Primary Malignant Melanoma of the Esophagus

Long-Term Survival After Radical Resection

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Abstract: A 55-year-old man had dysphagia of 4-month duration. Endoscopy revealed a large polypoidal pedunculated blackish mass located in the lower thoracic esophagus. A barium esophagogram revealed a lower esophageal mass. Computed tomography revealed a large polypoidal esophageal mass without any evidence of local invasion or distant disease. Endoscopic biopsy established the diagnosis of melanoma. A radical resection of the esophagus with three-field lymph nodal dissection was undertaken. The patient made an uneventful recovery. Histopathology confirmed the diagnosis of melanoma and positive lymph nodes. Adjuvant chemotherapy was given. He succumbed to systemic recurrence after 69 months.

Key Words: Esophagus, Melanoma, Malignant, Radical, Long-term, Survival, Esophagectomy, Three field, Primary.

Since the first description in 1906, approximately 250 cases of primary esophageal melanoma have been reported in the literature. Nevertheless, malignant melanoma of the esophagus as a primary tumor was accepted only after about 60 years, when De la Pava et al. demonstrated the presence of melanocytes in the esophageal mucosa. The presence of melanocytosis has been implicated as the precursor lesion by some workers. These tumors arising from mucosal melanocytes are known for their aggressive behavior. Despite aggressive treatment, the prognosis remains dismal. The reported 5-year survival is less than 5% and only about one-third of the patients survive beyond 1 year. Long-term survival after radical resection in the present case forms the basis of this report.

CASE REPORT

An Indian male, aged 55 years, presented with a 4-month history of gradually progressive painless dysphagia. He was in good health. General physical examination and systemic examinations were within normal limits. A detailed dermatological, ocular, and mucus membrane examination did not reveal any abnormality. Biochemical, hematologic, and coagulation studies were normal. An upper gastrointestinal endoscopy revealed a large polypoidal pedunculated blackish mass in the lower thoracic esophagus (Figure 1). Gastroesophageal junction and stomach were normal. Endoscopic biopsy clinched the diagnosis of malignant melanoma.

A barium contrast esophagogram revealed a polypoid filling defect with nodular outline involving the lower thoracic esophagus. The mucosal outline was irregular. Proximal esophagus was dilated with hold up of the contrast. A contrast-enhanced computerized tomogram of the chest revealed a well-defined heterogenous soft tissue density, an intraluminal lesion measuring 5 × 6 cm in the mid-thoracic and lower thoracic esophagus. Fat planes with the adjacent viscera were maintained (Figure 1). There was no evidence of distant disease.

A right posterolateral thoracotomy revealed a 9 × 4-cm mass lesion occupying the mid-thoracic and lower thoracic esophagus. The esophagus could be separated easily from the adjacent viscera. The patient underwent esophagectomy and three-field lymphadenectomy in a standard fashion. The gastrointestinal continuity was restored using a gastric conduit through the posterior mediastinal route.

Grossly, the specimen revealed an 8 × 4 × 3-cm, irregular, lobulated, polypoidal, and brownish black mass occupying the mid-esophagus and lower esophagus (Figure 2.4). Microscopically, malignant cells formed confluent masses within the tumor with abundant pigment deposition. Many clusters of cells were also seen infiltrating the overlying epithelium. The tumor cells showed S-100 positivity (Figures 2B–D). Hence, a diagnosis of malignant melanoma, possibly originating from the esophagus, was confirmed. Four of the 42 lymph nodes showed evidence of tumor deposits.

The patient made an uneventful recovery. A postoperative adjuvant chemotherapy of dacarbazine (800 mg/m² on day 1), cisplatin (30 mg/m² on days 2 to 5), and vinblastine (1.6 mg/m² on days 1 to 5) was administered intravenously (IV). The cycles were repeated every 4 weeks, and a total of six cycles were administered. The patient remained disease-free for 66 months as evidenced by periodic imaging and endoscopic examinations. Subsequently, he developed visceral (liver) metastasis and succumbed to recurrent disease after 69 months.

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DISCUSSION

Primary malignant melanoma of the esophagus is a rare disease, which accounts for 0.1 to 0.2% of the esophageal neoplasm. Li et al. in their hospital registry of about three decades reported seven cases of primary esophageal melanoma.

Patients usually present with dysphagia of short duration. The lesion is most often located in the middle and lower thirds of the esophagus, probably because of the greater concentration of melanocytes in this region. An upper gastrointestinal endoscopy reveals a tumor mass in the esophagus. Suzuki et al. detected early stage melanoma in two patients on endoscopic examination. A contrast esophagogram can reveal a large polypoid filling defect with or without the evidence of mucosal destruction. A computerized tomo-

FIGURE 1. Left, Endoscopic examination of the esophagus showing a large polypoidal mass in the esophagus. Note the brownish pigmentation of the mass as compared to the surrounding mucosa. Right, CECT chest at the level of inferior pulmonary vein showing a mass lesion occupying the esophagus. The lumen is chinked. A fat plane with aorta is well preserved. CECT, contrast-enhanced computerized tomogram.

FIGURE 2. Gross polypoidal black mass with narrow pedicle (A), multiple foci of rounded masses of neoplastic cells in the basal layer of squamous epithelium (B), large confluent foci of tumor cells in the deeper portion (C), and S-100 positivity in the tumor cells (D).
gram of the chest and the abdomen can illicit an esophageal mass.9 Endosonogram shows the tumors to be of variable ecogenicity.10 The incidence of the periesophageal lymph nodal metastasis is as high as up to 66%; however, it is unrelated to the depth of tumor invasion.3,4,6

On the basis of standard histologic examination, biopsy specimens are occasionally misdiagnosed as poorly differentiated carcinoma in the absence of melanin granules.11 Preoperative diagnosis can be established in only 54% of the cases.3,4 This mandates the use of special stains such as HMB-45 antibody, S-100 protein, and neuron-specific enolase.3,4,11 Grossly, the lesion may appear polypoidal and is covered with intact mucosa without ulceration. The presence of satellite tumor nodules have been reported in upto 12% of patients.3,4 The lesions are mostly pigmented, but nonpigmented amelanotic variety accounts for in 10 to 25% of the cases.11

Primary malignant melanoma of the esophagus has extremely poor prognosis with a mean survival of 13.4 months from diagnosis.1,3–5 The poor prognosis is due to the aggressive biologic behavior, advanced stage at presentation, and lack of effective therapy. Hematogenous and lymph node metastases are common with 30 to 40% of patients having clinical evidence of metastatic disease at the time of diagnosis.1,3,4,8

Over the years, radical resection for esophageal cancer has gained popularity. Volpin et al.6 in a thorough MEDLINE research of 25 cases of esophageal melanoma, in which patients underwent radical surgery, calculated a 5-year survival of 37%. In a series of seven cases of primary malignant melanoma of the esophagus, median survival of 8 months was achieved with varied therapeutic strategies.5 Nevertheless, one of the patients survived as long as 17 years, which is probably the longest reported survival in this disease.5 Suzuki et al.8 described the detection of early stage melanoma treated by esophagectomy, and they could achieve long-term survival. In all the patients surviving beyond 5 years, surgical extirpation of the tumor had been the main stay of treatment.5,6,8 The present patient also survived beyond 5 years.

A few of the reports mention the use of chemotherapy, radiotherapy, hormone therapy, and immunotherapy in the adjuvant and neoadjuvant setting.12 The role of systemic chemotherapy in adjuvant therapy is not well defined. Naomoto et al.12 reported a reduction of the tumor size to 70% using neoadjuvant DTIC-based chemotherapy. Nevertheless, the patients treated by chemotherapy, immunotherapy, and endocrine therapy are very few to advocate any recommen-

dations. The role of radiotherapy is also not well defined. Because of the lack of conclusive evidence, systemic chemotherapy was given in the present case in view of positive lymph nodes.

Approximately 85% of the patients die with disseminated disease. The most common sites involved at autopsy are the liver (39.3%), mediastinum (34.4%), lungs (24.6%), pleura (19.7%), supraclavicular lymph nodes (19.7%), peritoneum (14.8%), brain (13.1%), kidneys, and adrenals (11.5%), although no site is immune.1,3–6 Likewise, the present case developed systemic metastasis after 66 months.

Thus, primary malignant melanoma is a rare, aggressive tumor of the esophagus, which should be regarded as a potentially systemic disease at diagnosis. Surgical extirpation remains the mainstay of treatment. Although, overall prognosis of the condition is poor, long-term survival can be achieved by radical resection in selected cases. The role of adjuvant and neoadjuvant therapy is not well defined.

REFERENCES