Abstract. Double aortic arch (DAA) is an extremely rare vascular malformation which causes tracheal and esophageal compression, resulting in respiratory symptoms such as stridor and wheezing, or feeding problems such as dysphagia, usually during the first few months of life. In contrast, this disorder is rarely diagnosed in adults. We herein present an elderly case with thoracic esophageal carcinoma with DAA. To the best of our knowledge, this is only the second report of a successfully resected case of esophageal cancer associated with DAA in the English literature. Moreover, this is the first reported case of symptomatic DAA with esophageal carcinoma. Spiral computed tomography and three-dimensional reconstruction was very useful for preoperative assessment of the abnormal vessels.

Case Report

A 70-year-old man with a complaint of several months’ dysphagia visited a local physician’s office, where an esophageal tumor was detected by esophagoscopy. The patient was referred to our hospital for further examination. The patient had no breathing problems. His clinical examination was normal, and a chest X-ray demonstrated a right aortic arch with normal cardiac findings (Figure 1a). Esophagoscopy revealed an elevated tumor with irregular surface, occupying two-thirds of the esophageal wall between 35 cm and 39 cm from the incisors (Figure 1b). The histological diagnosis of the biopsy samples from the tumor revealed poorly-differentiated squamous cell carcinoma. An esophagogram revealed bilateral indentation of the upper thoracic esophagus due to the presence of right and left aortic arches and a 4-cm in diameter esophageal tumor located in the middle thoracic esophagus (Figure 2).

Chest-contrasting computed tomography showed a double aortic arch (DAA) and -descending aorta on the right side of the thoracic vertebrae. The trachea and esophagus were encircled with the right and left aortic arch, the descending aorta and the ascending aorta (Figure 3). The right aortic arch was larger than the left arch, while both aortic arches were patent. No esophageal tumor or metastatic lymph nodes were detected using this CT modality.

Three-dimensional reconstruction images of CT (3D-CT) showed that the ascending aorta was divided into a right anterior arch and a left posterior arch, which encircled the trachea and esophagus and joined posteriorly to form the descending aorta (Figure 4). The right carotid artery and subclavian artery originated from the right aortic arch. The left aortic arch gave rise to the left carotid artery and subclavian artery. The left aortic arch crossed the thoracic esophagus posteriorly and the descending aorta was on the right side of the thoracic vertebrae. The middle and lower thoracic esophagus was located on the left side of the descending aorta.

The patient was diagnosed with thoracic esophageal carcinoma accompanied by a DAA and underwent radical esophagectomy and three-field lymphadenectomy through a left thoracotomy.

Operative findings. A thoracic approach was followed by a left posterolateral thoracotomy via the fourth intercostal space. The DAA was recognized on preoperative 3D-CT. The trachea and esophagus were completely encased by a vascular ring consisting of the bilateral aortic arch and the descending aorta. The left recurrent laryngeal nerve passed behind the left aortic arch and ascended posteriorly (Figure...
We performed subtotal esophagectomy and radically dissected the paraesophageal, mediastinal, neck, and perigastric lymph nodes. Reconstruction was carried out by gastric conduit via the retrosternal route. The lymph nodes along the right recurrent laryngeal nerve were dissected through the neck. We did not perform division and ligation of the lesser aortic arch because there were no findings of tracheal compression of the bilateral aortic arch.

The postoperative course was uneventful except for minor leakage. Macroscopic and microscopic examination demonstrated an esophageal carcinoma, measuring 4.0×4.5 cm, which had invaded the submucosal layer. Two metastatic lymph nodes, one along the right recurrent laryngeal nerve and one at the supradiaphragm, were diagnosed histologically. The pathological stage was IIB (T1bN1M0) according to the TNM classification of the International Union Against Cancer version 7 (1).

Discussion

DAA is a congenital vascular anomaly and a common form of complete vascular ring, causing tracheal and esophageal compression. The ascending aorta is divided into left and right aortic arches, passing to each side of the trachea and the esophagus. Both arches are usually patent, although one is a usually larger than the other, the right arch being dominant in 70% of cases (2). The first report of symptomatic DAA was reported by Wolman in 1939 (3) and the first successful surgical treatment was published by Gross in 1945 (4). This abnormality tends to cause respiratory symptoms such as stridor and wheezing, which often require surgery in infancy or early childhood. By contrast, this anomaly is very rarely found in adults and fewer than 30 cases with symptomatic DAA have been reported during the last 10 years (5). Surgical repair of
symptomatic DAA is typically performed by division of the small atretic arch, which is the left arch in the majority of the cases, and also by ligation and division of the ductus arteriosus or ligamentum arteriosus, with mobilization of the trachea and esophagus (6).

We encountered a patient with esophageal cancer associated with DAA; to the best of our knowledge, this is the only second report of successful surgical treatment of esophageal carcinoma with DAA. In this case, the symptoms, such as dysphagia and nausea, were considered to have been caused not by the esophageal tumor itself, but by the DAA because the esophagography revealed bilateral indentations of the upper thoracic esophagus and the barium passage around the tumor was very smooth. Matano et al. (8) reported the first case of esophageal cancer accompanied by a DAA; the reported case was asymptomatic. Our case is the first case of symptomatic DAA with esophageal carcinoma.

In Japan, standard surgical treatment for patients with esophageal carcinoma involves esophagectomy, usually through a right thoracotomy, and radical three-field (neck, thorax and abdomen) lymphadenectomy (7). Resection of the lymph nodes along the right and left recurrent laryngeal nerve in the upper mediastinum, which have a high incidence of metastasis, is considered to be critical. Matano et al. (8) chose a right thoracotomy because it was a convenient routine approach, and considered that the left recurrent laryngeal nerve would be found more easily through a right thoracotomy because the right aortic arch was situated more cephalad than the left aortic arch, which meant that the aortic window on the right side was wider than that on the left side. However, we selected to undertake a left thoracotomy for this patient because the thoracic esophagus was located on the left side of the descending aorta. If a right thoracotomy had been chosen, the presence of the descending aorta would interfere with the mobilization of the esophagus and the lymphadenectomy. A left thoracotomy provided an excellent view of the thoracic cavity, and the left recurrent laryngeal nerve, which turned around at the lower edge of the left aortic arch and ascended along the tracheoesophageal groove, was identified and preserved.

Kinoshita et al. (9) reported a case of thoracic esophageal cancer associated with a right aortic arch (RAA). This patient underwent esophagectomy and mediastinal lymph node dissection via a left thoracotomy, along with a median
sternotomy for lymph-node dissection along the right recurrent laryngeal nerve. They stated this approach provided the surgical team with a good surgical view along the nerve and that a median sternotomy should be added to the left thoracotomy in cases with advanced thoracic esophageal cancer with RAA. In our case, the preoperative work-up showed no evidence of metastasis in the lymph nodes along the right recurrent laryngeal nerve. Moreover, this nerve was identified and preserved via neck incision, and the lymph nodes along its route were clearly dissected. Thus, in this case we managed to avoid sternotomy, which is considered to be very invasive and stressful for the patient.

To perform an operation safely in the presence of these vascular anomalies, it is very important to have a precise understanding of the vessel anatomy. Spiral CT and 3-D reconstruction imaging, a new technique, has now made possible rapid and continuous data collection, providing data that can be reconstructed to give 3-D images. This modality is less invasive and was useful as a preoperative vascular

Figure 4. Three-dimensional construction images of CT (3D-CT): the frontal view (a), the left lateral view (b), the back view (c). The right aortic arch and left aortic arch joined posteriorly and the right-sided descending aorta was found. RAA: Right aortic arch, LAA: left aortic arch, DA: descending aorta.

Figure 5. Intraoperative view at the upper mediastinum through the left thoracotomy. The left recurrent laryngeal nerve passed behind the left aortic arch and ascended posteriorly. RAA: Right aortic arch, LAA: left aortic arch, DA: descending aorta, LRLN: left recurrent laryngeal nerve, LSA: left subclavian artery, E: esophagus.
assessment of this patient. Indeed, this excellent modality could replace angiography because of its non-invasiveness and its provision of a good view through any angle.

In conclusion, we experienced a case with thoracic esophageal cancer accompanied by DAA and successfully performed an esophagectomy with extensive lymphadenectomy through a left thoracotomy. Preoperative 3D-CT imaging was considered to be a less invasive examination and was a helpful modality for understanding the complicated anomalies of the great vessels.

References