CASE REPORT

Management of Hemangiopericytoma Emulating a Thyroglossal Cyst: A Rare Presentation

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Abstract

Anterior midline neck masses that move with both deglutition and protrusion of tongue are commonly suspected to be thyroglossal cysts, subhyoid bursitis, or ectopic thyroid tissue. We report a middle-aged male patient with an unusual presentation of hemangiopericytoma in the subhyoid region, clinically mimicking a thyroglossal cyst, and its management.

Keywords: Hemangiopericytoma, Midline neck swelling, Subhyoid swelling.

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Background

Hemangiopericytomas are uncommon soft-tissue neoplasms that arise from pericytes, which are the cells surrounding the endothelial cells of vessels. The tumor was first described by Stout and Murray in 1942.[1] Hemangiopericytomas are usually found in adults in sixth and seventh decades, and only rarely in children—with no sex predilection.[2] These tumors occur most frequently in the lower extremity, the pelvis, and the retroperitoneum. Approximately 17% occur in the head and neck.[3] We report a patient with an unusual presentation of hemangiopericytoma.

Case Description

A 56-year-old male presented to our outpatient department with a painless swelling over midline of neck since past 6 months, insidious in onset gradually increasing in size. He also complained of deepening of voice since past 2 months.

On examination, a solitary ovoid swelling approximately 4 × 4 cm was noted over the upper midline of the neck, 2 cm from the anterior border of bilateral sternocleidomastoids, which moved with deglutition and protrusion of the tongue (Figs 1 and 2). The swelling was nontender, with no local rise in temperature. It was firm to soft in consistency, with smooth surface, well-defined borders,

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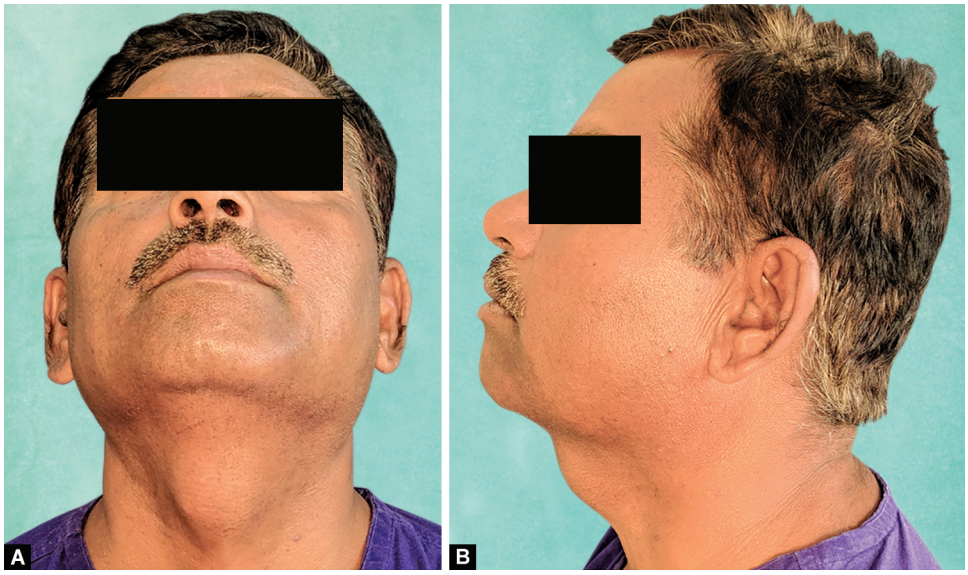


Figs 1A and B: Midline neck Swelling A frontal View, B- Lateral View

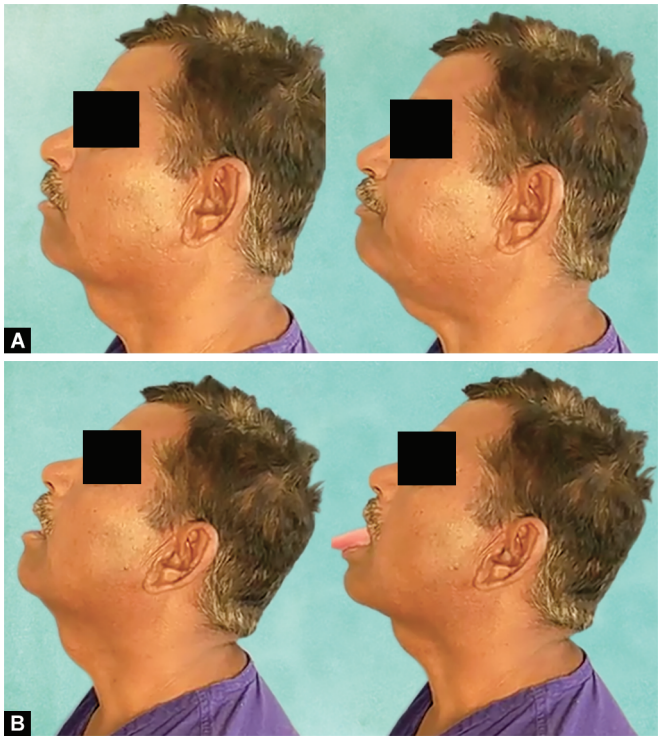
and was nontransilluminant. The skin over the swelling was normal. These features led to a clinical diagnosis of thyroglossal cyst.

Ultrasonography neck was suggestive of an infected thyroglossal cyst. Videolaryngoscopic examination was done to evaluate voice change, which revealed anteroposterior narrowing of supraglottis due to posterior displacement of epiglottis by the swelling. Vocal cords were not clearly visualized. Hence, contrast CT of head and neck was sought. It showed a well-defined homogeneously enhancing soft tissue mass, measuring approximately 4 × 4.1 × 3.9 cm, extending from the hyoid bone till the level of thyroid cartilage in the midline, also occupying pre-epiglottic space. It had well-maintained fat planes and no calcifications within the lesion—with a differential diagnosis of infection or neoplasm of the thyroglossal cyst (Fig. 3). To rule out malignant lesion, a USG-guided fine needle aspiration cytology (FNAC) was done, which yielded bloody aspirate containing neutrophils, lymphocytes, macrophages, and RBCs. No abnormal cells were seen, again suggestive of an infected thyroglossal cyst.

Routine blood investigations and thyroid function tests were within normal limits.

A novel simple technique of transthyrohyoid approach was planned. A curvilinear horizontal incision was taken over the swelling, and superficial fascia and the thyrohyoid muscle was dissected. The mass was found to be encapsulated with prominent blood vessels over the capsule. It was dumbbell-shaped, the superficial part occupying the subhyoid region, inferiorly abutting the thyroid cartilage, and the deeper part occupying the pre-epiglottic space (Fig. 4). Complete excision of the mass along with the mid third of the hyoid bone was done, without injury to the surrounding structures. His voice reverted back to normal within a few hours after surgery. The postoperative period was uneventful.

The cut section revealed a solid, yellow-white mass. Histopathological examination showed numerous vascular spaces surrounded by spindle-shaped cells, fanning out from the wall of the vascular space into the surrounding stroma. Reticulin stain surrounded the individual cells, suggestive of a tumor of pericytes



Figs 2A and B: Swelling moving with (A) Swallowing; (B) Protrusion of tongue

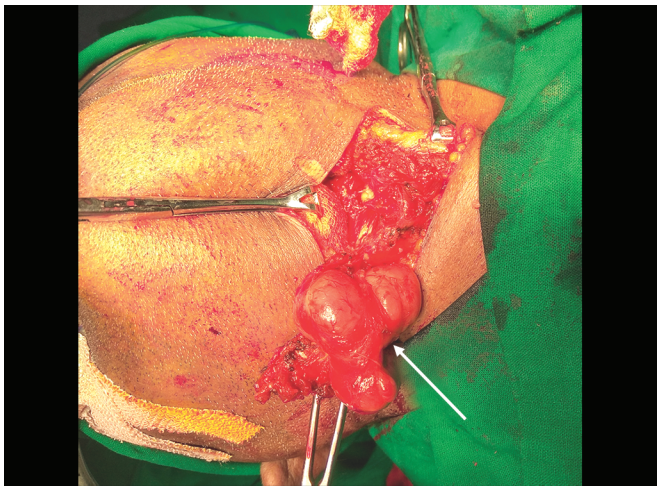
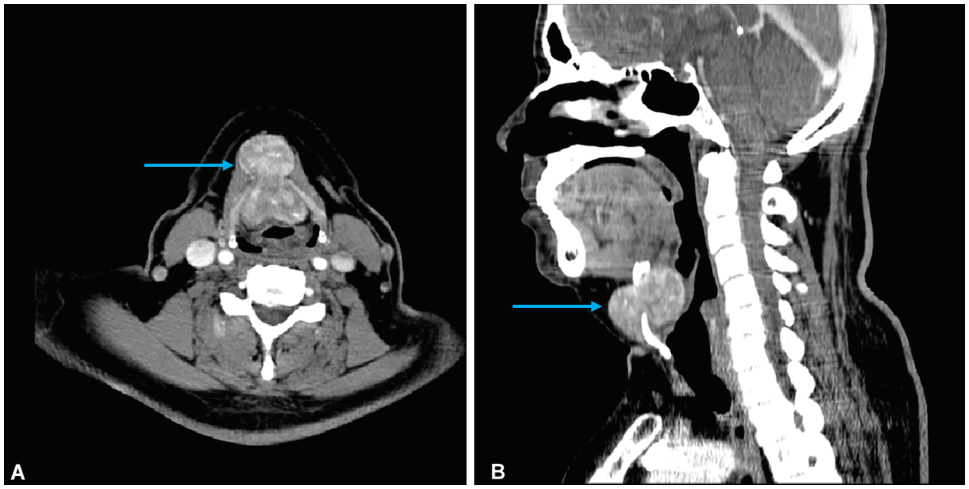
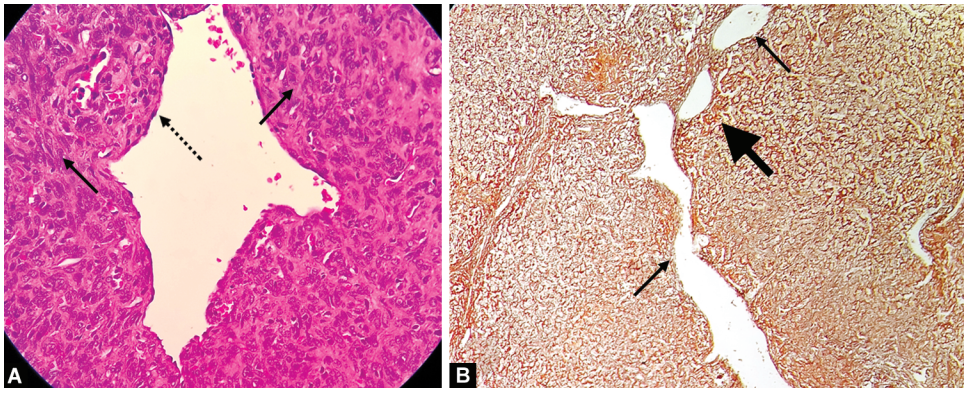


Fig. 4: Dumbbell-shaped mass excised with the mid third of the hyoid bone



Figs 3A and B: Contrast-enhanced computed tomography scans showing a well-defined homogeneously enhancing soft tissue mass in subhyoid region, extending into the pre-epiglottic space: (A) Axial section; (B) Sagittal section



Figs 5A and B: (A) Hematoxylin and eosin stain, 40x showing vascular spaces lined by endothelial cells (dotted arrow) and spindle-shaped cells fanning out from vascular wall (arrows); (B) Reticulin stain, 40x. Stain surrounding individual cells (arrow head) around the capillaries (arrows) "staghorn appearance"

(Fig. 5)—hemangiopericytoma. High mitotic activity and necrosis were absent.

Discussion

Zimmerman, in 1923, first described the pericyte as "a smooth muscle-related cell with contractile powers, although lacking myofibrils, and long processes that wrap around capillaries to change the lumen caliber."[4] It was not until 1942 that Stout and Murray coined the term "hemangiopericytoma" to describe a tumor composed of capillary buds that was surrounded by a rich reticulin network.[1] Though past history of trauma, prolonged steroid use, and hypertension may have etiological significance,[2] our patient was recently diagnosed to be hypertensive.

Enzinger and Smith[3] analyzed 106 cases of hemangiopericytoma and stated that the most common locations were the lower extremities (37), pelvis and retroperitoneum (26), head and neck (17), followed by trunk (15). According to Espat[5] et al., the most frequent anatomic sites were the extremities in seven patients (28%), the pelvis in seven patients (28%), and head and neck in six patients (24%).

In the larynx, hemangiopericytomas present as a supraglottic cyst-like mass that is firm, usually well circumscribed, in a submucosal location with size up to 4 cm. The surface is covered by intact epithelium with dilated vessels.[6]

Grossly, hemangiopericytoma is a well-circumscribed, brown, spongiform lesion, surrounded by a pseudo-capsule. Small satellite nodules are often present. They can be lobulated or nodular, firmly attached to muscle or fascia, and soft, firm, or friable.[5] Microscopic examination is characterized by small closely packed cells with ill-defined cytoplasm and darkly stained nuclei around slit-like or sinusoidal vascular spaces.[2] It is described as "staghorn pattern" with reticulin surrounding individual cells.[7]

McMaster[8] established a grading system as shown in Table 1. Enzinger and Smith[2] found that tumors with prominent mitotic activity, necrosis, hemorrhage, and increased cellularity were prone to recur or metastasize.

The mainstay of treatment for hemangiopericytomas is surgical excision. The indications for adjuvant treatment are controversial, preferred only in cases with incomplete resection, inoperable tumors, local recurrence, and metastasis.[2] Though there are several reports of adjuvant radiation and chemotherapy in a few case series, there are no large-scale studies looking at outcomes of postoperative adjuvant treatment. In addition, the clinical course and treatment options for laryngeal hemangiopericytomas are unknown due to the limited number of cases.[9] Since recurrence and metastasis can occur after many years, a lifelong regular follow-up is necessary.[2]

Table 1: Grading system for hemangiopericytoma

Grade	*Microscopic findings*
Benign or low-grade	Prominent vascular pattern with mostly spindle-shaped pericytes and no mitoses
Borderline or intermediate grade	More cellular with compressed vascular spaces, plumper tumor cells, and rare mitoses
Malignant or high-grade	Cellular anaplasia, a higher mitotic rate, and more compressed vascular spaces

Our case can be considered to be a low grade histologically, due to prominent vascular spaces, lower cellularity, and <4 mitoses/10 high-power field. Our patient did not receive any postoperative adjuvant treatment and has no evidence of disease recurrence after 1 year of surgery.

Conclusion

Hemangiopericytoma, though rare, should be considered as a differential diagnosis in case of atypical presentation of thyroglossal cyst, in elderly, as in our case. Wide excision should be considered as mainstay of treatment, depending on histopathological features. A case series with larger sample size is essential to expound the same.

Clinical Significance

An atypical presentation of a typical neck swelling must alert the head and neck surgeon to anticipate an uncommon diagnosis, such as a hemangiopericytoma.

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