Case Report

A RARE CASE OF MUCO EPIDERMOID CARCINOMA IN THE TRACHEA - IN A CHILD

Shankar Tati,¹ Shobhan Babu .A², Nagaraj.K³ Ranganathswamy.D⁴.

1. Professor, Department of ENT. Osmania Medical College, Koti, Hyd.

2.Associate Professor, Department of ENT. Osmania Medical College, Koti, Hyd.

3. Assistant Professor, Department of ENT. Osmania Medical College, Koti, Hyd.

4. Assistant Professor, Department of ENT. Osmania Medical College, Koti, Hyd.

Corresponding Author: Dr. T. Shankar, Professor, Department of ENT, Osmania Medical College / Govt. ENT Hospital, Koti, Hyderabad. Telangana State.

Abstract :

Of all primary tumors of the trachea, 80% are malignant, benign tumor can arise from any of the tissues present in the trachea, malignant tumors probably follow a carcinogenesis similar to that of lung cancers. Primary tracheal tumors are very rare, occurring in approximately 0.1 person per 1,00,000 population mostly 80-90% are malignant, treatment of choice is surgery, radiotherapy depending up on the histological type of malignancy, here we are presenting a rare case of mucoepidermoid carcinoma in the trachea in a 09 year old child, excised by endoscopic approach, with post operative radiotherapy, mucoepidermoid carcinoma is a rare tumor which arises from the bronchial glands. Patient doing well, still on tracheastomy tube we are planning for decanulation since post operative bronchoscopy is normal.

KEYWORDS :_adenoidcystic carcinoma, brachytherapy, cryosurgery, mucoepidermoid carcinoma, trachea, radiotherapy

INTRODUCTION

The trachea (Wind Pipe) is the tube that connects oropharynx to the Lungs. The Trachea is about 10-16cms, long and is made up of rings of tough, fibrous tissue (cartilage), it goes on to divide into the two airway which supply air to each lung, it lies in the neck infront of the oesophagus.



Figure No.1Anatomy of Trachea.

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Tracheal neoplasms occur infrequently, accounting for less than 1% of all malignancies ⁽¹⁾, of all primary tumors of the trachea 80% are malignant, sqamous cell carcinoma 44.3%, followed by adenoid cystic carcinoma 16.3%, the remainder is widely varied and includes both malignant and benign ⁽²⁾, despite their rarity usually insidious on set often leads to a delay in making these potentially treatable lesions difficult to treat and often fatal, thus, early diagnosis is the most important factor affecting overall survival, the mucoepidermoid carcinoma is rarest (13%) form of malignant tumors present in the trachea, and in our case we have totally excised by endoscopic debridment after preliminary tracheastomy.

CASE REPORT :

A 09 year old female child presented with complaint of cough with hemoptysis, shortness of breath, history of weight loss since 06 months, no other complaints were present.



Figure No.2

After all necessary investigations patient was subjected for elective tracheastomy and under endoscopic guidance tumor identified arising from the posterior wall of trachea on (R) side, the tumor meticulously dissected and removed from the trachea under general anesthesia, specimen sent for HPE, patient recovery was uneventful patient was kept on tracheastomy tube the HPE report confirmed as mucoepidermoid carcinoma of low grade, patient was sent for radiotherapy. Patient is under follow up for 1year, the diagnostic bronchoscopy was done, after 1year no recurrence is found, decanulation is planned.





Figure No.3 Endoscopic picture

Figure No.4 Post OP Photo with tracheostomy.

<u>HPE REPORT :</u> Multiple and serial sections studied shows histological features of mucoepidermoid carcinoma – Low grade - Trachea







HPE Report.

DISCUSSION :-

Primary tracheal tumors are very rare, the incidence of tracheal Carcinoma is much lower than laryngeal or endo bronchial cancer, lung cancers are 180 times more common than tracheal malignancies, smoking is a commonly associated risk factor ⁽³⁾, tracheal tumors are 3 times more common in male than in females, peak incidence occurs in the 5th and 6th decades of life ^(4,5), in adults, approximately 90% of primary tracheal tumors are malignant ^(6,7), the most common of these malignant neoplasms are sqamous cell carcinoma and adenoid cystic

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carcinoma, together comprising approximately 75% of all primary tumors of the trachea. Squamous cell carcinoma are invariably associated with cigarette smoking. Grossly they range from localized exophytic lesions to diffuse ulcerating tumors. Unfortunately, tracheal squamous cell carcinomas are frequently locally advanced and unresectable at the time of presentation. Grillo reported approximately one third of patients with either mediastinal or pulmonary metastases at initial diagnosis, further more metachronous lesions were also common as 40% reportedly had prior, concurrent, or later carcinoma of either the oropharynx, larynx, or lung.

In contrast, adenoid cystic carcinomas are not associated with cigratte smoking. These tumors have a propensity to spread along both submucosal and perineural planes. Regional lymph node metastases are reported in 10% of patients an remote metastases to lung, bone, and brain have been observed ⁽⁸⁾. Despite these malignant features, adenoid cystic carcinoma often follows a prolonged course. Slow and insidious progression, often over several years, is characteristic of even untreated cases.

A variety of rare, primary, tracheal malignancies including carcinosarcoma, chondrosarcoma, leiomyosarcoma, mucoepidermoid carcinoma, and carcinoid tumors have also been reported. The overall experience with these tracheal tumor has been so limited that generalizations with regard to natural history and optimal management cannot confidently be made. Tracheal invasion from adjacent larvngeal, thyroid, lung, or esophageal carcinoma is more commonly encountered than primary tracheal malignancy. Unfortunately, these secondary tumors are often not amenable to curative tracheal resection. Squamous cell carcinoma of the larynx may involve the upper trachea by direct extension or recur at the tracheal stoma after laryngectomy. In this situation, local irradiation is the treatment of choice. Radical resection of these recurrences with creation of mediastinal tracheostomies has been reported. Significant morbidity and high tumor recurrence rates, however, fail to justify this extensive surgical approach ⁽⁹⁾. Similarly, resection of the trachea for direct involvement by esophageal carcinoma is generally not advised. The extent of local involvement and the likelihood of distant disease recurrence are usually so great that a curative resection is unlikely in such cases. The secondary tumor perhaps most amenable to tracheal resection and cure is thyroid carcinoma. Papillary and follicular thyroid carcinomas are reported to directly invade the airway in 1% to 6.5% of patients ⁽¹⁰⁾. Tracheal invasion tends to be more common in poorly differentiate tumors and in older patients. In addition, the prognosis of thyroid carcinoma correlates with both the site and depth of tracheal invasion (11,12). Anaplastic thyroid carcinoma is often widely invasive at initial presentation and usually not resectable.

The most common symptoms of Tracheal cancer are.

- A dry cough.
- A hoarse voice.
- Breathlessness.
- Difficulty in Swallowing.
- Coughing of blood.
- Wheezing
- Fever, chills and chest infection that are keep coming back.

Cancer of the Trachea is rare and can be difficult to diagnose, it may be mistaken for asthma or bronchitis, which sometimes results in a delay in the diagnosis.

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Tomography and CT scan are the most helpful methods of radiologic examination of tracheal tumors ⁽¹³⁾, routine chest X-rays on the other hand are relatively insensitive in detecting tracheal pathology, CT is currently considered the standard imaging modality for diagnosis and staging of tracheal tumors. CT demonstrates the intraluminal and extraluminal extent of tumor and delineates the relationship of the tumor to adjacent structures. Contrast esophagography, when performed with CT, may help to identify esophageal invasion. Threedemensional (3D) helical CT scanning is a very useful imaging modality which has replaced the conventional tomogram ⁽¹⁴⁾, to date magnetic resonance imaging (MRI) provides no clear advantage over CT imaging in the evaluation of tracheal tumors. Pulmonary function testing may suggest the presence of upper airway obstruction. An obstructive flow pattern which does not respond to bronchodilator therapy should arouse suspicion of a fixed upper-airway obstruction. Flow-volume loops, with characteristic flattening of both inspiratory and expiratory phases, may provide further evidence of upper airway obstruction. Bronchoscopy is the mainstay of diagnosis and staging of tracheal neoplasms, biopsies are obtained for tissue diagnosis and histological classification of malignancy which helps in planning of treatment.

TREATMENT OF TRACHEAL CANCERS. The treatment for tracheal cancers depend on a number of features including, general health of the patient, position and size of the cancer and whether it has spread anywhere else in the body. The main treatment for cancer of the trachea are.

- Surgery •
- Radiotherapy
- Chemotherapy

SURGERY: In early, small cancers an operation may be able to completely remove the tumor. This is specialized surgery and is only carried out in specialist centres. However, in many cases too much of the length of the trachea is affected to remove the cancer and re-join the cut ends of the trachea (Tracheal resection). As well as removing the cancer, the surgeon also usually removes some healthy looking tissue surrounding it (known as a clear margin). Surgery represents the primary curative treatment for tracheal tumors. Current tracheal mobilization techniques, including suprathyriod laryngeal release ⁽¹⁵⁾. Radiotherapy may be given after surgery to try to reduce the chances of the cancer coming back. It may also be done if there were any cancer cells left behind after the operation.

RADIOTHERAPY : Radiotherapy uses high energy x-rays to destroy cancer cells, while doing as little harm as possible to normal cells. It can be used on its own to cure people with early, low-grade cancer of the trachea who are unable to have surgery, radiotherapy is also given after surgery to reduce the chances of the cancer coming back (adjuvant radiotherapy) or to relieve symptoms (palliative radiotherapy).

CHEMOTHERAPY: chemotherapy is the use of anti-cancer (cytotoxic) drugs to destroy the cancer cells. They work by disrupting the growth of cancer cells. Chemotherapy is usually used to help control the cancer or its symptoms (palliative chemotherapy). Chemotherapy is rarely used for adenoid cystic cancers of the trachea.

LASER TREATMENT : Laser treatment relieves symptoms by burning the tumor with a laser light. Its carried out under a general anaesthetic and a flexible fibre is passed through the bronchoscope to aim the laser beam at the tumor. The beam is aimed at the tumor and www.earthjournals.org Volume 3, Issue 4, 2014

destroys as much of it as possible. There aren't usually any side effects and patient can go home the following day or the same evening.

<u>CRYOTHERAPY</u>: Cryotherapy uses liquid nitrogen, which is extremely cold, to freeze and destroy cancer cells. It's carried out under general anaesthetic. Using a bronchoscope, instrument called a cryoprobe is placed close to the tumor. Liquid nitrogen is then circulated through the probe to kill off parts of the tumor.

PHOTODYNAMIC THERAPY (PDT) : Photodynamic therapy (PDT) uses laser, or other light sources, combined with a light-sensitive drug to destroy cancer cells. The light-sensitive drug is given as liquid into a vein. After waiting for the drug to be taken up by the cancer cells, the laser light is directed at the tumor using a bronchoscope. This starts the drug working to destroy the cancer cells. PDT will make the patient temporarily sensitive to light and the patient will need to avoid bright light for between a few weeks and a few months, depending upon the photosensitizing drug that is used. PDT is relatively new treatment and is only available at some centres.

<u>AIRWAY STENTS</u>: Sometimes the airway can become narrow due to pressure on it from the outside. This can sometimes be relieved using small device called a stent, which is put inside the airway to hold it open. The most commonly used stent is small wire frame. It's inserted through a bronchoscope in a folded up position and as it comes out of the end of the bronchoscope it opens up, like an umbrella. This pushes the walls of the narrowed airway open. Side effects from an airway stent can include infection, blockage with mucus and irritation of the airway.

<u>**CONCLUSION**</u>: Malignant tracheal tumors are infrequently in clinical practice, tracheal involvement usually results from secondary invasion by adjacent thyroid, laryngeal, oesopharyngeal or lung carcinomas. Primary tracheal tumor are rare and usually are either adenoid cystic carcinoma or scquamous cells carcinoma in our case the case is mucoepidermoid carcinoma is a one of the rarest form of malignancy in the trachea (13%) that to in a 09 years old child and treated by endoscopic excision after doing preliminary tracheastomy, patient also had post operative radiotherapy, patient is followed for one year no recurrence is found and planning for decannulation.

REFERENCES :

1. Gaissers HA, Mark EJ. Tacheobronchial gland tumors. Cancer Control. Oct 2006;13 (4):286-94. (Medline).

2. Urdaneta AI, Yu JB, Wilson LD. Population based cancer registry analysis of primary tracheal carcinoma. Am J Clin Oncol. Feb 2011;34(1):32-7. (Medline).

3. Velez Jo ET, Morehead RS. Hemoptysis and dyspnea in a 67 – year old man with a normal chest radiograph. Chest . Sep 1999; 116(3):803-7. (Medline).

4. Pearson FG, Todd TRJ Cooper JD. Experience with primary neoplasms of the trachea. J Thorac Cardiovasc Surg 1984;88:511-516.

5. Grillo HC, Mathisen DJ. Primary tracheal tumors: treatment and results. Ann thorac Surg 1990;49:69-77.

6. Culp OS. Primary carcinoma of the trachea. J.Thorac Cardiovasc Surg 1938:7:471-478.

7. Ampil FL. Primary malignant tracheal neoplasms: case report an literature radiotherapy review.J surg Oncol 1986;33:20-23.

8. Pearson FG, Thompson DW, Weissberg D et al. Adenoid cystic carcinoma of the trachea. Ann Thorac Surg 1974: 18:16-29. <u>Cross Ref Medline</u>

9. Krespi YP, Winster CF, Sisson GA. Immediate reconstruction after total larynopharyngoesophagectomy and mediastinal dissection. Laryngoscope 1985; 95:15-161.

10. Lawson W, Som MP, Biller HF. Papillary carcinoma of the thyroid invading the upper air passages. Ann Otol Rhinol Laryngol 1977; 86:751-755.

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11. Tsumori T, Nakao K, Miyata M et al. Clinicopathologic study of thyroid carcinoma infiltrating the trachea. Cancer 1985; 56:2843 – 2848.

12. Shin DH, Mark EJ, Suen HC et al. Pathological staging of papillary carcinoma of the thyroid with airway invasion based upon the anatomic manner of extension to the trachea. Hum pathol 1993;24:866-870.

13. Li W, Ellerbroek NA, Libshitz HI. Primary malignant tumors of the trachea: a radiologic and clinical study. Cancer 1990;66:894-899.

14. Kauczor H, Wolcke B, Fischer B et al. Three – dimensional helical CT of the tracheobronchial tree: evaluation of imaging protocols and assessment of suspected stenoses with bronchoscopic correlation. AJR 1996; 167:419-424.

15. Montgomery WW. The surgical management of supraglottic and subglottic stenosis. Ann Otol Rhino Laryngol 1968;77:534-546.