CHONDROSARCOMA OF THE LARYNX IN A 36-YEAR OLD WOMAN: A CASE REPORT

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ABSTRACT

Chondrosarcomas are malignant tumors of cartilaginous tissue that mostly located in the pelvis and upper extremities. Chondrosarcomas of larynx are very rare, slow-growing tumors that causes destruction of the surroundings tissues by the mass effect. They accounts for 0,07 to 2% of all laryngeal tumors and are the most common nonepithelial neoplasm of the larynx. We present a clinical case with histopathologically confirmed laryngeal chondrosarcoma in young 36-years-old female. Asthma was diagnosed at first and she was treated for six months, because of the history of hoarseness and dyspnoea. Endoscopy examination showed a smooth tumor located in subglottis region near left arytenoid cartilage, and paralysis of left vocal cord. CT scan confirmed a prominence in the left arytenoids cartilage. The surgical procedure revealed that the tumor arose from cricoid cartilage and filled in almost all lumen of the larynx. At follow-up tree months later endoscopy revealed a lesion like a granulation at the same site where the original lesion had arisen. CT scan confirm irregular swelling of the tissue within calcification in the subglottis region on the left side. She is following up regularly and the time of the writing has been asymptomatic for one and half year. The etiology of chondrosarcomas is unknown and several theories have been proposed. Because the tumor grows slowly, patient may experience a subtle symptoms for several years before a diagnosis is made. Physical examination of a patient with chondrosarcoma mostly revealed the subglottic or submucosal mass, vocal fold paralysis or displacement of arytenoids cartilage by the tumor. The diagnosis and management of these tumors are challenge, because of their rare occurrence and slowly growth.

Key words: Chondrosarcoma, laryngeal neoplasm

INTRODUCTION

Chondrosarcomas are malignant tumors of cartilaginous tissue that are usually located in the pelvis and upper extremities. Chondrosarcomas of larynx are very rare, slow-growing tumors that causes destruction of the surroundings tissues by the mass effect. They
account for 0.07 to 2% of all laryngeal tumors and are the most common nonepithelial neoplasm of the larynx. Actually, real incidences are not clear, as low grade chondrosarcomas are often misinterpreted as chondromas. The aetiology is unknown, but initial, disordered ossification of the laryngeal cartilage has been suspected to be responsible. Correct diagnosis of laryngeal cartilaginous tumors requires a complete examination of the entirely resected tumor. According to the Figi even the most experienced otolaryngologist will encounter not more than one cartilaginous tumor of the larynx during his work. We present a clinical case with histopathologically confirmed laryngeal chondrosarcoma in young female.

CASE REPORT

A 36-years-old woman with 6 months history of hoarseness and dyspnoea. Asthma was diagnosed at first and she was treated for six months. Laryngoscopy revealed an infiltration in region of posterior commissure and immobile of left vocal cord. Endoscopy examination showed a smooth tumor located in subglottis region near left arytenoid cartilage and immobile of left vocal cord (Figure 1). A computed tomography (CT) scan of the neck revealed a tumor located in the subglottis region on the left side. The mass of the tumor demonstrated calcification within the tumor and its capsule. The tumor caused the osteolysis of cricoid cartilage on the left side and it also infiltrated posterior commissure (Figure 2 and Figure 3). CT scan confirmed a prominence in the left arytenoids cartilage. Enlarged of lymph nodules alongside the artery of the neck was diagnosed. Their dimension were about 18 mm.

Figure 1. Endoscopy examination of the larynx before operation. Tumor located in subglottic region on the left and posterior part of the larynx.
Chondrosarcoma of the larynx

Figure 2. CT scan of the tumor in larynx before operation.

Figure 3. CT scan of the neck before operation. Tumor localized in the left part of larynx, narrows the breathing space.
Chondrosarcoma of the larynx


Figure 4. Pictures of laryngoscopy examination, 7 days after surgical treatment. On the site of the resection is swelling of the tissue, left vocal cord is motionless.

We undertook a surgical approach by open laryngofissure. The surgical procedure revealed that the tumor arose from cricoid cartilage and filled in almost all lumen of the larynx. The tumor had extremely hard consistency. We resected the tumor with a margin of normal tissue. In anticipation of postoperative oedema a tracheotomy was performed. Histopathology revealed a low-grade chondrosarcoma. The patient’s postoperative course was uncomplicated. She didn’t have trouble with swallow. Seven days after the operation the endoscopy examination showed a swelling of the tissue in the site of the resection and immobile of left vocal cord (Figure 4). Control CT revealed a postoperative scarf in the site of the resection and restriction of true glottis.

At follow-up tree months later endoscopy revealed a lesion like a granulation at the same site where the original lesion had arisen. CT scan confirm irregular swelling of the tissue within calcification in the subglottis region on the left side. To assess the radical resection we decided to make PET-CT examination. PET-CT scan of the larynx revealed no
increase metabolism of glucose in the site of the resection. It showed swelling of the left aryepiglottica fold and slightly increase of glucose metabolism in this region (Figure 5). Patient did not agree for second operation. She is following up regularly and the time of the writing has been asymptomatic for one and half year.

Figure 5. PET – CT scan 3 months after operation.

DISCUSSION

Chondrosarcoma of the head and neck region constitute about 10 % of all chondrosarcomas tumors of the body 4. The first case of the cartilaginous tumor of the larynx was reported in 1822 by Heusinger 5. The first case of laryngeal chondroma was presented in 1816 by Travers to the Medico-Chirurgical Society in London 6, and the first case of a laryngeal chondrosarcoma was reported by New in 1935 7.
The etiology of chondrosarcomas is unknown and several theories have been proposed. It has been suggested that laryngeal chondrosarcomas may originate from an abnormal ossification on laryngeal cartilages starting in the third decade. Areas of ossification in hyaline cartilages show a predilection for the development of laryngeal chondrosarcomas. Another theory is that chondrosarcomas are caused by malignant changes secondary to polytetrafluoroethylene (Teflon) injections or exposure to radiation. Our patient is 36 years old female which is unusual since the literature indicates a clear male predominance mostly between 50-80 years. The age of onset of CS is reported in the literature to range from 33 to 91 years-old, with an average of about 60 years-old. The tumor presents of lobules of hypercellular atypical chondrocytes in a basophilic cartilaginous matrix material. Lobules of neoplastic cartilage invade bone within the cartilage. The cells have a relatively high nuclear-to-cytoplasmic ratio and hyperchromatic nuclei. Mitotic figures are not common, and necrosis is infrequent. Diagnosis of CS is based on criteria for malignant cartilaginous tumors of bone origin first set down by Lichtenstein and Jaffe. Based on their cellularity and degree of anaplasia, chondrosarcomas are classified as either low-, medium-, or high-grade tumors. Most reported chondrosarcomas of the larynx are of low grade. The differential diagnosis includes chondroma, hamartoma, and polyps. Spindle-cell carcinoma can contain metaplastic or malignant cartilage. Laryngeal chondrosarcomas usually arise from the posterior or posterolateral area of the cricoid cartilage, followed by the inferolateral wall of thyroid cartilage, very rarely from the arytenoids, vocal folds and epiglottis. Because the tumor grows slowly, patient may experience subtle symptoms for several years before a diagnosis is made. Symptoms depend on location of the tumor. Lesions arising from the cricoid cartilage usually present with hoarseness, dyspnea and dysphagia. Thyroid cartilage chondrosarcomas may present as an asymptomatic neck mass. As a submucosal tumors it can arise from any of the supporting structures of the larynx. Physical examination of a patient with chondrosarcoma mostly reveals the subglottic or submucosal mass, vocal fold paralysis or displacement of arytenoids cartilage by the tumor. The literature reported that chondrosarcomas presents as a subglottic smooth mass covered by intact mucosa during endoscopy. Radiologically, chondrosarcomas may present as a mass of variable density, showing endolaryngeal spread or extralaryngeal (growth into the surrounding soft tissue). On CT scan chondrosarcomas present punctuate, stippled to coarse (“like-popcorn”) calcification within the tumor. Macroscopically chondrosarcomas are smooth, lobular and they are "crunchy" when cut. Preservation of laryngeal function with surgical eradication of the tumor is the preferred treatment. Conservative surgical approach is the best option. Tumor resection with a margin of normal cartilage is possible by open laryngofissure, thyroectomy, organ preservation with partial laryngectomy, or endoscopic resection. Total laryngectomy is indicated for large primary tumors, that involve more then half of cricoid cartilage and for recurrent tumors not amenable to partial laryngectomy. Conservative surgical
management is supported by the low-grade of this tumor and small difference in survival is reported between patients treated with total laryngectomy (86%) and patients with recurrent tumors treated with salvage surgery (77%). Another option for resection of primary or recurrent tumor is endoscopic CO2 laser. Radiotherapy and chemotherapy does not seem to be effective. Chondrosarcomas are considered to be radioresistant. Metastasis rate range from 2% to 8%. Higher tumor grade is directly correlated with metastasis. The lung is the most common site of metastasis. Tumor-free margin does not exclude a possible recurrence, rates between 18%-40% have been reported. Recurrence is related to the tumor grading. A long term follow-up is necessary to detected a tumor recurrence as early as possible.

CONCLUSION

The etiology of chondrosarcomas is unknown and several theories have been proposed. Because the tumor grows slowly, patient may experience a subtle symptoms for several years before a diagnosis is made. Physical examination of a patient with chondrosarcoma mostly revealed the subglottic or submucosal mass, vocal fold paralysis or displacement of arytenoids cartilage by the tumor. The diagnosis and management of these tumors are challenge, because of their infrequent and indolent pattern of growth.

CONSENT

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

COMPETING INTERESTS

The authors declare that they have no competing interests.

REFERENCES

Chondrosarcoma of the larynx