Embryonal Rhabdomyosarcoma of the Larynx: a case report

Wen-Pin Chen1 Chun-Liang Tung2 Wei-Hsiung Huang3 Chun-Lin Huang1 Joseph-Hang Leung1

Department of Radiology1, Department of Pathology2, Department of Otolaryngology3, Chia-Yi Christian Hospital

Rhabdomyosarcoma (RMS) represents approximately 4% of all childhood tumor. A third of all RMSs occur in the head and neck region. Laryngeal involvement by RMS is extremely rare. Herein we report a case of a 21-year-old man with the complaint of dysphagia and hoarseness for 2 months. The magnetic resonance imaging (MRI) demonstrated a well-demarcated laryngeal mass with non-specific signal intensities and enhancement. Pathologic result revealed embryonal RMS.

More than 95% of laryngeal tumors in adults are squamous cell carcinoma (SCC). RMS is the most common soft tissue sarcoma and the second most frequent head and neck malignancy in children and adolescents. But it is rarely occurred in the larynx [1-3]. The diagnosis of RMS was established on the basis of the pathologic examinations. Herein we report a case of embryonal RMS involving the larynx and demonstrate the findings of laryngoscope, MRI and pathology.

CASE REPORT

A 21-year-old man was admitted to our hospital with the complaint of dysphagia and hoarseness for 2 months. He also presented with voice change, lumping throat and dyspnea. On physical examination, there were no palpable neck mass or lymphadenopathy. The medical history and laboratory examinations were unremarkable.

The flexible laryngoscope revealed a large, smooth, lobulated mass occupying the larynx and hypopharynx (Fig. 1). Submucosal tumor was impressed. The neck MRI (1.5T, Signa® Horizon LX system; GE) demonstrated a well-demarcated, lobulated mass, measuring 4.0 × 3.7 × 3.2 cm in size, involving the supra-glottic larynx and hypopharynx. The mass appeared isointense on T1-weighted images (T1WI) and hyperintense on T2-weighted images (T2WI) when compared with the muscle. Inhomogenously rim enhancement of the mass was noted after intravenous administration of gadolinium (Fig. 2). These pictures were not typical for laryngeal SCC.

He underwent tracheostomy and laryngotomy and tumor excision. Surgical findings disclosed a large submucosal tumor involving left-sided false vocal cord, aryepiglottic fold, arytenoid cartilage, medial wall of pyriform sinus and post-ericoid area. Pathologic examination revealed grouping proliferation of small round cells with hyperchromatic nuclei and scanty cytoplasm, and focal myxoid stroma. Scattered spindle cells with eosinophilic strape-like...
cytoplasm, tumor necrosis and frequent apoptosis were seen. Immunohistochemical staining showed positive reaction for vimentin, desmin and myogenin (Fig. 3). Embryonal RMS was diagnosed.

**DISCUSSION**

RMS represents approximately 4% of all childhood tumors. RMS is the most common soft tissue sarcoma (60%) and the second most frequent head and neck malignancy next to lymphoma in children and adolescents. A third of all RMSs occur in the head and neck region. It is rarely occurred in the larynx (2%). Three broad anatomic groups have been identified: orbital, parameningeal, and non-parameningeal. There are 3 major categories of RMS: embryonal (58%), alveolar (31%) and pleomorphic. Embryonal and alveolar variants are more commonly occurred in children and adolescents, but pleomorphic variant is usually seen in adult patients [4].

In the literature, there were less than 20 case reports of embryonal RMS of larynx since 1980 [5, 6]. There were only few case reports of laryngeal RMS imaged with MRI. MRI was incapable of distinguishing RMS from other benign and malignant soft tissue tumors [7]. In our case, the tumor revealed nonspecific MR features with hyperintensity on T2WI and inhomogenous contrast enhancement. But well-demarcated and lobulated contour of RMS were not typical for SCC.

More than 95% of laryngeal tumors in adults are SCC. The other tumors with unusual histology account for 2% to 5% of laryngeal tumors. These rare laryngeal tumors include vasoformative tumors, chondrogenic tumors, hematopoietic tumors, salivary gland tumors, fatty tissue tumors, metastases to larynx, neurogenic tumors, myogenic tumors and fibrohistiocytic tumors [8-10]. In some of these rare tumors, computed tomography (CT) and MRI may allow a specific diagnosis. Hemangiomas have very high signal intensity on T2WI and strong enhancement at MRI. Phleboliths, which are pathognomonic

Figure 1. Appearance of gross tumor on flexible laryngoscope. E: epiglottis.

Figure 2. MR images at laryngeal level. Arrow indicates the tumor mass. E: epiglottis; H: hyoid bone; P: pyriform sinus. a. Axial fast spin-echo (FSE) T2WI (3500/100 [TR/TE]) with fat suppression reveals a large lobulated mass with hyperintensity. b. Coronal FSE T1WI (416/10) shows a large lobulated mass isointense relative to muscle. c. Sagittal gadolinium-enhanced FSE T1WI (566/10) with fat suppression demonstrates inhomogenously rim-like enhancement of the mass.
for hemangiomas are easily identified at CT. Chondrogenic tumors typically manifest with coarse or stippled calcifications at CT. At MRI, they have very high signal intensity on T2WI, because of high water content. Small areas of low signal intensity on T1WI and T2WI correspond to stippled calcifications. Lipomas typically show homogeneous nonenhancing lesions with attenuation and signal intensity like subcutaneous fat at both CT and MRI. Paragangliomas and hypervascular metastases such as renal carcinoma typically demonstrate strong enhancement and signal voids at MRI. Metastases from melanotic melanoma usually have high signal intensity on T1WI and low signal intensity on T2WI due to paramagnetic properties of melanin [8]. Although radiologic findings are usually nonspecific in most other non-SCC of the larynx, they should be included in the differentiated diagnosis of laryngeal RMS.

According to the IRS (Intergroup Rhabdomyosarcoma Studies)-IV, the treatment of RMS had changed from extensive surgery toward organ preservation with the multimodality treatment protocols. The role of surgery is limited to biopsy or palliation of airway obstruction. Embryonal RMS of larynx appears to be highly responsive to chemoradiation and has a good prognosis [9-11].

In conclusion, RMS is a rare laryngeal malignancy and larynx is a rare location of head and neck RMS. It presented nonspecific MRI appearance, and only biopsy can confirm the diagnosis. When we meet an unusual laryngeal mass in a young adult, the possibility of RMS should be considered.

**REFERENCE**


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**Figure 3.**

a. Low power photomicrograph showed the submucosal tumor of larynx (hematoxylin and eosin stain, x40).

b. Histologically, the tumor was composed of hypercellular primitive round neoplastic cells and occasional elongated rhabdomyoblasts with eosinophilic mofibrils (hematoxylin and eosin stain, x400).

c. Those neoplastic cells revealed positive staining of myogenin by immunohistochemical stain.
喉部胚胎型横纹肌肉瘤：病例报告

陳文彬 1   董俊良 2   黃咸雄 3   黃駿麟 1   梁恆 1

嘉義基督教醫院 影像醫學科 1   病理科 2   耳鼻喉科 3

横纹肌肉瘤约占小孩子肿瘤百分之四的比例，三分之一病例发生於頭頸部区域，發生於喉部則極為少見。我們報告一位 21 歲男性，主訴吞嚥困難及聲音沙啞有 2 個月之久。磁振造影影像顯示一個界限清楚之喉部腫塊，沒有特異性之訊號及顯影。病理結果証實是胚胎型横纹肌肉瘤。