—Case Reports—

Downhill Varices in the Hypopharynx of a Patient with a Large Thyroid Tumor: A Case Report

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Downhill varices are usually caused by superior vena cava (SVC) obstruction due to bronchogenic carcinoma or mediastinal tumors. These structures exhibit retrograde blood flow and are located in the proximal esophagus. Varices in the hypopharynx resulting from mediastinal thyroid tumor are extremely rare. A 70-year-old man with a 35-year history of a growing thyroid tumor on the right side of his neck visited a local hospital. Fine-needle aspiration cytology of the tumor revealed benign goiter. Contrast-enhanced computed tomography showed a huge tumor (13 × 10 × 5 cm) in the right to left lobe of the thyroid that extended into the mediastinum. A well-enhanced mass mimicking hypopharyngeal cancer was identified in the hypopharynx. Endoscopic examination showed varices in the postcricoid region, so biopsy was contraindicated. The preoperative diagnosis was adenomatous goiter and hypopharyngeal varices caused by obstruction of the internal jugular and brachiocephalic vein by the goiter. Total thyroidectomy was performed and the hypopharyngeal varices had disappeared by the next day. The histopathological diagnosis of the thyroid tumor was poorly differentiated carcinoma. Mediastinal thyroid tumor rarely causes downhill varices due to SVC obstruction. However, signs of SVC obstruction were absent in this case, and varices were present in the hypopharynx, not in the upper esophagus. Obstructed venous flow from the thyroid plexus might circulate via the superior laryngeal vein and cause varices in the postcricoid region. When a patient with a large mediastinal tumor has a tumor-like lesion in the hypopharynx, downhill varices should be considered before scheduling a biopsy. (J Nippon Med Sch 2023; 90: 408–413)

Key words: downhill varix, hypopharynx, superior vena cava obstruction, poorly differentiated thyroid carcinoma, mediastinal thyroid tumor

Introduction

Uphill varices, which develop in the distal esophagus, are a frequent complication of portal hypertension. Downhill varices are usually caused by superior vena cava (SVC) obstruction, which is mainly due to bronchogenic carcinoma and mediastinal tumors. These varices exhibit retrograde blood flow and are located in the proximal esophagus. Varices in the hypopharynx resulting from mediastinal thyroid tumor are extremely rare. We experienced a case of downhill varices due to a large thyroid tumor that was suspected of being a hypopharyngeal tumor but was easily differentiated by laryngoscopy.

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https://doi.org/10.1272/jnms.JNMS.2023_90-601
Journal Website (https://www.nms.ac.jp/sh/jnms/)

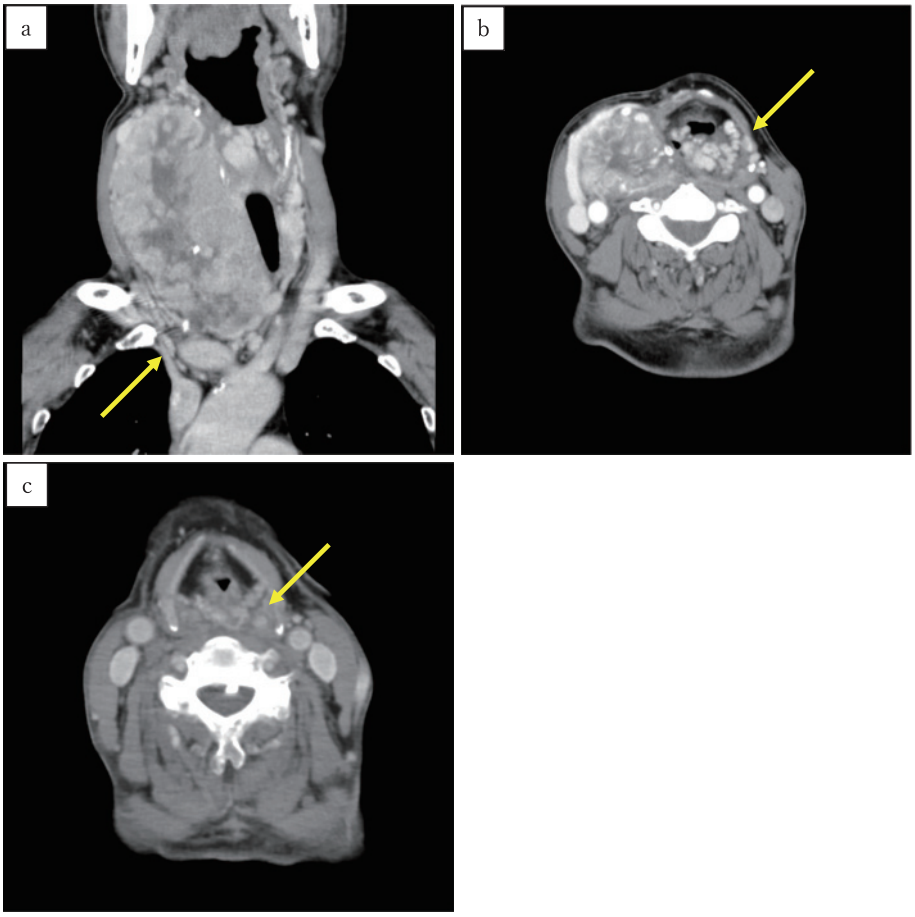

Fig. 1 Contrast-enhanced computed tomography scans of the neck
a) Coronal view. A large tumor (13 × 10 × 5 cm) in the right lobe of the thyroid extends into the mediastinum and obstructs the right internal jugular and brachiocephalic vein (*arrow).
b) Axial view. A well-enhanced mass mimicking hypopharyngeal cancer (*arrow) is present in the posterior hypopharynx.
c) Coronal view after surgery. The varices have disappeared (*arrow).

Case Presentation

A 70-year-old man visited a local hospital for assessment of a large and growing thyroid tumor on the right side of his neck. He first noticed the tumor more than 35 years previously. He had hypertension, but his medical history was otherwise unremarkable. His drinking history was 40 years of drinking one can of beer daily, and he reported smoking 60 cigarettes per day for 40 years. Physical examination revealed a large, firm, nodular mass in the right side of the neck. Laboratory findings, including thyroid function tests, were within normal ranges, except for thyroglobulin (73,336 ng/mL; normal, ≤33.7 ng/mL) and antithyroglobulin antibody (438.9 IU/mL; normal, ≤40 IU/mL). No liver dysfunction was detected. Fine-needle aspiration cytology of the tumor revealed benign goiter. Contrast-enhanced computed tomography (CE-CT) showed a large tumor (13 × 10 × 5 cm) in the right to left lobe of the thyroid. The tumor extended into the mediastinum and obstructed the right internal jugular and brachiocephalic veins (**Fig. 1a, 1b**). Furthermore, a well-enhanced mass mimicking hypopharyngeal cancer was identified in the hypopharynx, and he was referred to our department. Endoscopic examination showed varices in the postcricoid region (**Fig. 2a**) and we therefore avoided biopsy. Gastrointestinal endoscopy was likewise avoided, to prevent any risk of variceal bleeding.

The preoperative diagnosis was adenomatous goiter and hypopharyngeal varices caused by obstruction of the internal jugular and brachiocephalic veins by the goiter. Total thyroidectomy was performed, sparing the bilateral recurrent laryngeal nerves and four parathyroid glands; tracheostomy was not performed. The internal jugular vein and brachiocephalic vein were preserved. Operation time was 223 minutes and total intraoperative blood loss was 995 mL. Veins on the surface of the thyroid gland were markedly distended, causing easy bleeding (**Fig. 3**).

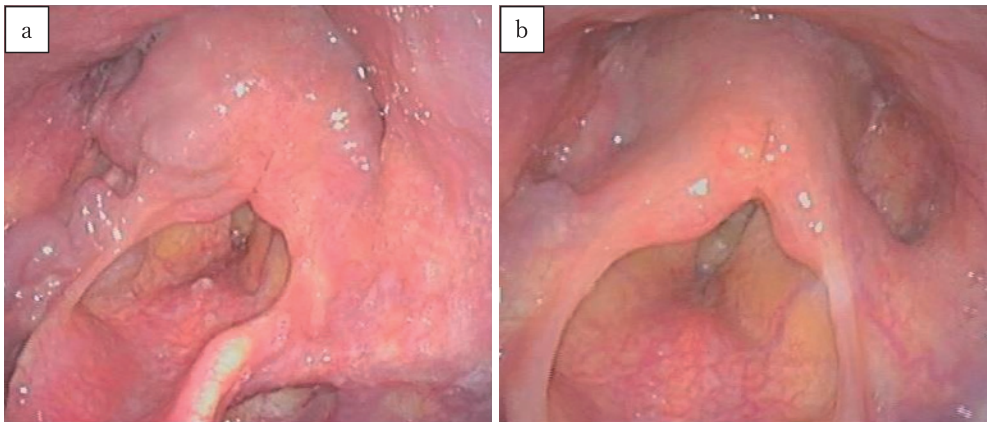

Fig. 2 Endoscopic examination of the hypopharynx
a) Preoperative findings. Varices are evident in the dorsal wall of the hypopharynx. **b**) Postoperative findings. The varices have disappeared on the day after surgery.

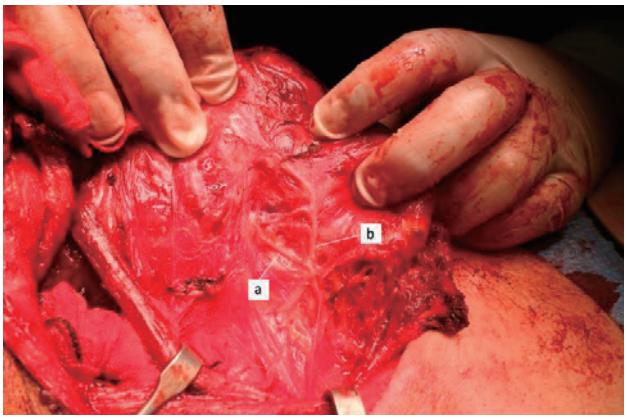

Fig. 3 Intraoperative findings
a) Right recurrent laryngeal nerve. **b**) Right inferior thyroid artery. Veins on the surface of the thyroid gland are markedly distended.

There was no evidence of tumor invasion of surrounding tissue. The thyroid specimen weighed 249 g and the histopathological diagnosis of the thyroid tumor was poorly differentiated carcinoma (**Fig. 4**). Varices in the hypopharynx had disappeared by the next day (**Fig. 1c, 2 b**), and the patient was discharged 5 days after surgery.

Four years postoperatively, a thoracic vertebral metastasis was detected and high-dose radioiodine therapy was administered, but no accumulation in the tumor was identified. At this writing, 6 years postoperatively, the thoracic spine metastases remain but he is asymptomatic and receiving denosumab. The patient declined external-beam radiation therapy.

Discussion

This case of downhill varices due to a large thyroid tumor was suspected of being a hypopharyngeal tumor on CE-CT but was easily differentiated by laryngoscopy. The downhill varices resolved after thyroidectomy.

Downhill varices of upper esophagus are usually caused by SVC obstruction due to bronchogenic carcinoma, mediastinal tumor, surgical caval ligation, central venous catheter therapy, or a thyroid mass[1]. Occlusion of the SVC results in blood flow reaching the esophageal venous plexus via the azygos vein, which causes downhill varices with downward blood flow (**Fig. 5a**). Downhill varices of the upper esophagus can also form without obstruction of the SVC—by obstruction of the inferior thyroid vein, for example, or because of stenosis or hypoplasia of the internal jugular vein[2–4]. In a study of 1,051 patients with cervical and retrosternal goiter, Schmidt reported that 3% of patients developed nonbleeding downhill varices[5]. Lagemann reported development of downhill varices in 4% of goiter cases, 12% of nonrecurrent post-thyroidectomy cases, and 54% of recurrent goiter cases (ie, recurrent goiter from a residual thyroid gland after lobectomy or subtotal thyroidectomy)[3]. These findings suggest that the probability that a mediastinal thyroid tumor will result in downhill varices due to SVC obstruction is 3-4%. A potential reason why varices form in most patients with recurrent goiter is that the inferior thyroid vein, which should function as an outflow vessel for the abundant blood flow from the enlarged thyroid gland, becomes obstructed by ligation or scar formation after surgery, resulting in the development of downhill varices as drainage veins.

Vascular malformation can cause hemangioma in the pharynx[6–8], but for our patient, the primary concern was varices because of the presence of a very large thyroid tumor. Downhill varices of the hypopharynx developed

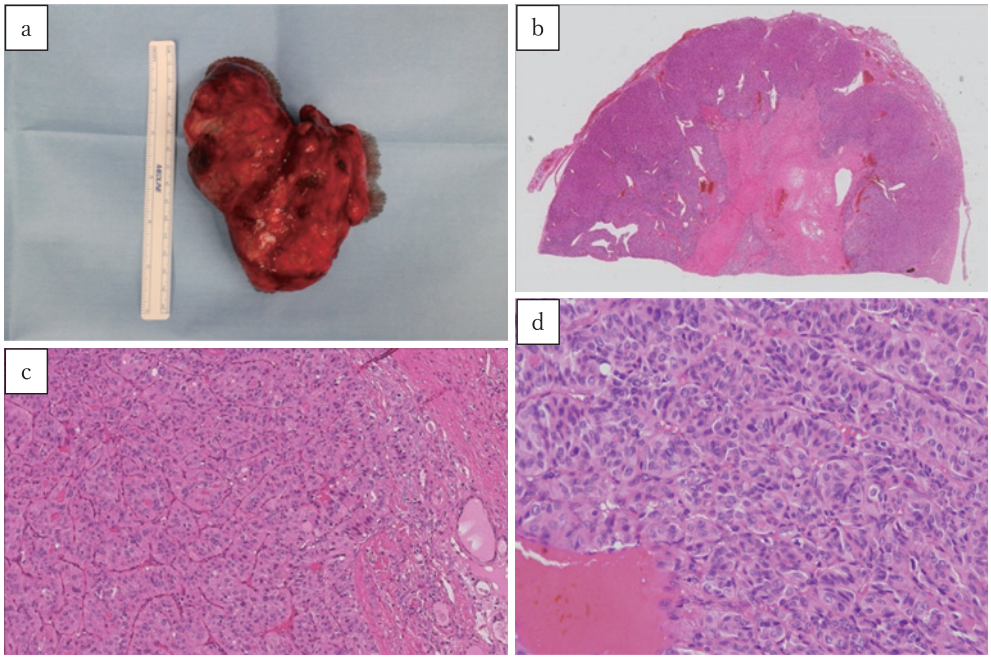

Fig. 4 Histopathological findings
a) Macroscopic appearance of tumor. Gross examination reveals a very large, hard mass measuring 13 × 10 × 5 cm. **b**) Hematoxylin-eosin stain (low-power field). Histologically, the tumor is a massive growth of solid nests with focal necrosis. **c, d**) Hematoxylin-eosin stain (high-power field). Tumor cells are arranged in an insular and follicular pattern, with enlarged atypical nuclei. These histological findings are consistent with a diagnosis of poorly differentiated carcinoma of the thyroid.

without SVC obstruction but with stenosis of the internal jugular and brachiocephalic veins. The inferior thyroid vein was probably also obstructed by pressure from the tumor, thus resulting in the formation of varices due to collateral blood vessels. However, the varices were located in the hypopharynx, not in the upper esophagus. Although we could not directly observe the esophageal mucosa by upper gastrointestinal endoscopy, CE-CT showed no evidence of varices in the upper esophagus. Blood flow to the esophageal venous plexus was probably also blocked by pressure from the tumor. Blood flow from the thyroid gland, which had lost its outflow destination because of occlusion of the internal jugular, brachiocephalic, and inferior thyroid veins, reached the hypopharynx via the superior laryngeal vein and formed varices (**Fig. 5b**).

Unlike uphill varices associated with portal hypertension, downhill varices rarely result in bleeding. Papazian et al.[9] reported a bleeding rate of only 7.6%. The potential reasons for this are as follows. First, in patients with uphill varices, coagulation capacity may be reduced because of concomitant liver disease and an inherent increase in bleeding tendency. Second, exposure to esophagogastric reflux damages distal rather than proximal varices. Third, because distal uphill varices predominantly distend subepithelial levels rather than the submucosal location of downhill varices in the midthoracic and proximal esophageal wall, variceal rupture near the esophagogastric junction is much more likely[10]. We identified 12 reports of hemorrhagic downhill esophageal varices related to thyroid tumors but no cases of death due to hemorrhage[2,4,10–19]. In 10 of those 12 cases (excluding one patient treated with esophageal balloon tamponade[10] and one patient treated with sclerotherapy[16]), varices resolved after thyroidectomy. Similar to the present case, there was also a report of downhill varices that was detected without bleeding[20].

In our patient, fine-needle aspiration cytology revealed benign tissue, but the pathological diagnosis was poorly differentiated carcinoma. Although the preoperative diagnosis was a benign tumor, the blood thyroglobulin level was very high, and follicular thyroid carcinoma could not be ruled out, so total thyroidectomy was performed. The presence of distant metastasis also suggested that the tumor was a poorly differentiated thyroid carcinoma derived from follicular thyroid carcinoma.

Surgical thyroidectomy is the standard treatment for varices of the hypopharynx, as the mechanism of forma-

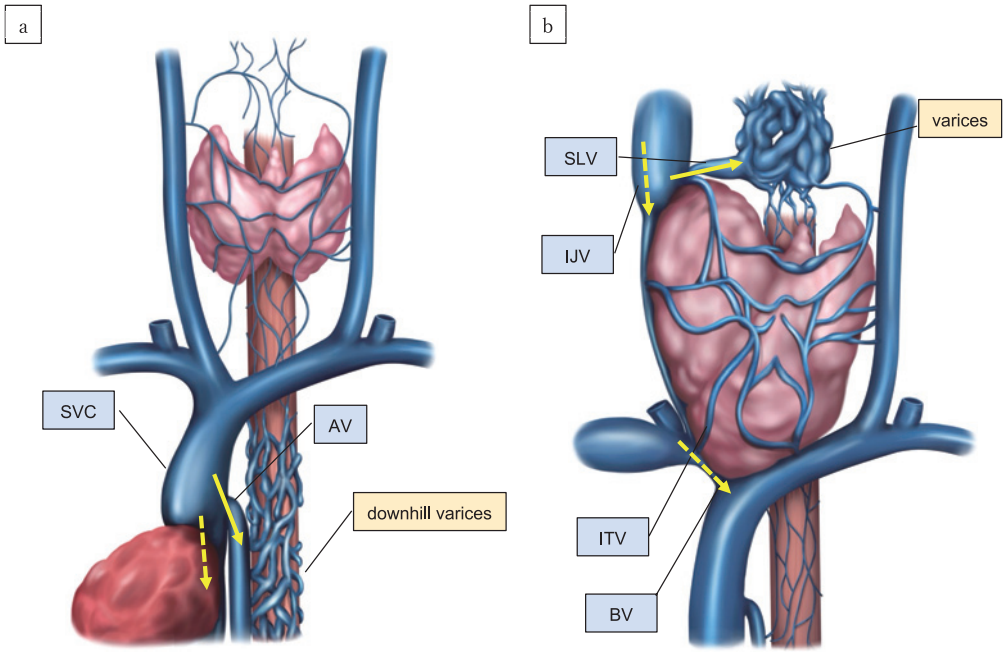

Fig. 5 Mechanism of downhill varices

a) Typical downhill varices. Because of occlusion of the superior vena cava (SVC), obstructed blood flow likely reached the esophageal venous plexus via the azygos vein (AV).

*dashed arrows, obstructed blood flow

**solid arrows, blood flow

b) Downhill varices in the hypopharynx. Blood flow from the thyroid gland, which had lost its outflow destination because of occlusion of the internal jugular vein (IJV), brachiocephalic vein (BV), and inferior thyroid vein (ITV), reaches the hypopharynx via the superior laryngeal vein (SLV) and forms varices.

*dashed arrows, obstructed blood flow

**solid arrows, blood flow

tion for varices in such cases appears to be similar to that for downhill varices. If a varix is misdiagnosed as a neoplastic lesion and puncture aspiration cytology or biopsy is performed, the risk of serious bleeding is substantial. Adequate observation by laryngeal fiberscopy can identify varices, and dangerous cytological examinations can thus be avoided.

Conclusions

Mediastinal thyroid tumor rarely causes downhill varices due to SVC obstruction. However, in the present case, signs of SVC obstruction were absent and varices were present in the hypopharynx, not in the upper esophagus. Obstructed venous flow from the thyroid plexus might circulate via the superior laryngeal vein and cause varices in the hypopharynx. When a patient with a large mediastinal goiter displays a tumor-like lesion in the hypopharynx, downhill varices should be considered before conducting a biopsy.

Acknowledgements: We are grateful to Dr. Chihoko Yamagata, of the Department of Design for Medical and Health Care, Kawasaki University of Medical Welfare, Okayama, Japan, for creating the beautiful medical illustrations.

Conflict of Interest: None declared.

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(Received, March 14, 2022)
(Accepted, July 8, 2022)
(J-STAGE Advance Publication, October 21, 2022)