Esophageal Cancer Associated with Right Aortic Arch: A Case Study

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Abstract. *Right aortic arch is a rare congenital vascular anomaly, thus, the surgical resection of esophageal cancer in a patient with this anomaly is extremely uncommon. The surgical treatment of esophageal cancer was performed in a 73-year-old man with right aortic arch. An upper GI series and endoscopy revealed Type 2 esophageal cancer in the lower thoracic esophagus. Chest computed tomography revealed a right aortic arch with an aberrant subclavian artery and was classified as an aortic arch anomaly of Type IIIB according to the Edwards' classification. The middle and lower portions of the intrathoracic esophagus were resected through a left thoracoabdominal incision. Type IIIB is the most common form of right aortic arch. For surgical resection, a left thoracotomy was the most common method. It is difficult to adequately visualize the right recurrent laryngeal nerve through a left thoracotomy alone and an additional incision, i.e., a midline sternotomy, may be necessary.*

Right aortic arch is a rare condition and is reported to affect 1 - 2 per 1,500 persons in Japan (1) or 0.03 - 0.04% of autopsy cases in Western countries (2). Consequently, esophageal cancer surgery is very rarely performed in patients with right aortic arch and only 27 such cases have been reported to date (3-19). In patients with a right aortic arch, the aorta surrounds the esophagus and trachea, causing the compression and deviation of these structures, as well as deviation of the recurrent laryngeal nerve. Because of such deviations and compression, great care must be taken when esophageal cancer is treated surgically in these patients. Recently, we performed esophagectomy for esophageal cancer in a patient with right aortic arch. This case is reported here with discussion of the relevant literature.

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Case Report

A 73-year-old man noted difficulty with swallowing in May 2004, and consulted a local physician. He was referred to our department after a diagnosis of esophageal cancer was made from an upper GI series.

Physical findings. The patient was undernourished, but there was no anemia or jaundice. Palpations the neck, chest and abdomen were normal.

Laboratory tests on admission. The hematology and biochemistry tests gave normal results and the tumor markers (SCC and CYFRA) were within the normal range.

Chest X-ray film. The aortic arch was on the right side (right aortic arch; Figure 1).

Upper GI series. A 5-cm long irregular ulcerated lesion with marginal elevation was seen in the lower thoracic esophagus, involving almost half of the esophageal wall circumference. This was diagnosed as esophageal cancer and the extent of tumor invasion was estimated to be T2 or T3. The right aortic arch was located behind the upper thoracic esophagus and pushed it forward (Figure 2).

Esophagoscopy. An ulcerated localized lesion which covered almost half of the circumference of the esophageal wall was seen at a point 30 - 35 cm from the incisors.

Chest computed tomography. A 5-cm long circumferential wall-thickness of the esophagus was recognized in the lower thoracic esophagus. There were no metastases in the lungs or lymph nodes. Because the presence of a right aortic arch was demonstrated with an aberrant subclavian artery, it was classified as an aortic arch anomaly of Type IIIB according to the Edwards' classification (or Type II according the Stewart classification) (20). A concomitant small Kommerell diverticulum was also recognized (Figure 3).

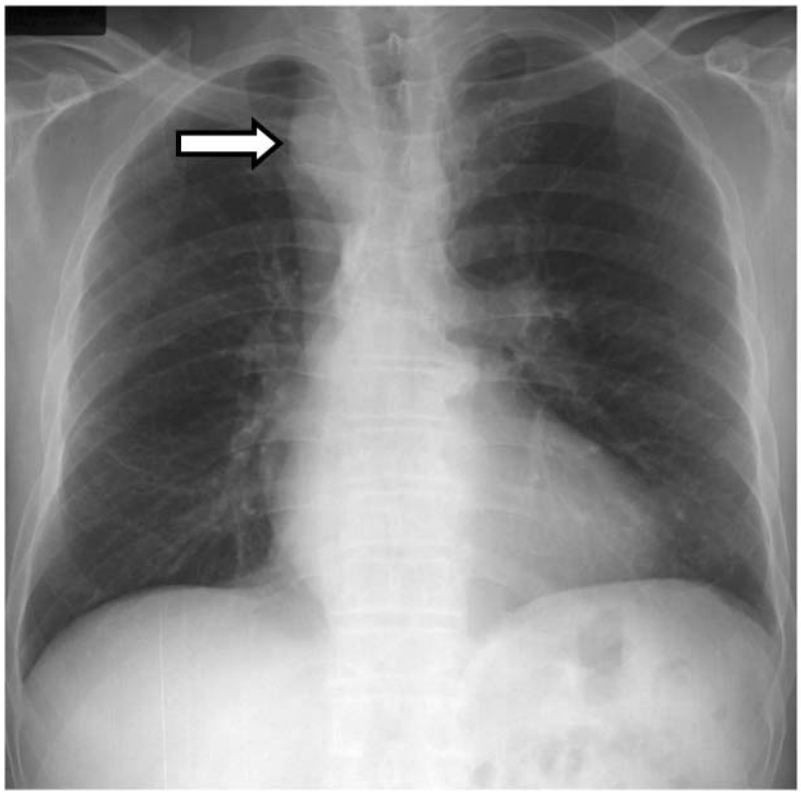


Figure 1. *Chest X-ray. The aortic arch was located on the right side (⇨).*

Abdominal computed tomography. No metastases in the liver or lymph nodes were demonstrated.

From the findings described above, a diagnosis was made of advanced esophageal cancer (T2N0M0, stage IIA by UICC) associated with right aortic arch. Surgery was performed in July 2004.

Surgical procedure. The patient underwent subtotal esophagectomy and regional lymphadenectomy *via* left thoracotomy and laparotomy. Intrathoracic esophago-gastrostomy was performed for reconstruction.

Surgical findings. The esophagus was exposed under an incision made in the parietal pleura of the superior mediastinum. The left recurrent laryngeal nerve was identified as it ran around the left ductus arteriosus and was preserved when the left recurrent laryngeal nerve lymph node chain was dissected.

The right recurrent laryngeal nerve could not be identified through the pleural incision. Consequently, additional midline sternotomy would have been needed to perform right recurrent laryngeal nerve lymphadenectomy for the operation to be as curative as possible. Because of the patient's advanced age and the operative finding that there were no lymph node metastases, however, this lymphadenectomy was not performed (Figure 4).*Histological findings.* The histological diagnosis was poorly-differentiated squamous cell carcinoma of the esophagus (pT3, p N0, pStage IIA) and the operation was curative.

The post-operative course of the patient was uneventful. On post-operative day 2, the endotracheal tube was extracted. On post-operative day 10, the patient began to take food orally. About one month after surgery, he was discharged. At 1.5 years after surgery, there has been no sign of recurrence.

Discussion

Right aortic arch is a congenital vascular anomaly. The classification of aortic anomalies by Edwards *et al.* (20, 21) is based on their theoretical concept of the development of the aortic arch (Figure 5). Right aortic arch belongs to Group III of this classification. Its variations are divided into three types and our case was Type IIIB. According to a similar classification proposed by Stewart (22), however, this case is Type II (Figure 6). This is the most common type of right aortic arch and is seldom associated with congenital heart disease. Each type of right aortic arch is further divided into three subtypes by Stewart, depending on the position of the ductus arteriosus. Our case was subtype IIIB1 because the left recurrent laryngeal nerve ran around one side of the left ductus arteriosus. The major symptoms of right aortic arch

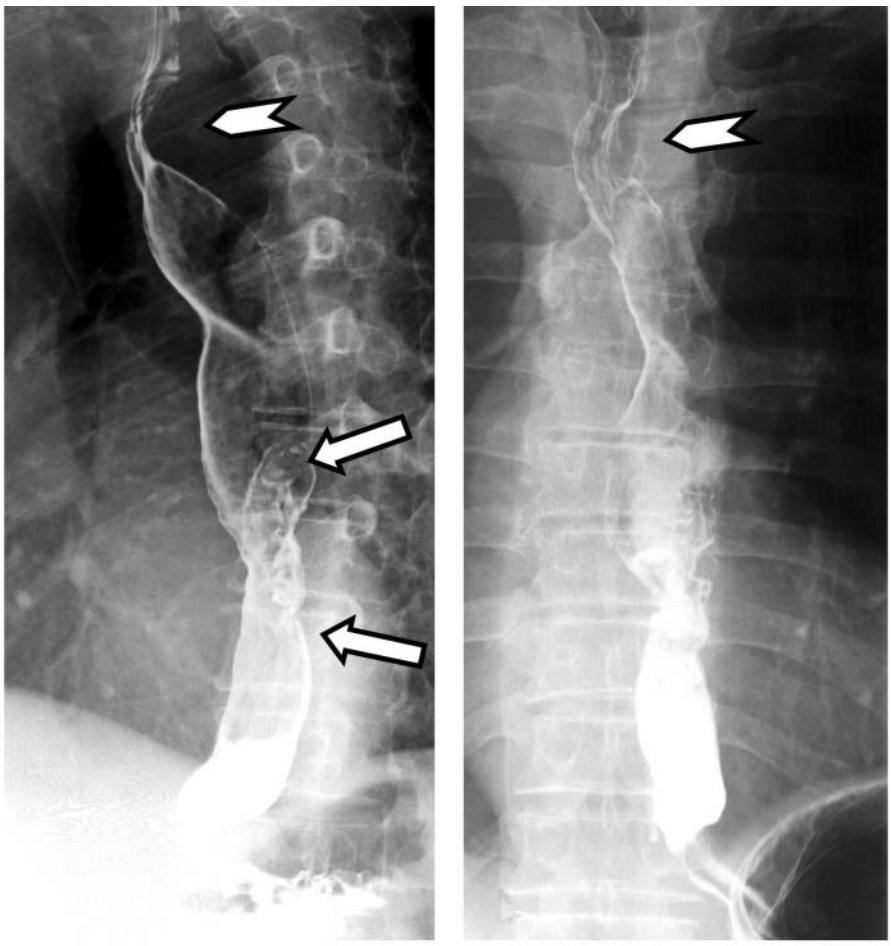


Figure 2. *Upper GI series. A 5-cm long irregular ulcerated lesion with marginal elevation was seen in the left of the lower thoracic esophagus and was diagnosed as esophageal cancer (T2 or T3) (⇨). The right aortic arch pushed the upper thoracic esophagus forward (⇶).*

include dysphagia due to compression of the esophagus by the vascular ring that the arch creates. In our patient, however, dysphagia was not noted before the development of esophageal cancer.

Our search of the literature revealed only 27 of esophageal cancer resection in patients with right aortic arch, not including our case, indicating that this is an extremely uncommon combination (Table I). The 28 patients ranged from 30 to 78 years in age (mean: 63.3 years), consisting of 26 men and one woman (gender was not mentioned in one report). According to the Edwards' classification, the most common type was Type IIIB (as in our case) in 19 patients, followed by Type IIIA in 8 patients. The type was unknown in one patient. The most common approach for resection of esophageal carcinoma was left thoracotomy in 20 patients, followed by a combination of left thoracotomy and midline sternotomy in 4, the left open door method in 2 and bilateral thoracotomy and the transhiatal esophageal approach in 1 case each. Although the most common approach for resection of esophageal cancer is right thoracotomy, it was not used in any of these 28 patients. This spectrum of surgical procedures reflects the anatomical characteristics of right aortic arch. It is difficult to identify the right recurrent laryngeal nerve *via* left thoracotomy alone. Because lymph nodes around both recurrent laryngeal nerves are common sites of metastasis, however, to make the operation as curative as possible, midline sternotomy should be added or a left open door method should be used. In our patient, no lymph node metastases were recognized before or during the operation. If he had been younger in age and in better physical condition, however, a midline sternotomy should have been added.

Conclusion

We treated esophageal cancer in a patient with right aortic arch, a very rare combinationd. Deviation of the recurrent

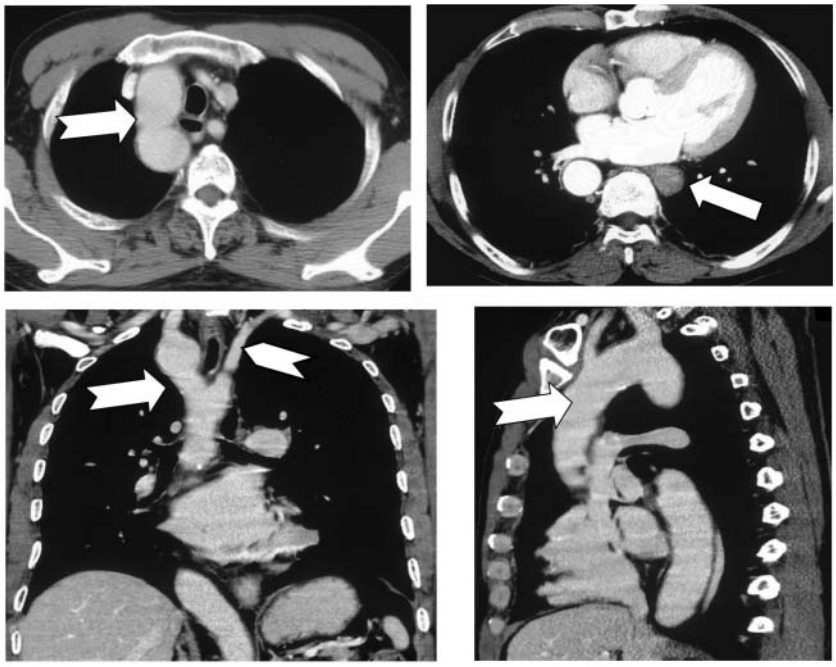


Figure 3. *Chest computed tomography. A 5-cm long circumferential wall-thickness of the esophagus was recognized in the lower thoracic esophagus (⇨). The right aortic arch (⇨) was demonstrated with an aberrant subclavian artery (⇨).*

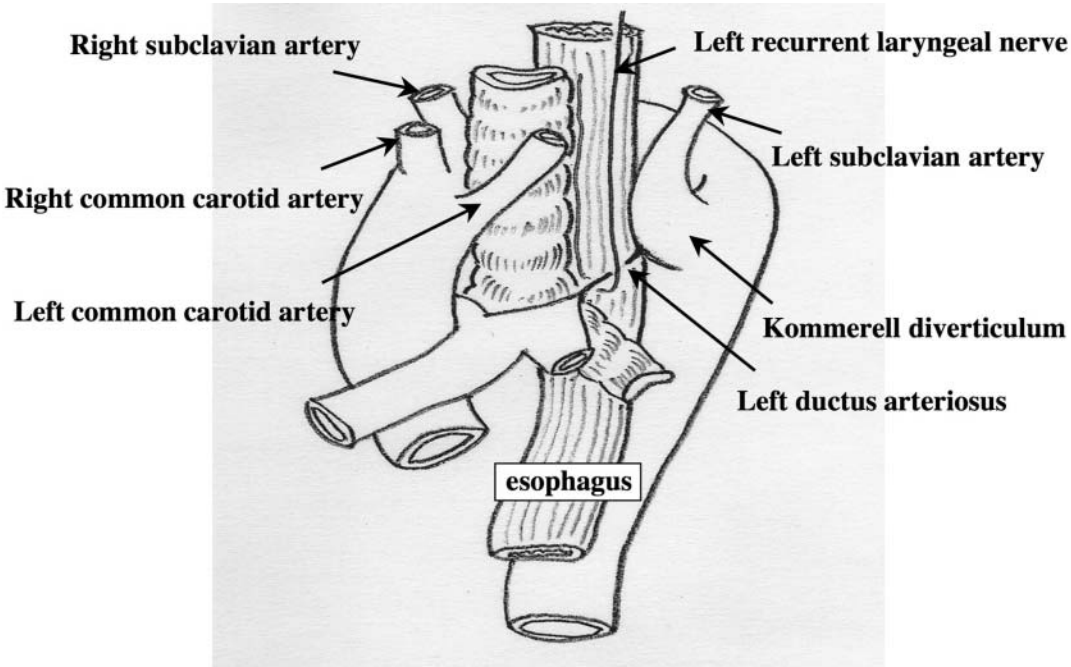
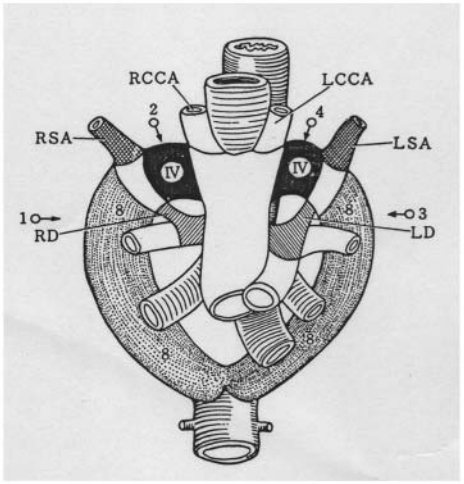


Figure 4. *Schematic illustration of the surgical findings.*

laryngeal nerve associated with right aortic arch should be taken into consideration when such patients undergo surgery for esophageal cancer. To allow complete lymphadenectomy in the superior mediastinum, a midline sternotomy or another incision is necessary to provide good access.



The theoretical schematic picture of the development of the aortic arch. (by Edwards)

Edwards's classification of the aortic arch

I	double aortic arch (Neithers does regression)	A	both aortic arch patency
		B	one side aortic arch close
II	Left aortic arch (arrow 1 or 2 is regression)	A	nomal branch
		B	aberrant rt.subclavian artery
		C	isolation of rt.subclavian artery
III	Right aortic arch (arrow 3 or 4 is regression)	A	mirror image branch
		B	aberrant lt.subclavian artery
		C	isolation of lt.subclavian artery
IV	others		

Each subgroup is classified in side of ductus arteriosus.
1: lt. ductus arteriosus
2: rt. ductus arteriosus
3: bil. ductus arteriosus

Figure 5. *The classification of malformation of the aortic arch.*

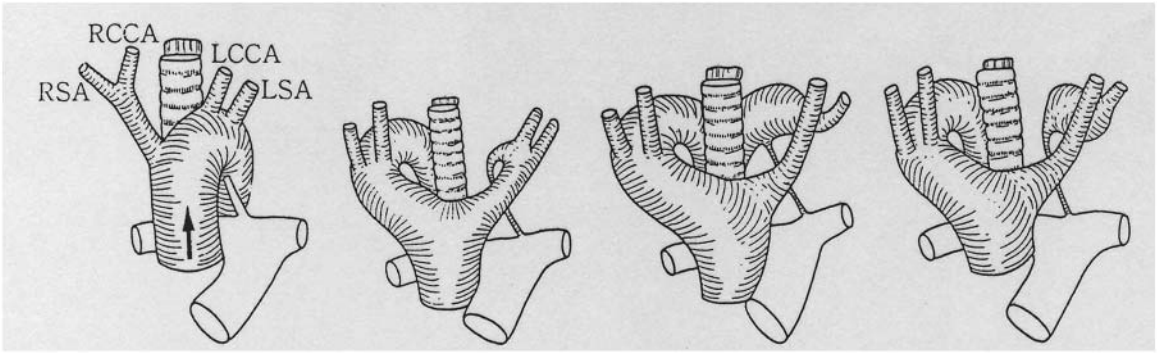


Figure 6. *Stewart's classification of right aortic arch.*

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Table I. *Reported cases of resected esophageal cancer with right aortic arch.*

Case	Year	Author	Age	Gender	Location	Type	Approach (Edwards)	Reconstruction (thoracotomy)	Stage	Recurrent nerve	
										Left	Right
1	1974	Soma *et al.* (3)	62	M	Mt	IIIA1	Left	retrosternal/eso-gastric	III	0	-
2	1989	Ito *et al.* (4)	-	-	-	IIIB1	Left	-	-	-	-
3	1992	Tanaka *et al.* (14)	56	M	Mt	IIIB1	Left	-	-	-	-
4	1993	Nakamura *et al.* (6)	56	M	Mt	IIIB1	Left	retrosternal/eso-gastric	IVb	Ductus arteriosus	-
5	1993	Nakamura *et al.* (6)	52	M	Ut	IIIA1	Left	post. mediastinal/eso-gastric	I	Ductus arteriosus	-
6	1997	Terasita *et al.* (7)	58	M	Ut	IIIB1	Left	-	II	-	-
7	1997	Tomita *et al.* (17)	59	M	Ce	IIIA	transhiatal	-	-	-	-
8	1998	Yano *et al.* (8)	52	M	Mt	IIIB1	Left	retrosternal/eso-gastric	IVb	Ductus arteriosus	-
9	1998	Kinosita *et al.* (9)	61	M	Mt	IIIB1	Left+sternotomy	retrosternal/right colon	III ~ IV	Ductus arteriosus	Aortic arch
10	1999	Matsui *et al.* (10)	65	M	MtLt	IIIB1	Left	-	II ~ III	-	-
11	1999	Saito *et al.* (11)	68	M	UtMt	IIIA1	Left	-	III	-	-
12	1999	Saito *et al.* (11)	60	M	UtMt	IIIA1	Left+sternotomy	-	IVb	-	-
13	1999	Tamura *et al.* (12)	60	M	Ut	IIIB1	Bilateral	retrosternal/eso-gastric	III	-	-
14	1999	Guillem *et al.* (13)	70	M	Ut	IIIB1	Left	-	II	-	-
15	1999	Guillem *et al.* (13)	59	M	Ut	IIIB1	Left	-	II	-	-
16	1999	Guillem *et al.* (13)	30	M	Lt	IIIA1	Left	-	IVb	-	-
17	1999	Yamatsuzi *et al.* (14)	62	M	UtCe	IIIA1	Lt. door open	ant. thoracic/right colon	III	Ductus arteriosus	Aortic arch
18	2001	Sunada *et al.* (15)	67	M	MtLt	IIIB	Left	post. mediastinal/eso-gastric	-	Lt. subclavian a.	-
19	2001	Yamatsuzi *et al.* (14)	71	M	MtUt	IIIA	Lt. door open	intrathoracic/eso-gastric	III	Ductus arteriosus	Aortic arch
20	2002	Kobayashi *et al.* (6)	62	M	Ut	-	Left	-	–	Ductus arteriosus	Aortic arch
21	2002	Tamai *et al.*	46	M	LtAeG	IIIB1	Left	-	–	Ductus arteriosus	-
22	2003	Isiguro *et al.* (16)	58	M	Ut	IIIB1	Left	retrosternal/eso-gastric	0	Ductus arteriosus	-
23	2003	Amano *et al.* (17)	60	M	Mt	IIIB1	Left	retrosternal/eso-gastric	IVa	Ductus arteriosus	-
24	2003	Amano *et al.* (17)	66	M	UtMtCe	IIIB1	Left+sternotomy	retrosternal/eso-gastric	0	Ductus arteriosus	Aortic arch
25	2003	Amano *et al.* (17)	78	F	Mt	IIIB1	Left+sternotomy	retrosternal/eso-gastric	IVa	Ductus arteriosus	Aortic arch
26	2003	Hanazono *et al.* (18)	62	M	Ut	IIIB1	Left	post. mediastinal/eso-gastric	II	Ductus arteriosus	-
27	2004	Simoda *et al.* (19)	64	M	Mt	IIIB1	Left	retrosternal/eso-gastric	IVa	Ductus arteriosus	-
28	2005	Ours	73	M	Lt	IIIB1	Left	retrosternal/eso-gastric	II	Ductus arteriosus	-

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