery with partial ethmoidectomy and polypectomy.

We received several dark-brown, irregular, rubbery tissue fragments with an aggregate diameter of 3 cm. Histopathologic examination shows sheets of spindly tumor cells beneath the respiratory epithelial lining. These spindle cells are closely packed and arranged in short fascicles and storiform clusters surrounding hyalinized large vessels or thin-walled submucosal blood vessels. *(Figures 1 and 2)* There is no atypia or necrosis. Immunohistochemical studies show strong immunoreactivity to muscle specific actin, and focal reactivity to S-100. *(Figure 3)* Stains for CD34, caldesmon, cytokeratin, and desmin, are negative. *(Figure 4)* Based on these features, we diagnosed the case as glomangiopericytoma.

Glomangiopericytoma is a rare tumor arising from the pericytes surrounding capillaries, and accounts for less than 0.5% of all sinonasal tumors.[1] It has a very slight female preponderance, with a peak incidence during the seventh decade of life. The most common symptom is nasal obstruction, or epistaxis, with accompanying difficulty breathing, sinusitis and headache. A mass, or polyp is the most common clinical finding.[2]

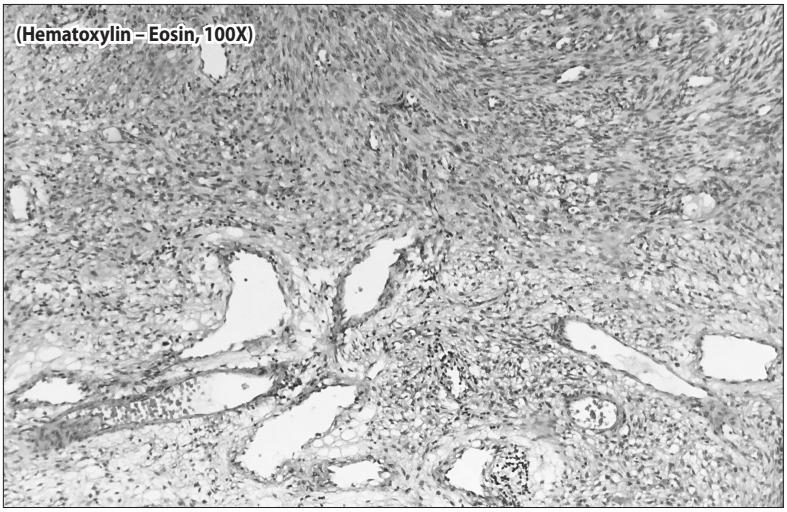

Figure 1. Cellular spindle cell tumor with branching thick blood vessels (Hematoxylin-eosin, 100X magnification)

Correspondence: Dr. Jose M. Carnate, Jr.
Department of Pathology
College of Medicine, University of the Philippines Manila
547 Pedro Gil St. Ermita, Manila 1000
Philippines
Phone (632) 526 4450
Telefax (632) 400 3638
Email: jmcjpath@gmail.com

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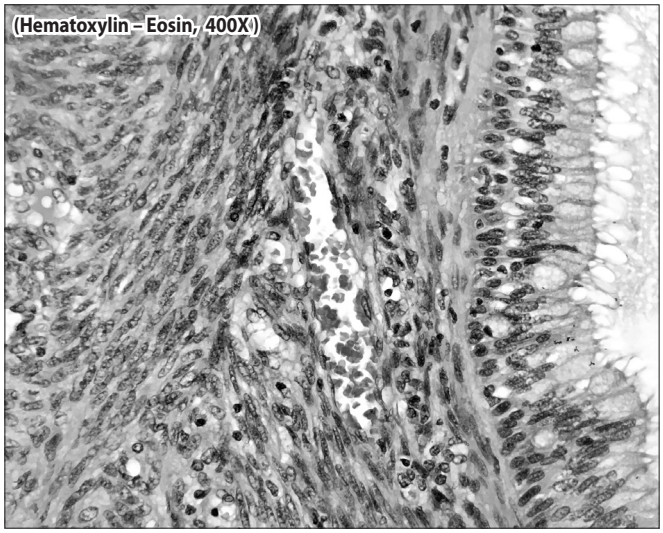

Figure 2. Closely packed spindle cells, in short fascicles surrounding a thin-walled blood vessel (Hematoxylin-eosin, 400X magnification).

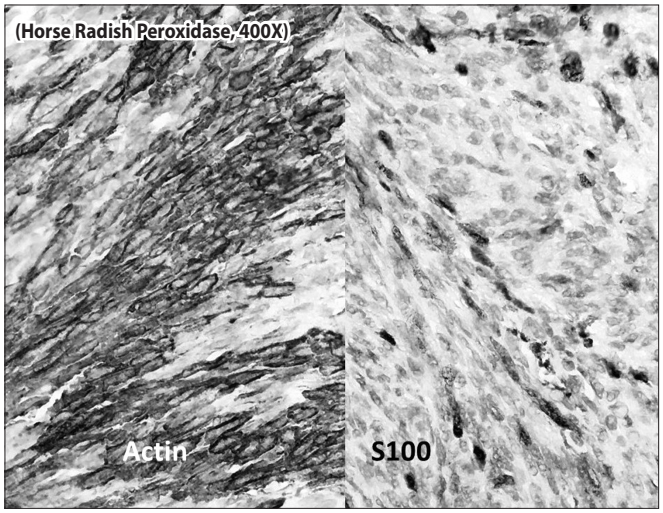

Figure 3. Tumor cells strongly positive for smooth muscle actin and focally positive for S100 (Horse radish peroxidase method, 400X magnification).

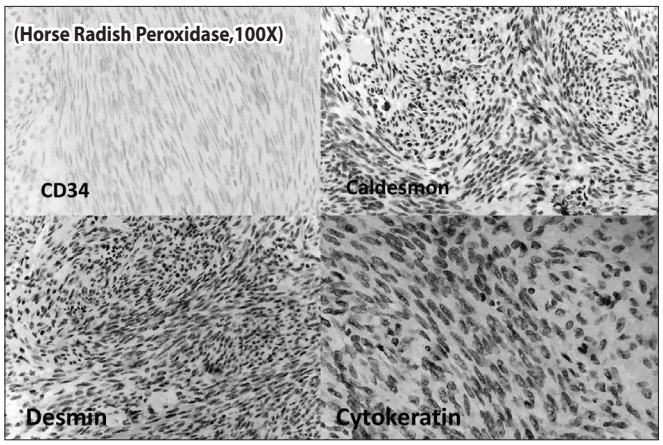

Figure 4. Tumor cells negative for CD34, caldesmon, desmin, and cytokeratin. Focal caldesmon positivity is seen in associated capillaries (Horse radish peroxidase method,100X magnification).

Hematoxylin–eosin staining shows a well-delineated but unencapsulated cellular tumor underneath the normal respiratory epithelium that effaces or surrounds adjacent normal structures.[2] The tumor is composed of closely packed, uniform, oval to spindle-shaped cells, in short fascicles and in storiform, whorled or palisaded patterns. The cells surround numerous branching thin-walled, blood vessels, thus the morphologic resemblance to soft tissue hemangiopericytoma/ solitary fibrous tumor. However, in contrast to hemangiopericytoma, glomangiopericytoma shows diffuse reactivity to muscle actins, and non-reactivity to CD34, while hemangiopericytoma shows the reverse reactions. Desmin and caldesmon are likewise non-reactive, distinguishing the tumor from leiomyomas or leiomyosarcomas of the upper aerodigestive tract. Cytokeratin non-reactivity distinguishes it from spindle cell carcinoma. S100, although typically negative, can be focally and weakly positive in a small percentage of tumor.[3]

Glomangiopericytoma is categorized as a borderline low malignancy tumor with an overall survival of >90% in 5 years but which tends to recur in up to 30% of cases. Strict follow-up is thus required, especially if complete resection is not achieved.[1]

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