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Case Report

Spindle cell rhabdomyosarcoma in the hypopharynx of an adult

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Abstract: We herein present the first-reported case in the English-language literature of a spindle cell rhabdomyosarcoma in the hypopharynx of an adult. A 58-year-old male presented with a progressive, 7-month history of discomfort during swallowing, accompanied by hoarseness for 1 month. Computed tomography and magnetic resonance imaging revealed a tumor in the left pyriform sinus; frozen section evaluation suggested that it was rhabdomyosarcoma. The tumor was removed completely via left lateral cervical incision. Section margins were negative, and laryngeal function was preserved. The postoperative pathological results indicated spindle cell rhabdomyosarcoma. The patient received postoperative radiotherapy (6,000 cGy in 200 cGy fractions, delivered over 30 days). However, he was also required to undergo urgent tracheostomy due to severe laryngeal obstruction, precipitated by laryngeal edema, 3 months subsequent to radiotherapy. The patient was free of disease 25 months postoperatively but is still unable to close his tracheostomy.

Keywords: Spindle cell rhabdomyosarcoma, hypopharynx, adult, radiotherapy

Introduction

Rhabdomyosarcoma (RMS), while being particularly rare in adults, represents the most common soft tissue sarcoma in children [1]. Although RMS most frequently arises in the head and neck, mainly in orbit and parameningeal sites, non-orbit, non-parameningeal cases are rare [2]. RMS of the hypopharynx in adults is extremely uncommon, and is only ever reported either as a part of large studies [3, 4] or in separate case reports [5]. Accordingly, scant information is available to guide clinicians in the treatment and diagnosis of hypopharyngeal RMS.

Three general types of pathologic RMS have been identified: embryonal, alveolar, and pleomorphic [6]. Spindle cell RMS is a variant of embryonal RMS, which was first described in 1992 [7]. Compared with the other subtypes, the spindle cell variant has a favorable outcome in children; however, in the adult population, prognosis is poor [6, 7].

It is unclear whether hypopharyngeal RMS behaves similarly to histologically different tumors that occur at other sites. In the present study, an adult case of evolved hypopharyngeal spindle cell RMS was assessed, and the characteristics, diagnosis and treatment of such tumors were discussed.

Case report

A 58-year-old male presented with a progressive 7-month history of discomfort during swallowing, accompanied by hoarseness for 1 month. He had no history of sore throat, cough, difficulty in breathing, or fever. He had a 30-year history of smoking (20 cigarettes per day) but no history of drinking alcohol. He consulted a local clinic, who administered drug therapy for "chronic pharyngitis" in April 2012. He then consulted our clinic. Computed tomography (CT) revealed a mass in the left pyriform sinus (**Figure 1**); he was admitted to our department on May 4, 2012. Indirect laryngoscopy and stroboscopic laryngoscopy revealed an irregular swelling in the left aryepiglottic fold and left pyriform sinus, as well as attenuation in the motion of the left vocal cord. An irregular, firm 3×4-cm mass in the posterosuperior area of the left thyroid cartilage was palpated; the

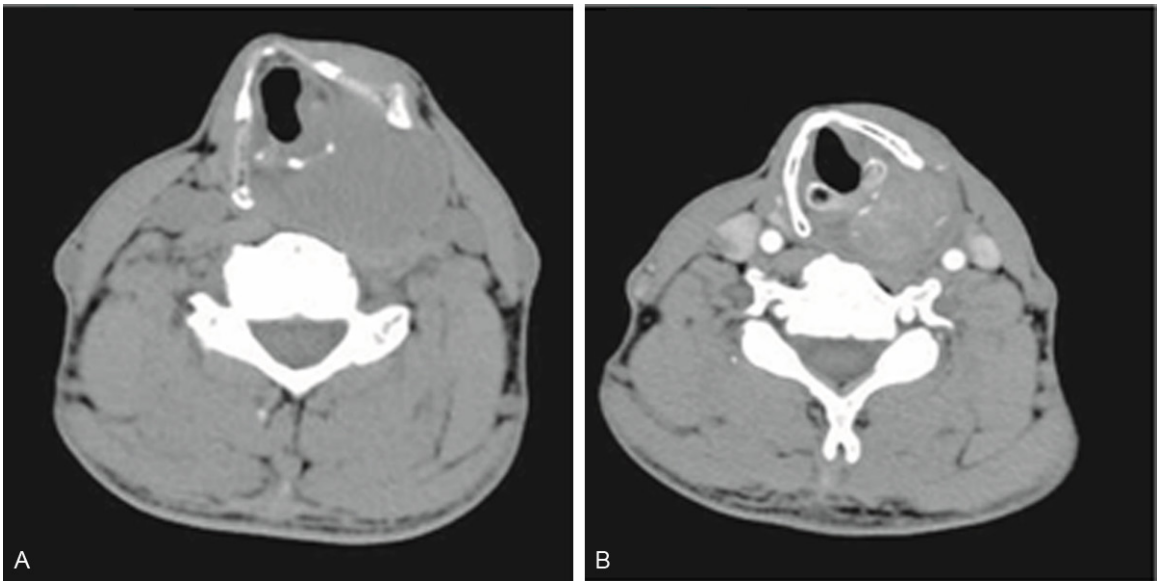

Figure 1. CT revealed a mass in the left pyriform sinus. A. A plain scan showed that the mass was homogenous; the CT value was set at 50 Hu. B. Contrast-enhanced imaging showed that the lesion had heterogeneous mild enhancement.

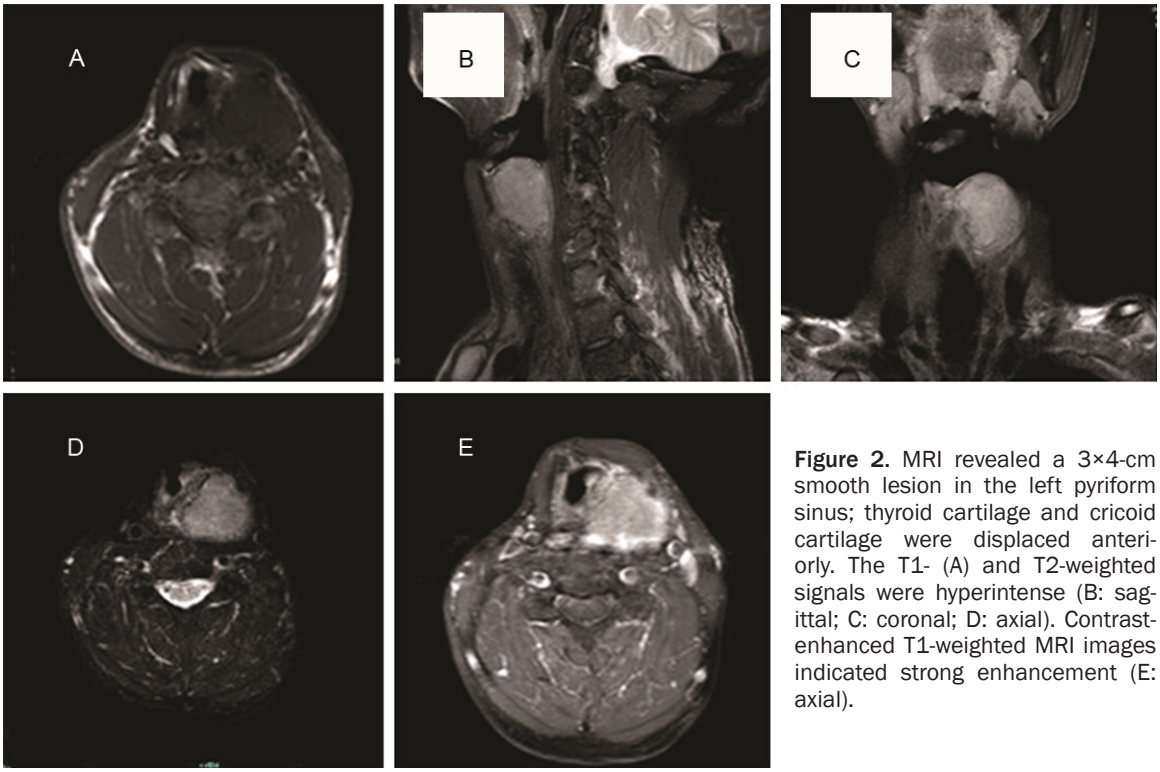

Figure 2. MRI revealed a 3×4-cm smooth lesion in the left pyriform sinus; thyroid cartilage and cricoid cartilage were displaced anteriorly. The T1- (A) and T2-weighted signals were hyperintense (B: sagittal; C: coronal; D: axial). Contrast-enhanced T1-weighted MRI images indicated strong enhancement (E: axial).

regional lymph node was not palpated. Magnetic resonance imaging (MRI) revealed a smooth 3×4-cm lesion in the left pyriform sinus; the thyroid cartilage and cricoid cartilage were displaced anteriorly. T1- and T2-weighted signals were hyperintense. Contrast-enhanced T1-weighted MRI images demonstrated pronounced enhancement (**Figure 2**).

On May 11, 2012 the biopsy was performed via suspension laryngoscopy under general anesthesia. The frozen section evaluation suggest-

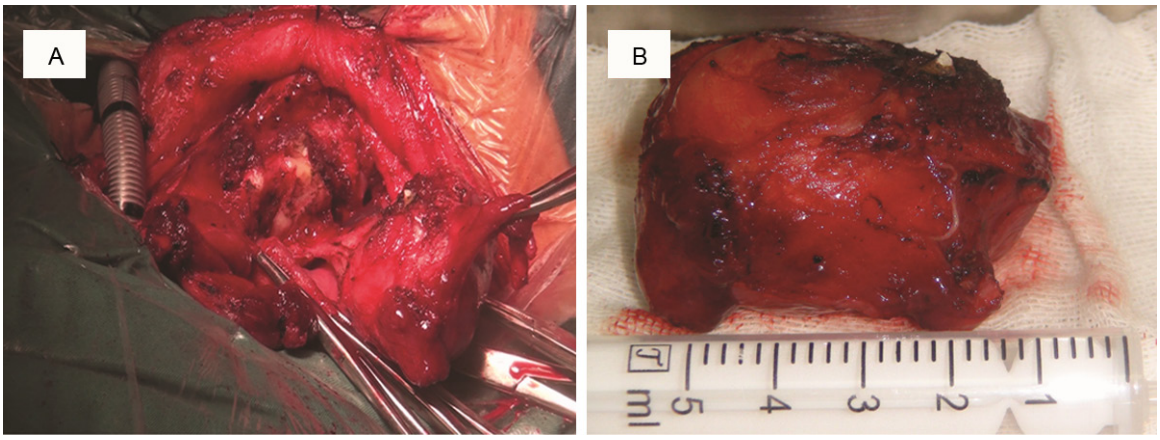

Figure 3. The tumor was removed completely (including 2 cm of mucosa external to the tumor) via left lateral cervical incision (A). During the operation, we discovered that the 4.5×5.5-cm tumor was located in the inner lateral area of the left pyriform sinus. The mass was pink, smooth and encapsulated (B).

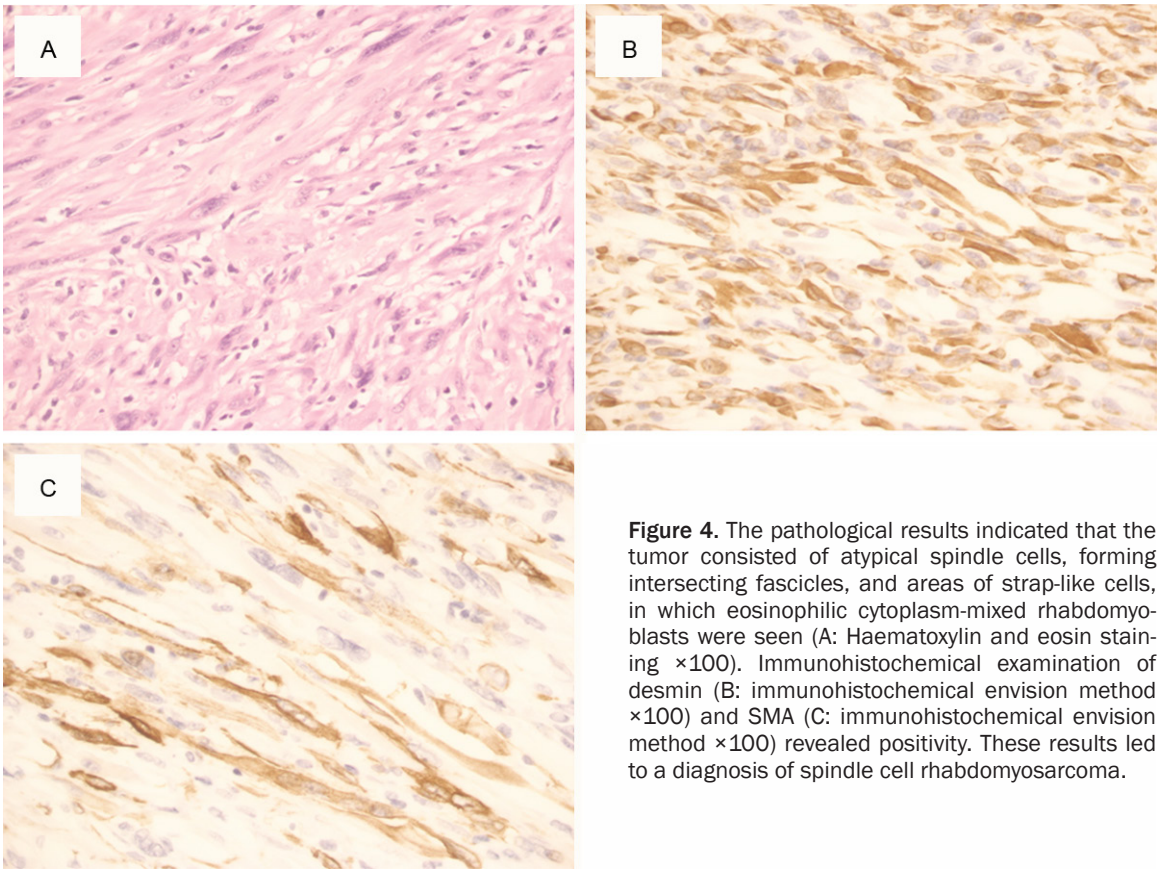

Figure 4. The pathological results indicated that the tumor consisted of atypical spindle cells, forming intersecting fascicles, and areas of strap-like cells, in which eosinophilic cytoplasm-mixed rhabdomyoblasts were seen (A: Haematoxylin and eosin staining ×100). Immunohistochemical examination of desmin (B: immunohistochemical envision method ×100) and SMA (C: immunohistochemical envision method ×100) revealed positivity. These results led to a diagnosis of spindle cell rhabdomyosarcoma.

ed that the tumor was a soft tissue sarcoma; rhabdomyosarcoma was suspected initially. The tumor was removed completely (including 2 cm of mucosa external to the tumor) via a left lateral cervical incision (**Figure 3A**). During the operation, we discovered that the 4.5 cm × 5.5 cm tumor was located in the inner lateral area of the left pyriform sinus. The mass was pink, smooth and encapsulated (**Figure 3B**). Section margins were negative, and laryngeal function was preserved. Neck dissection was not performed, because no regional lymph node metastases were revealed by either CT or MRI. The patient was classified as Group I according

to the Intergroup Rhabdomyosarcoma Study Committee (IRS) [8].

Postoperative pathological results revealed that the tumor consisted of atypical spindle cells, forming intersecting fascicles, and areas of strap-like cells with eosinophilic cytoplasm-mixed rhabdomyoblasts. Immunohistochemical examination revealed both desmin and α-smooth muscle actin (SMA). CD117, S-100, CD34, cytokeratin (AE1/AE3), MyoD1 and caldesmon were not present. These results led to a diagnosis of spindle cell rhabdomyosarcoma. The resection margins were tumor-free (**Figure 4**).

The postoperative period was uneventful. The patient was discharged 18 days post-surgery with closure of his previous tracheostomy. His swallowing function was normal. Postoperatively, the patient received radiotherapy (6,000 cGy in 200-cGy fractions delivered over 30 days). However, he subsequently underwent an urgent tracheostomy procedure, due to a severe laryngeal obstruction caused by laryngeal edema 3 months after radiotherapy. The patient has been closely followed up with CT scans and laryngoscopy at 3-month intervals and was free of disease 25 months postoperatively. However, he also remains unable to close his tracheostomy.

Discussion

Rhabdomyosarcomas are extremely rare in adults, especially in the hypopharynx. To the best of our knowledge, fewer than 10 cases of pharyngeal involvement have been reported in the English-language literature [3-5, 9]. In 1956, Gammell et al. provided the first report of hypopharyngeal RMS. In 2009, another hypopharyngeal RMS case was reported by Hasan et al. [9]. Owing to the rarity of hypopharyngeal RMS, little is known about the behavior of these aggressive tumors. Chemotherapy and radiotherapy have been recommended as the standard therapeutic approaches by the IRS Committee and, in many cases, result in relatively favorable prognoses [1-5].

Spindle cell RMS is a neo-variant of embryonal RMS [6, 10]. Histopathologically, the tumor consists of long fascicles of elongated spindle cells containing eosinophilic cytoplasm, in addition to admixed rhabdomyoblasts [6, 10, 11]. These characteristics and immunochemical markers may differentiate it from other spindle cell malignant tumors: leiomyosarcoma, spindle cell carcinoma, desmoplastic melanoma, and fibrosarcoma [6, 10, 11]. Spindle cell RMS consistently tests positive for desmin, SMA, and Myf-4 and negative for S100, h-caldesmon, CD34, and glial fibrillary acidic protein [6, 10, 11]. In the present case, the tumor consisted of atypical spindle cells, forming intersecting fascicles, and areas of strap-like cells where eosinophilic cytoplasm admixed rhabdomyoblasts were seen. Immunohistochemical examination confirmed the presence of Desmin and α-smooth muscle actin (SMA). CD117, S-100, CD34, cytokeratin (AE1/AE3), MyoD1 and caldesmon were not detected.

Spindle cell RMS always involves young males and has a more favorable outcome compared with the other forms of RMS [6, 10]. The IRS reported that the 5-year survival of young patients with spindle cell RMS in the paratesticular area was 95.5%, compared with 80% in patients with non-spindle cell RMS [11]. However, some reports have indicated adverse results in adults with spindle cell RMS.

In 1998, Rubin et al. provided the first description of spindle cell RMS affecting adults in two cases, one arising in the cheek and the other in the left hemidiaphragm. Although both patients were treated with surgical resection and chemotherapy, they died shortly after diagnosis (13 and 27 months) [12]. In 2005, Nascimento et al. reported 16 adult cases of spindle cell RMS. Two patients died from the disease 13 and 7 months after diagnosis; 25% of patients developed metastases, and 40% experienced uncontrolled local disease [10]. In 2006, Mentzel et al. documented seven adult cases of spindle cell RMS; two patients, both with lung metastases, died from the disease 12 and 24 months following diagnosis [13]. To our knowledge, there has been no report of spindle cell RMS in the hypopharynx in adults.

Treatment of spindle cell RMS in adults is the same as it is for other types of RMS and, thus, includes a combination of surgery, chemotherapy, and radiotherapy [6, 10, 12, 13]. In the present case, the patient was categorized as Group I and received postoperative radiotherapy. The patient was alive and disease-free 25 months after surgery.

In summary, spindle cell RMS in adults is rare. This represents the first report on spindle cell RMS in the hypopharynx of an adult.

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Disclosure of conflict of interest

None.

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