Clinical Case Report

Medicine®

OPEN

Sonography used in the infantile desmoid fibromatosis of postcricoid area

A case report

Chunxia Xia, MD[a], Qiang Zhu, MD, PhD[a,*], Changli Yue, MD[b], Minxia Hu, MD[a], Pingdong Li, MD[c], Zheng Li, MD[d]

Abstract

Rationale: Infantile desmoid fibromatosis of the postcricoid area is a rare disease and is characterized by a proliferation of fibrous tissue with non-metastasis, local infiltration, and a high rate of recurrence after surgical resection. Currently, ultrasound is scarcely used in the hypopharynx and larynx area.

Patient concerns: A 4-year-old boy presented with hoarseness, deep voice and snoring for 2~4 years without any surgical history. On sonography, the lesion was found in the postcricoid area, and the left larynx showed impaired mobility in real time observation. Complete excision with a negative margin in this pivotal anatomic area is impossible, and necessitates a long-time surveillance.

Diagnoses: Infantile desmoid fibromatosis of the postcricoid area was diagnosed according to surgery and histopathology.

Interventions: Local excision was carried out to relieve the upper airway narrowing.

Outcomes: Relieved hoarseness and snoring were reported on the latest follow-up. A residual lesion was seen in the surgical bed and maintained a stable extent on ultrasound and MR imaging after a year.

Lessons: Considering the non-radiation merit and diagnostic ability, ultrasonography is advocated as a valuable supplementary imaging method to CT, MR and laryngoscopy in the juvenile larynx and hypopharynx.

Abbreviations: CT = computed tomography, MR = magnetic resonance.

Keywords: desmoid fibromatosis, hypopharynx, infantile, sonography, tumors

1. Introduction

Infantile desmoid fibromatosis is a rather rare disease and is considered to be virtually indistinguishable from the adult form of fibromatosis (desmoid tumor). This disease is characterized by a proliferation of fibrous tissue with non-metastasis, local infiltration, and a high rate of recurrence after surgical resection.[1] Lesions of infantile desmoid fibromatosis most frequently involve the head and neck and the preferred sites include tongue, mandible, and mastoid process, whereas hypopharynx and larynx are scarcely reported.[1,2] Computed tomography (CT) and magnetic resonance (MR) imaging are commonly the primary imaging examinations performed in patients with suspected pharyngeal and laryngeal lesions. To the best of our knowledge, sonography has not been used for infantile fibromatosis of the pharynx and larynx in the literature up to now. In this case report, we describe a case of infantile desmoid fibromatosis of the postcricoid area and discuss the role of sonography compared with CT and MR imaging.

2. Case report

A 4-year-old boy was referred to our hospital, presented with hoarseness and deep voice without any surgical history. He had been complaining of snoring for last 2 years, which got worse when lying flat, but there was no buccal breathing or stuffing occurring in sleep. On physical examination, an enlarged adenoid was observed occupying about two-thirds of the posterior naris. On laryngoscopy, a submucosal swelling was seen behind the left aryepiglottic fold with an intact larynx mucosa noted. Overall visualization of glottis could not be achieved due to the associated displacement by the swelling.

On initial sonograms, an oval mass with a measurement of 4.5 × 2.1 × 1.4 cm (maximum length × transverse diameter × antero-posterior diameter) was found mainly in the middle and left aspects of the postcricoid area, extending from the level of arytenoids downward to the inferior margin of cricoid. It had a well-circumscribed margin and homogeneous echogenicity resembling that of cervical and posterior cricoarytenoid muscles, and the mass was surrounded by hyperechoic structures consistent with fatty or fibrous tissue. The left arytenoid and vocal cord were displaced

Editor: Jimmy T. Efird.

The authors declare no conflicts of interest.

[a] Department of Diagnostic Ultrasound, Beijing Tongren Hospital, Capital Medical University, [b] Department of Pathology, Beijing Tongren Hospital, Capital Medical University, [c] Department of Otorhinolaryngology Head and Neck Surgery, Beijing Tongren Hospital, Capital Medical University, [d] Department of Diagnostic Ultrasound, Beijing Aerospace General Hospital, Beijing, China.

[*] Correspondence: Qiang Zhu, Department of Diagnostic Ultrasound, Beijing Tongren Hospital, Capital Medical University, Beijing 100730, China (e-mail: qzhu_mail@126.com).

Medicine (2017) 96:44(e8500)

Received: 9 February 2017 / Received in final form: 2 October 2017 / Accepted: 7 October 2017

http://dx.doi.org/10.1097/MD.0000000000008500

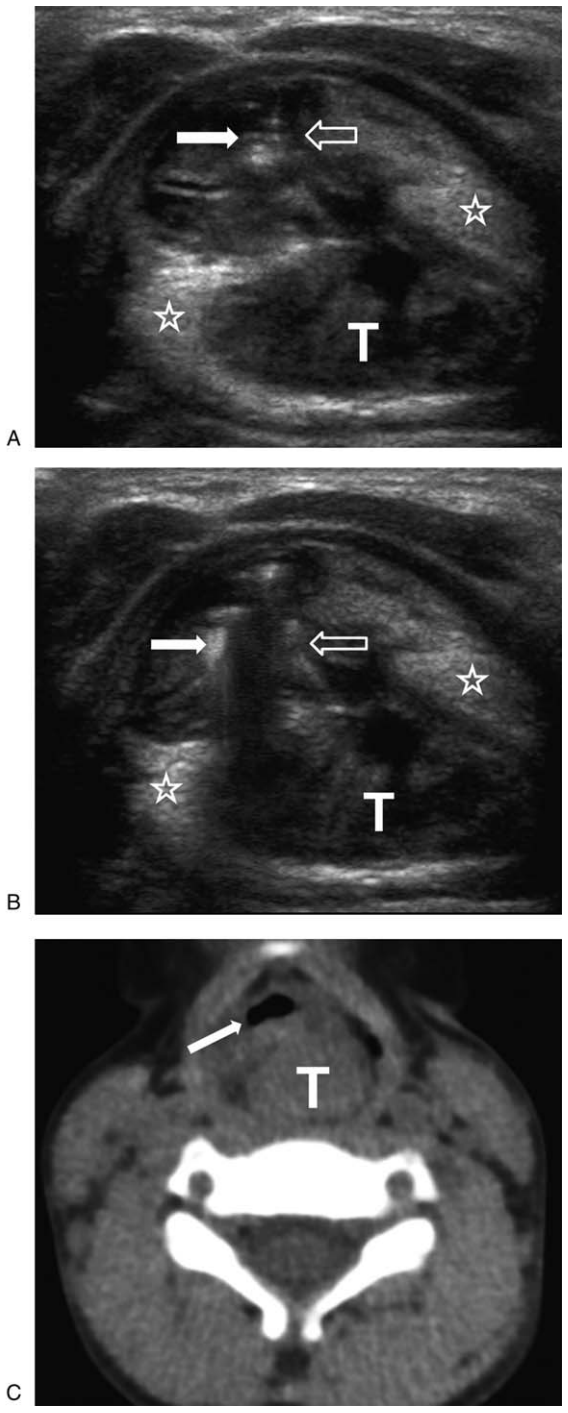

Figure 1. Infantile desmoid fibrosis of postcricoid area in a 4-y-old boy before surgery. (A, B) Sonographic appearance. A hypoechoic tumor (T) is located in the postcricoid area with hyperechoic fatty-fibrous structures (star) surrounded. (A) During breath holding, the glottis is closed with the right vocal cord (solid arrow) adducted, whereas the left one presents a rigid status in the midline (open arrow). (B) During breath relaxation, the right vocal cord adducts normally (solid arrow), whereas the left vocal cord (open arrow) almost keeps fixed in the adductive state probably owing to the compression to the left arytenoid by the tumor. (C) On axial view of unenhanced CT, an oval lesion (T) is localized posterior and lateral to the left aryepiglottic folds. It appears as well-defined, homogeneous with moderate attenuation resembling that of muscle in the neck. The larynx is displaced anteriorly with the presence of luminary narrowing (solid arrow).

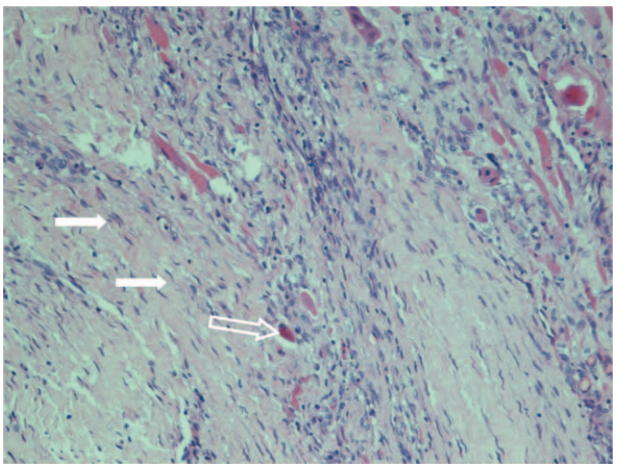

Figure 2. Photomicrograph of the resected tumor. Proliferation of fibroblasts (solid arrows) are surrounded and separated from one another by collagen. Remnants of striated muscle fibers (open arrow) are seen entrapped in the tissue. Hematoxylin-eosin ×200.

slightly forward and upward with decreased mobility during breathing and phonation shown on the real-time imaging (Fig. 1A and B). On color Doppler, some dotlike flow was detected peripherally. The contralateral structures and the laryngeal mobility were grossly unremarkable. On axial view of unenhanced CT, an oval lesion was localized posterior and lateral to the left aryepiglottic folds with extension to the proximal segment of esophagus. It appeared as well defined, homogeneous with moderate attenuation resembling that of muscle in the neck, measuring 57 HU. The larynx was displaced anteriorly with the presence of luminary narrowing (Fig. 1C). Contrast agent was not administered considering the patient's young age.

An endoscopic biopsy was performed and the subsequent pathologic finding indicated that it was a mass originating from fibrous tissue without any evidence of malignancy. Twenty-two days after the endoscopic biopsy, a tumor biopsy and local excision surgery was carried out from an access of the left lateral neck. During the operation, a tough submuscosal mass with a pseudo-capsule was detected in the postcricoid area continuing to the upper esophagus. Frozen section examination was performed and the non-malignant nature was confirmed. To relieve the upper airway narrowing, the main part of the tumor was resected from the postcricoid area. Histopathologically, proliferating fibroblasts were found in bundles invading the striated muscle tissue (Fig. 2). The immunohistochemical stains showed positive for smooth muscular actin (SMA) and Vimentin, but Desmin, MyD1, CD117, and S-100 protein were negative. Some mitotic activity was found with 1 to 2 counts per high-power field. On the 6-month follow-up unenhanced CT, a residual lesion was seen in the surgical bed and involving upper esophagus, whereas the narrowing of the upper airway was relieved compared with the preoperative CT (Fig. 3A). The lesion maintained a stable extent on ultrasound and MR imaging after another 6 months (Fig. 3B and C). Given the juvenile growth of the patient and the benign or intermediate nature of this disease, a wait-and-see strategy was applied and the patient is still under review. Symptoms newly complained were not found and relieved hoarseness and snoring were reported on the latest follow-up on call.

This case report was approved under the review of the Ethics Committee of Beijing Tongren Hospital, affiliated to Capital Medical University.

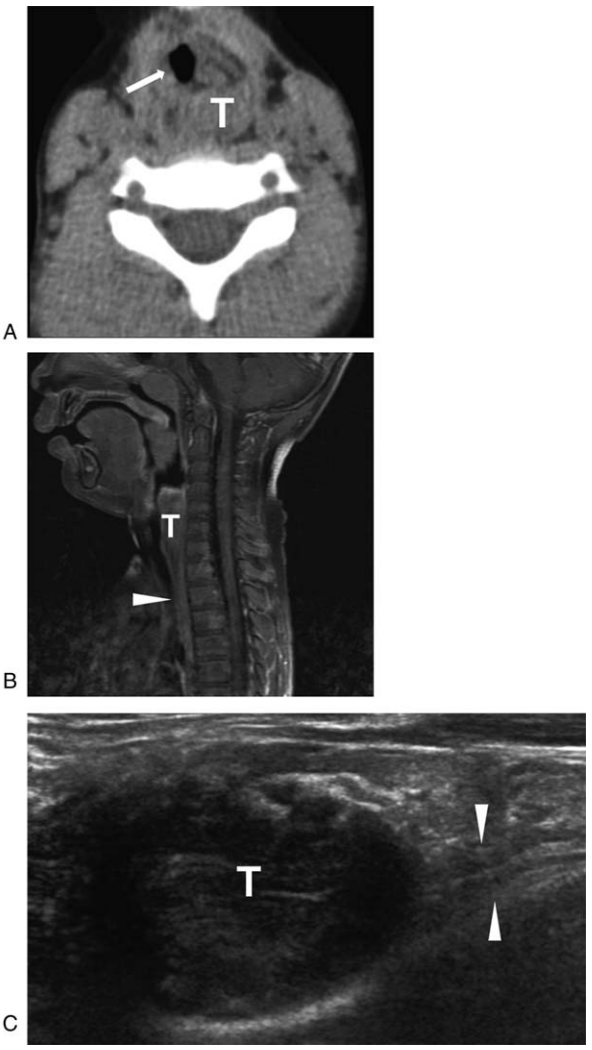

Figure 3. Postoperative findings on surveillance imaging. (A) An uneradicated lesion (T) is seen in the surgical bed on 6-mo follow-up by unenhanced CT, while the narrowing of the upper airway is relieved compared with the preoperative CT (Fig. 1C). (B) The lesion (T) maintained a stable extent with slight enhancement on the contrast T1-weighed MR image after another 6 mo. (C) On sonography for 1-y follow-up, hypoechoic tumor (T) adjacent to the upper esophagus (arrowheads) is noted on longitudinal view.

3. Discussion

In this case report, we find ultrasound could visualize the juvenile hypopharynx and larynx with a high resolution image and could supply valuable information on the diagnosis of desmoid fibromatosis of the postcricoid area. The non-radiation, real-time nature, and diagnosis capability for this disease suggest that ultrasound could be used as a repeatable imaging method in the long-term follow-up for the pediatric patients after surgery.

Fibromatosis is a rare disease and the estimated incidence is 2 to 4/1,000,000/y in the general population.[3] It is defined as an intermediate tumor that rarely occurs in the head and neck of children.[4] Lesions in the head and neck are reported in 12% of all cases, and 71% occur in the first decade of life.[2,5] There has been challenge to the management of fibromatosis in these pivotal anatomic areas because complete excision with a negative margin is usually impossible, and thus frequent local recurrence is common in many cases. In this case, the diffused extension of the lesion in the postcricoid area downward to the upper esophagus limited the successful surgery and the residual lesions were found in the postoperative imaging. Radio- or chemotherapy, reported by some authors,[6,7] was not applied in view of the controversial effects and the potential hazard to the growth of a young patient.

Considering the biological behaviors of the infantile fibromatosis, imaging plays an important role throughout the whole clinical course including the preoperative evaluation and the post-therapy follow-up. CT and MR imaging are usually the modalities of choice, although these 2 imaging methods have some disadvantages especially in children. Radiation dose intake is the major concern for CT and limits its application in the long-term follow-up in this case. For another, sedations should be applied during the performance of MR. Even under this condition, the motion artifacts caused by respiration is inevitable and degrade the imaging quality as limited characterization of the lesions on CT and MR in some cases.

Generally speaking, sonography is scarcely used in the investigation of pharynx and larynx, because calcification of thyroid cartilage and air within the laryngeal cavities are usually regarded as the most important unfavorable factors to the ultrasonography examination. Fortunately, rare thyroid cartilage calcifications occur in the juvenile individuals and thus the juvenile larynx is predominantly sonolucent.[8,9] Noticeably, in this case, the patient underwent a preceding ultrasound examination followed by CT evaluation before the operation. The thyroid cartilage and neck soft tissue were used as acoustic windows to image the larynx and pharynx with the high-frequency ultrasound. The lesion of the postcricoid area was delineated on high resolution sonography showing the muscle-like echogenicity, and the surrounding hyperechoic structures suggested of fibrous tissue. Consideration of the clinical history and the sonographic features combined threw light on the impression of the lesion being a neoplasm in benign etiology originating from the muscular and/or fibrous tissue. In addition, assessment of vocal fold mobility with real-time ultrasound is of great benefit because few young children will tolerate laryngoscopy. Ultrasound showed the impaired mobility of the left hemilarynx during the maneuvers of respiration and phonation before surgery and it was relieved to some extent after the local excision surgery. We therefore supposed that ultrasound could be used as the first-line method for the initial diagnosis and long-term follow-up in children with larynx and pharynx diseases.

High-frequency ultrasonography could be used to examine the diseases in the juvenile larynx and hypopharynx, and prove to be a valuable supplementary imaging method to CT, MR, and laryngoscopy in the field. Taking advantage of the non-radiation nature, sonography is advocated as the first-line diagnostic imaging modality in the larynx and hypopharynx area in children.

References

[1] Goldblum JR, Folpe AL, Weiss SW. Goldblum JR, Folpe AL, Weiss SW. Fibrous tumors of infancy and childhood. Enzinger and Weiss's Soft Tissue Tumors 6th ed.Saunders Elsevier, Philadelphia, PA:2013;272–6.
[2] Kruse AL, Luebbers HT, Grätz KW, et al. Aggressive fibromatosis of the head and neck: a new classification based on a literature review over 40 years. Oral Maxillofac Surg 2010;14:227–32.
[3] Reitamo JJ, Häyry P, Nykyri E, et al. The desmoid tumor. I. Incidence, sex-, age- and anatomical distribution in the Finnish population. Am J Clin Pathol 1982;77:665–73.
[4] Miyashita H, Asoda S, Soma T, et al. Desmoid-type fibromatosis of the head and neck in children: a case report and review of the literature. J Med Case Rep 2016;10:173.

[5] Tostevin PM, Wyatt M, Hosni A. Six cases of fibromatosis of the head and neck in children. Int J Pediatr Otorhinolaryngol 2000;53: 235–44.
[6] Hourani R, Taslakian B, Shabb NS, et al. Fibroblastic and myofibroblastic tumors of the head and neck: comprehensive imaging-based review with pathologic correlation. Eur J Radiol 2015;84:250–60.
[7] Wang W, Koirala U, Ma S, et al. Age-based treatment of aggressive fibromatosis in the head and neck region. J Oral Maxillofac Surg 2014;72:311–21.
[8] Loveday EJ. Ultrasound of the larynx. Imaging 2003;15:109–14.
[9] Ongkasuwan J, Devore D, Hollas S, et al. Laryngeal ultrasound and pediatric vocal fold nodules. Laryngoscope 2017;127:676–8.