Primary tracheal adenocystic carcinoma and tracheal tumors during pregnancy

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Abstract

Cancer complicates approximately 0.1% of all pregnancies. Primary tracheal carcinoma is one of very rarely seen tumors and the rate of its being seen makes up approximately % 0.2 of all tumors of respiratory tract. The patient, 28 years old, who has 28-weeks-pregnant, was diagnosed with primary tracheal adenocystic carcinoma. Patient was made operation as thoracotomy and tracheal tumor was removed at the 28th week of pregnancy. Patient was delivered with sectio abdominale at the 39th week of pregnancy. Primary tracheal adenocystic carcinoma is very rarely seen tumors and it is the first tracheal ACC with pregnancy case in literature to have been detected and surgically treated during pregnancy. We discussed primary tracheal adenocystic carcinoma and tracheal tumors during pregnancy with literature.

Introduction

Cancer complicates approximately 0.1% of all pregnancies and, in order of frequency, the incidence of cancers by site are breast, melanoma, thyroid, cervix, lymphoma, and ovary.[1] Prevellance of tracheal tumors determine approximately % 0,2 of all tumors of respiratory tract.[2] Primary malignant tumors in the tracheobronchial tree can produce symptoms of airway obstruction (dyspnea, wheezing, stridor), mucosal irritation and ulceration (cough, hemoptysis), or direct invasion and involvement of adjacent structures (recurrent laryngeal nerve palsy, dysphagia). Signs and symptoms from distant metastases are uncommon, being seen in less than 10% of patients.[3,4]

Adenocystic carcinoma (ACC) is a salivary gland tumor which is slowly progressive and make metastasis to lungs, brain and bone metastasis in late period. Because of the slowly progressing, it can not give any symptoms until airway obstruction.[5] ACC is the most common type cancer of salivary gland origin in the central airway.[6] ACC has an equal sex distribution and commonly occurs in the 4th and 5th decades of life.[7] Metastases are very unusual, with recurrence more often being local.[3] ACC tends to occur in the central airways such as the trachea, main bronchus, or lobar bronchus; a peripheral or segmental location is uncommon. These tumors have a striking tendency toward submucosal extension and manifest with circumferential and infiltrative growth.[6-8]

In this study, we indicated primary tracheal ACC with pregnancy as a rare case and it was discussed tracheal tumors during pregnancy with literature.

Case Report

The patient was a 28-years-old woman who had a 28-weeks-intrauterine single pregnancy. The patient was taken diagnosis as asthma bronchiale for 4 years ago. The patient was came to emergency service, she had complaint with dyspnea and cyanosis the the patient was hospitalized. She was transferred to intensive care unit because of cyanosis, lethargy and decreasing of blood partial oxygen saturation on the second day of hospitalization. Endotracheal tube was put and mechanichal ventilation was started because of decline of blood oxygen saturation. On the third day of hospitalization, the cardiopulmonary arrest was developed, cardiopulmonary resuscitation was done immediately. However, O_2 saturation was detected as lower in spite of mechanic ventilation and metabolic acidosis was detected in blood gas test analysis. When the endotracheal aspiration, abandon bleeding was detected, thus thorax computerized tomography was planned. In tomographic evaluation, it was seen solid, well contoured, 29×36 milimeters lesion and closely localizated tracheal bifurcation. The lesion almost completely obstructed to tracheal lumen in tomographic imagination. (Figure 1)

In bronchoscopic evaluation indicated that straight, noduler lesion localizated near of tracheal bifurcation and 90% obstruction of tracheal lumen was determined. Thoracotomy was done and tracheal lesion were completely removed. In intraoperative evalution, it wasn't seen any detectable lesion in thorax. Obstetrical ultrasonographies were done in preoperative and postoperative periods and no problems were detected. On the second day of postoperation the patient was disconnected from mechanic ventilator and since her spontaneous breathing and O_2 saturation were normal she was tranferred to chest diseases clinic. On the seventh day of postoperation she got discharged pregnant. On the tenth day of postoperation, pathology result primary tracheal adenocystic carcinoma came out. Cytologic evaluation with hematoxylene eosin was demonstrated that adenoid cystic carcinoma with uniform hyper chromatic basaloid cells surrounding acellular spaces containing mucoid and hyaline material, tumoral infiltration of tracheal epitelium was determined by ACC, widespreadly tumoral infiltration of submucosal tracheal site and tracheal cartilage was infiltrated by tumor (Figure 2). Because of the operation was done under urgent circumstances and tracheal mass demonstrated benign morphologic feautures, it had been made local tumoral resection. But pathology report was came as ACC, radiotheraphy was planned after the surgery. However, it was scheduled after the birth. At postoperative period pregnancy follow up was found normal. However, the patient was enlighted that because of hypoxic circumstances in time of preoperative and operation periods, the baby could be affected and developed permanent sequel. On the 39th week of pregnancy, a 3680 gr healthy male baby was delivered with sectio abdominale. In observation of the baby one year after birth, no developmental problems were observed. It was not detected any lymph node involvement and metastasis in thorax and other organs with PET scan and MRI imagination after the postpartum 1th month.

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Radiotheraphy was applied by the radiation oncology department after the 6 months after birth. When the radiotherapy was started, lactation was stopped.

Discussion

Primary carcinomas of the trachea are rare tumors, occurring at a percent of 2.6 new cases per 1,000,000 people per year. Adenoid cystic carcinoma (ACC) was the second most common tumor (16.3%) among primary tracheal cancers.[9] As symptoms, pulmonary metastasis are the most common ones but brain, bone, liver, kidney, skin even abdomen and heart metastasis have been reported as well.[10] 40-50% of primary tracheal ACC tumors are located in lower trachea and tracheal bifurcation, 30-35% in the upper and only 10-15% in the middle trachea.[11] Histologically ACC are typical, long, cylindrical structures lined by small cuboidal cells with deeply eosinophilic cores of basement-membrane-like material. As a result of its this feauture it is also cylindroma.[2] Fifty-one patients, including 44 (%86) malignant and seven(%14) benign tumors of airway of were managed in a 14-year period. Among 18 patients with adenoid cystic carcinoma 13 (72%) were resected. Overall 1-, 2-, 5-, and 8-year survival was 90.9%, 90.9%, 77.9%, and 19.5%, respectively.[12] It was reported two cases. Case 1 is a 34-year-old female who presented with squamous cell carcinoma of the tongue at 29 weeks' gestation. Partial glossectomy, selective neck dissection was performed at 31 weeks. She underwent induction and early delivery at 38 weeks prior to receiving radiotherapy. Case 2 is a 36-year-old female who presented with carcinoma of the cervical esophagus complicated by tracheal invasion at 13 weeks' gestation. Pregnancy was terminated at 16 weeks. She received a course of neoadjuvant chemoirradiation.[13] It was indicated that one case of adenoid cystic carcinoma of trachea metastatic to the placenta during pregnancy in literature until now.[14] Treatment options of ACC are surgery alone, radiation alone and combination of surgery and radiation. Studies showed that survival rates of 5 years was determined % 66-100 and % 51-62 for 10 years after the primary surgery.[10] 12 laryngotracheal, 58 tracheal and 38 carinal resections for primary ACC in 108 consecutive operative survivors between 1962 and 2007 was conducted. Median overall survival and disease-free survival for the entire group were 17.7 and 10.2 years, respectively. It was concluded that after tracheal resection for ACC, limited tumor extent and complete resection are associated with longer overall and disease-free survival.[15] 43 patients of primary tracheal or bronchus adenoid cystic carcinoma treated from 1958 to 2007. The 1-yr, 3-yr, 5-yr survival rates of the 43 patients above were 100% (41/41), 89.5% (34/38), 87.1% (27/31), respectively. It was concluded that the best treatment is early detection and taking measures of operation plus radiotherapy.[16]

Squamous Cell Carcinoma (SCC) is the most common primary tracheobronchial malignancy and develops mainly in the 6th and 7th decades of life.[17] This tumor is strongly associated with habitual cigarette smoking, affecting men two to four times more frequently than women.[3] Approximately 40% of tracheobronchial SCCs have been reported to occur before, concurrently with, or after carcinoma of the oropharynx, larynx, or lung.[3,18] Management of such patients must consider the gestational age of the pregnancy and must include alternatives to current treatments to accommodate the individual's wishes regarding her pregnancy.[1] There are a few articles about squamous cell carcinoma in respiratory tract during pregnancy. It was indicated that 31-year-old patient with squamous cell carcinoma with multiple bone metastases and a 34-year-old patient with poorly differentiated lung carcinoma with brain metastasis and left hemiparesis had developed initially during the third trimester.[19] It was supposed that the pregnancy could have contributed to the evolution of the disease, especially because of the increased levels of gestational hormones, particularly the estrogen, because its receptors have an important role in regulating growth and in the differentiation of several tissues facilitating like this, the development of the neoplasia, and complicating its early diagnosis.[20]

Pulmonary or bronchial carcinoid tumors account for over 25% of all carcinoid tumors and for 1-2% of all pulmonary neoplasms.[21] Carcinoid tumors in the tracheobronchial tree are frequently typical and usually involve the main, lobar, or segmental bronchi.[22] It was indicated two cases of tracheal carcinoid during pregnancy.[23,24] A woman 29 weeks pregnant presented with acute respiratory insufficiency and massive hemoptysis of unknown origin. An emergency cesarean section was performed to avoid hypoxic fetal damage during episodes of maternal hypoxemia. The tumor obstructed the distal portion of the trachea, leading to life-threatening complications during tracheal intubation because of its histological characteristics and placement.[24]

Mucoepidermoid carcinoma of the tracheobronchial tree is a rare airway tumor, composing only 0.1-0.2% of the primary lung malignancies. The tumor is believed to originate

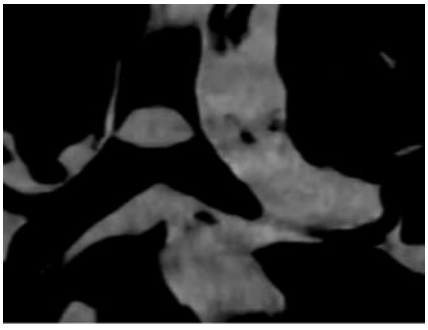

Figure 1. Tracheal lesion in computed Tomography imagine. Lesion size was 29×36 mm and closely localizated tracheal bifurcation. The lesion almost completely obstructed to tracheal lumeni.

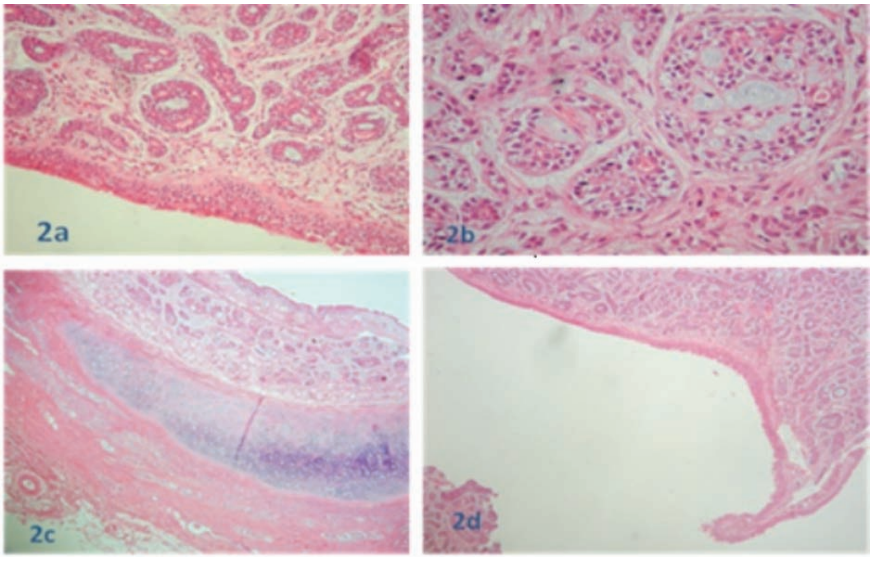

Figure 2. Primary tracheal Adenocystic carcinoma. (a) Widespreadly tumoral infiltration of submucosal tracheal site (H. E. x200); (b) Adenocystic carcinoma tumor tissue(H. E. x400); (c) Tracheal cartilage was infiltrated by tumor (H. E. x40); (d) Tumoral infiltration of tracheal epitelium (H. E. x40).

from the minor salivary glands lining the tracheobronchial tree.[25,26] Two mucoepidermoid carcinoma in pregnancy were described in literature. Case 1; a 23-year-old pregnant female in her 39th week.[27] Case 2; A 30-year-old, 36-week pregnant, female was admitted to the hospital with progressive asthma and wheezing. Following ventilating tracheal bronchoscopy with dilatation of the trachea and establishment of an airway through the tumor, a cesarean section was performed with resultant delivery of a healthy, female, and infant. A partial abruptio placenta was noted.[28] Malignant granular cell tumor is a rare neoplasm reported to occur at various sites in the body.[29] Granular Cell Tumors are rare mesenchymal soft tissue tumors that arise throughout the body and are believed to be of neural origin. They often present as asymptomatic, slow-growing, benign, solitary lesions but may be multifocal. 1-2% of cases are malignant and can metastasis.[30] It was indicated two cases of malignant granular cell tumor during pregnancy.[31,32] The patient was a 21-year-old woman, who was 5 months pregnant. The tumor occurred in the retrotracheal space, extending from the level of the larynx to the thoracic inlet. The patient refused further treatment and died 7 months later. The diagnosis of a malignant granular cell tumor should be considered in cases with aggressive clinical findings and some histologic features, such as necrosis, nuclear atypism, and mitotic activities, which could suggest the malignant behavior of this neoplasm.[31]

Kaposi's sarcoma, confined primarily to the trachea, is one of the rarest tumors. The one case was described of Kaposi's sarcoma during pregnancy in literature till now. The clinical and histological picture of a case of Kaposi's sarcoma of the trachea in a young, pregnant woman, presenting with severe airway obstruction, is described here. The emergency tracheoscopy dislodged a mass from the trachea, which turned out to be a Kaposi's sarcoma on histology. Although Kaposi's sarcoma is one of the manifestations of AIDS, this patient had neither any underlying immunodeficiency nor any skin, visceral or lymphatic lesions.[33]

Inflammatory myofibroblastic tumor, also known as inflammatory pseudotumor, is a benign lesion predominantly found in the lung and abdomen. Sporadic cases have been reported in the trunk, genitourinary tract, and extremities as well as in the head and neck. Of critical importance is this entity's correct histopathologic diagnosis that differentiate it from malignant neoplasms such as spindle cell carcinoma and fibrosarcoma, benign tumors such as neurofibroma, and other pseudoneoplastic lesions such as nodular fasciitis. It was indicated a unique case of inflammatory myofibroblastic tumor of the trachea presenting with acute upper airway obstruction in a pregnant woman.[34]

Paraganglioma with tracheal location is a rare tumor. Tumors tend to invade bordering structures and may also form metastasis. Up to 50% of patients are asymptomatic and diagnosis is incidental. Presenting symptoms are related to catecholamine hypersecretion or to a mass effect. Complete surgical resection remains the standard of care due to malignant potential of the tumor and poor response to chemotherapy or radiation. Strategic location of the tumor in proximity to great vessels, trachea, and recurrent laryngeal nerve poses challenge for the surgeon.[35] Twelve cases with involvement of the trachea have been reported in literature, including six cases of solely tracheal paragangliomas. It was indicated a case of a paraganglioma confined to the tracheal wall in a 33-year-old pregnant woman. The tumor was locally resected. Follow-up 17 months later showed no evidence of relapse.[36]

The cancer and cancer surgery in pregnancy is seen very rarely. The timing of treatment is an important determinant on fetal wellbeing. Diagnostic and treatment modalities may harm the fetus, while delaying or choosing suboptimal treatment in order to preserve fetal health may worsen maternal outcome. A multidisciplinary approach should be adopted to enable parents and clinicians to make the best clinical decision. In literature, we have not come across a primary tracheal ACC case detected during pregnancy. Previous studies indicated that the rate of detection a cancer during pregnancy is 1/1000 pregnancies and 118 pregnancies with cancer has been detected at last ten years, only one of which was found as oropharyngeal ACC.[37] We indicated and treated that the first primary tracheal ACC with pregnancy case have been detected and surgically treated in literature. Although, patient was made a major surgery and malign diagnosis, we waited till 39th week of pregnancy then patient was delivered with section abdominale. Radiation treatment was applied after the lactation period and the patient remained healthy for one year after treatment. Although fetus was exposed to intense maternal hypoxia at intrauterine period, there was not detected any sequel or developmental problem for baby's observations for one year after birth . Even though it is rare case for women during pregnancy, one should keep rarely seen systemic diseases in mind.

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