

Case Report

Oesophageal varices in a case of compressive goiter

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SUMMARY

Downhill oesophageal varices are venous submucosal dilations developing in the upper oesophagus, draining upper body venous flow “downwards” and usually bypassing superior vena cava obstruction. Extrinsic compression from a thyroid goiter is a rare cause with only 18 cases reported in the literature. We report the case of an 84 year old woman with a long history of multinodular goiter admitted for the exploration of severe iron-deficiency anemia. She had no history of hematemesis or melena. The endoscopic exam found grade II upper oesophageal varices with no sign of active bleeding, as well as a non hemorrhagic gastric ulcer. No other obvious explanation for anemia could be found. Goiter investigation included a cervical and thoracic CT scan which evidenced a voluminous thyroid goiter with massive endothoracic extension, causing oesophageal compression. The optimal treatment was considered to be radioiodine therapy followed by surgery. Downhill oesophageal varices are a rare clinical entity seen in patients with goiter. We propose a clinical observation of this pathology in a patient explored for chronic anemia.

Key words: downhill oesophageal varices, goiter, digestive bleeding, anemia

INTRODUCTION

Oesophageal varices are typically associated with portal hypertension and are a primary cause of severe upper

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gastrointestinal bleeding. Less frequently, downhill oesophageal varices (DEV) are described in cases of superior vena cava obstruction usually associated with mediastinal masses. We report the case of an 84 year old woman with severe anemia. Downhill oesophageal varices related to external compression from an endothoracic multinodular goiter were found.

CASE REPORT

An 84 year old female was admitted for investigation of severe anemia diagnosed in a clinical context of asthenia. She had an 8 year history of a slow-growing multinodular cervical goiter with no reported difficulty in swallowing or breathing. Her medical history included chronic pleurisy secondary to earlier asbestos exposure and chronic hypertension. Her medication consisted of neomercazole and verapamil. When physically examined, she was pale and presented a painless multinodular cervical goiter. (Fig. 1) The superficial venous circulation was highly distended in the cervical area as well as on the anterior upper thoracic wall (Fig. 2). There was no overt bleeding and no clinical signs of portal hypertension.

Laboratory findings included severe normocytic anemia with low iron levels and normal-low ferritin levels. Thyroid function tests were within normal limits even after neomercazole was discontinued. All other laboratory test values, including liver function tests and prothrombin levels, were normal (**table 1**). Finally, the pathogenic investigation of her anemia was completed with tests for occult bleeding. The upper digestive endoscopy revealed upper oesophageal grade II varices with no red spots or other endoscopic signs of recent bleeding. The presence of a non hemorrhagic gastric ulcer was also noted. A colonoscopic examination did not evidence any hemorrhagic lesions. The abdominal ultrasound showed the liver, spleen

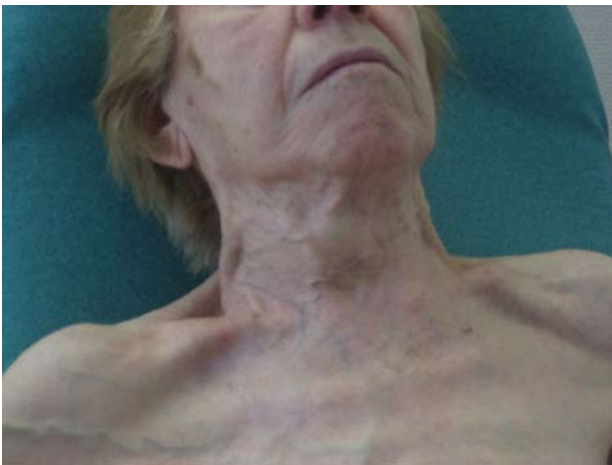


Figure 1. Voluminous cervical goiter



Figure 2. Collateral venous circulation involving both cervical and upper thoracic area

and portal vein to be normal in size, thus providing no evidence for portal hypertension. Workup for her goiter included a cervical and thoracic CT scan which showed a voluminous thyroid goiter (with an estimated volume of 525 cc³) filling the entire cervical area and extending into the upper mediastinum, causing oesophageal compression (Fig. 3, 4). As radioiodine therapy was proposed, we performed an ultrasound-guided thyroid biopsy which confirmed the non malignant nature of the goiter.

The patient had received blood transfusions, long-term proton pump inhibitor treatment and triple therapy to eradicate *H. pylori*. During her stay in hospital, we observed two episodes of acute respiratory distress without any acute pulmonary or cardiac concomitant disease. Given the position-related nature of her dyspnea, the logical

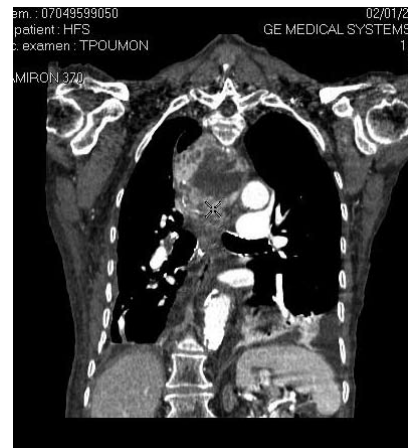


Figure 3, 4. Thoracic CT scan showing an important mediastinal goiter with oesophageal compression

explanation seemed to be sudden tracheal compression caused by her extremely large goiter.

After consultation with the thoracic surgeons, rapid surgery to remove this goiter was deemed necessary. The patient was extremely reluctant to undergo open thoracic surgery and radioactive iodine therapy was proposed until her consent for the intervention was obtained. Unfortunately, she died soon afterwards of an episode of acute respiratory failure most likely caused by the tracheal compression described above.

DISCUSSION

During investigation of severe anemia in an elderly patient with no gastrointestinal signs, we discovered varices in the upper third of the oesophagus. Downhill oesophageal varices (DEV) are submucosal venous dilatations that arise most frequently in response to a superior vena cava obstruction. They direct the blood flow away from the superior venous circulation to the azygos vein when the superior vena cava is blocked or even to the portal circulation when the azygos vein is also involved in the obstruction.¹ The clear pathophysiological distinction between “regular” portal varices and downhill oesophageal varices was

Table 1. Laboratory findings

Laboratory test	Patient's results	Normal range
RBC	2.05	2.8-5.8 10 ⁶ /l
Hemoglobin	6.1	11.5-16 g/dl
MCV	93	80-100 fl
MCHC	29.7	27-32 pg
Platelet count	378	150-450 10 ³ /l
Reticulocytes	87	25-100 10 ³ /l
C reactive protein	3	<5 mg/l
Ferritin	13	3-105 µg/l
Iron level	1	10-26 µmol/l
Folic acid	5	3-17 µg/l
Vitamin B12	198	150-980 ng/l
GPT	13	<31 UI/l
GOT	10	<34 UI/l
PT	78%	80-100%
TSH	1.44	0.2-4 mUI/l

first acknowledged by Felson et al. in 1964.² Although accessibility to upper endoscopy and venous Doppler studies has facilitated the diagnosis of this particular entity, in 1983 there were only 119 cases described in the literature.³ No major incidence study was conducted after these cases and the remaining DEV reported as isolated cases were almost exclusively linked to superior vena cava syndrome.^{4,7} The presence of downhill oesophageal varices in cases of superior vena cava syndrome appears to be related both to the duration of compression and to the localization of the obstruction. The overwhelming majority of downhill varices are described in different SVC syndromes related to pulmonary and breast cancers, lymphomas, thymomas, mediastinal fibrosis and Behcet disease, or even as a complication of a hemodialysis catheter.^{3-8,10}

In the case reported here, we discovered downhill varices in a patient with a large goiter and no other mediastinal pathology. The literature reports 18 cases of DEV of thyroid origin, most of which are related to benign recurrent goiters.⁹ A cervical goiter can cause direct compression on the jugular vein leading to the development of collateral circulation. In addition, the inferior thyroid veins usually drain into the brachiocephalic veins thus promoting formation of an oesophageal submucosal variceal system which is "forced" to bear the high systemic circulation pressures. Mediastinal goiters are not different from any mediastinal tumors acting as direct compressive masses on the superior vena cava leading to SVC syndromes of benign etiology.

In our patient, DEV was diagnosed as a result of a systematic search for occult gastrointestinal bleeding in a case of unexplained iron deficiency anemia. As with uphill portal varices, the major complication of DEV is upper gastro-

intestinal hemorrhage. All the cases reported confirm that the upper gastrointestinal bleeding from DEV is both less severe and less frequent than that related to uphill portal varices. In a Spanish series reported in 2005, DEV represented only 0.1% of all variceal bleeding.¹⁴

Our patient was a clear candidate for surgical treatment of her goiter. The treatment of DEV is based on the same principles as that proposed for "regular" varices and includes endoscopic band ligation in cases of recurrent bleeding.¹³⁻¹⁵ The prognosis is better than in the case of cirrhotic varices because of the correctable underlying pathology. In eight of the ten cases of DEV secondary to benign thyroid compression, surgical removal of the goiter led to complete regression of the oesophageal varices.⁹ Given the unexpected death of our patient, we were unable to report a similar result.

The patient in the case presented here had chronic anemia and a slow-developing thyroid goiter. The endoscopic confirmation of downhill oesophageal varices allowed us to establish a relationship between the pre-existing goiter and the varices. The patient had a goiter, whose compressive character was clinically suggested by the impressive superficial collateral circulation involving all cervical and upper thoracic area (Fig. 1, 2) and was later confirmed in morphological studies.

Endoscopy showed that she also had a peptic ulcer. Although we found no endoscopic signs of active bleeding, we were more inclined to consider this lesion to be responsible for the anemia. Both the colonoscopy and abdominal scan were normal and we were unable to find any other explanation for her anemia. A more thorough small bowel study wasn't performed.

The discovery of DEV in this particular context of anemia suggests the possibility of a relationship between the slowly developing anemia and the varices observed. The sparse data in the literature suggest that DEV bleeding is not as severe as portal variceal bleeding. However, there are no reports of related occult digestive bleeding. Consequently, is it reasonable to consider that the anemia is caused by intermittent variceal bleeding or should the blame be laid entirely on the peptic ulcer, which is a well known cause of occult bleeding? The fact that the peptic ulcer did not respond to treatment was an interesting argument in favour of this but, on the other hand, we were not able to prove that the patient's anemia regressed after she received treatment for oesophageal varices.

CONCLUSION

Downhill oesophageal varices are a rare clinical entity. They should always be taken into consideration in the standard diagnostic workup for patients with SVC syndrome. Although much less common, DEV should be considered in cases of thyroid goiter particularly when there are other clinical signs of compression such as voluminous goiter, chest wall collateral veins, tracheal dyspnea or dysphagia. Their most serious complication is upper gastrointestinal bleeding usually manifesting as hematemesis or melena.

Recognizing this rare pathology is particularly relevant because in most cases treatment is successful, resulting in resolution of the varices after the goiter has been surgically removed.

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