Good Prognosis of Early Stage Malignant Melanoma of the Esophagus: A Case Report

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ABSTRACT

Malignant melanoma of the esophagus is a rare tumor with poor prognosis. The survival of patients is generally less than one year after diagnosis. A case of primary malignant melanoma of the esophagus is presented, who after radical resection of the tumor, is in excellent health, with no evidence of disease 14 months after surgery.

Keywords: Melanoma, Esophagus, Esophageal tumor Govaresh/ Vol. 14, No.3, Autumn 2009; 203-204

INTRODUCTION

Malignant melanoma of the esophagus is a rare tumor and accounts for 0.1-0.2% of all benign and malignant tumors in this organ (1). Only 262 cases have been documented by June 2005 worldwide (2). It often presents as a polypoid mass (3) and occurs most frequently in the middle or lower third of esophagus, especially in middle-aged or elder population (4). Prognosis is poor and does not seem to be related to tumor thickness (2). Mean survival period is 8 (5) to 10 (6) months after diagnosis. Long term survival has been exceptional (1,7). Hereby, we report a case of primary esophageal malignant melanoma who is in excellent condition 16 months after resection of the lesion.

CASE REPORT

A 65 year old male presented with progressive dysphagia since 2 months before and 12 kg

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Department of Pathology, Firoozgar Hospital, Beh-Afarin st, Vali-e-asr sq., Tehran, Iran Tel:+98 21 88942661-4 Fax: +98 21 88942622 E-mail: azaremehrjardi@yahoo.com **Received:**31 May 2009 **Edited:** 28 Jul. 2009 **Accepted:** 29 Jul. 2009 weight loss. There was no history of nausea or vomiting nor any alteration in bowel habits. Endoscopic examination showed 2 masses at 25 cm and 30 cm from incisors. Barium swallow revealed pressure effect in the middle third of esophagus. Ultrasonographic studies of the liver, and chest X-ray were normal. Histopathologic evaluation of biopsy of the esophageal mass was reported as undifferentiated carcinoma. Trans-hiatal total esophagectomy (Orringer procedure) was performed. Esophagus was 13 cm in length, featuring a large polypoid mass in the mid-part, measuring 4.5x2x1.3 cm with a stalk of 0.5 cm in diameter, and another small polyp of 1.5x1.2x0.2 cm, 1.2 cm distal to the first one. Both were ulcerated and showed a pleomorphic tumor composed of polygonal, plump to spindly cells with vesicular nuclei, and large nucleoli (figure 1).

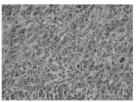


Figure 1: Tumor cells are large, pleomorphic and with distinct nucleoli (x400, H&E).

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Mitoses were frequent, but only occasional cells showed fine brownish cytoplasmic pigments. Adjacent mucosa in the stalk of the large polyp showed lentiginous proliferation of atypical melanocytic cells for a few millimeters (figure 2).

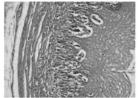


Figure 2: Tumor cells grow in lentiginous pattern in adjacent mucosa (x 400, H&E).

Despite the 13 mm thickness of tumor, it did not invade beyond submucosa. None of the four lymph nodes adjacent to the esophagus was involved by tumor, either. Tumor cells were strongly positive for S-100 protein, and for HMB-45 (figure 3), and were negative for cytokeratin. Sixteen months after surgery, the patient regained his previous weight and was in excellent conditions with no evidence of tumor.

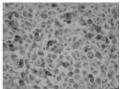


Figure 3: Many tumor cells are positive for HMB-45 (x400, IHC).

DISCUSSION (

The most common site of primary malignant melanoma is the skin. However, it can occur in other sites like in the GI tract and eyes. In the former, it is most common in the anorectal area (8). It is rare in the esophagus, and similar to esophageal carcinoma, dysphagia is the most common symptom. It may easily be missed in a small biopsy of an esophageal mass. Most often, it is not pigmented, and in histology, it is composed of epithelioid, spindled and anaplastic cells. Distinguishing primary from metastatic melanoma in the esophagus may also be difficult, but absence of history of malignant melanoma elsewhere, presence of radial growth phase, and epithelioid and spindle cell histology are in favor of primary esophageal melanoma (9).

In most cases, tumor has already advanced deep in esophageal wall, and lymphatic or distant metastases may be present; so survival of the patients is no more than a few months, despite multimodality treatments (5). The most common sites of metastasis are adjacent lymph nodes, liver, adrenal glands and lung (5). In our case, the tumor did not invade beyond submucosa, and despite its large size, the patient is currently free of tumor.

CONCLUSION

In this regard, it seems that malignant melanoma of the esophagus has a pattern of growth similar to squamous cell carcinoma, and not akin to cutaneous malignant melanoma.

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