OROPHARYNGEAL LEIOMYOSARCOMA

Ravinder Sharma[1], Isha Tyagi[2], C. K. Panday[3], Vini Tandon[4]

ABSTRACT A unique case of Oropharyngeal leiomyosarcoma in a 32 years old male is presented with the review of literature The details of clinical, radiological, histopathological and operative findings are discussed The treatment of choice is complete tumor removal The patient was symptom free with no evidence of loco regional or distant metastasis at one-year follow up

INTRODUCTION

Leiomyosarcoma refers to the malignant tumor of smooth muscle origin They are rare lesions of head and neck and are more commonly found in the uterus and grastrointestinal tract[5] The upper aeiodigestive tract is further much rarer location of this tunor because of paucity of smooth muscles in this area[8] A review of literature revealed 10 cases of leiomyosarcoma in the upper aerodigestive tract[8]

We present a unique case of a large oropharyngeal leiomyosarcoma The clinical features, operative findings, histopathology and CT findings are discussed with the review of literature

CASE REPORT

A32 year old male presented to us with history of gradually progressive dysphagia for solid food for last 18 months, progressive weight loss for last 10 months, change in voice for last 6 months, gradually progressive dyspnoea for last 3 months and stridor for last 10 days There was no history of cough with expectoration and hemoptysis

General examination was normal except for the lean and thin appearance of the patient Examination of oropharynx showed a large smooth surfaced exophytic penduculated

[1] Senior Resident [2]Associate Professor (Neurotology) Department of Neurosurgery [3]Assistant Professor Department of Anaesthesiology [4]Senior Resident, Department of Pathology Sanjay Gandhi Post Graduate Institute of Medical Sciences Lucknow- 226 014

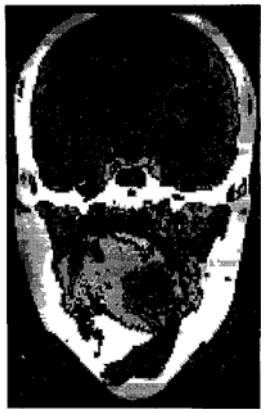

Fig I Coronal CT scan showing heterogeneously enhancing mass lesion with focal hypodense areas in oropharynx and extending lower down in hypopharynx

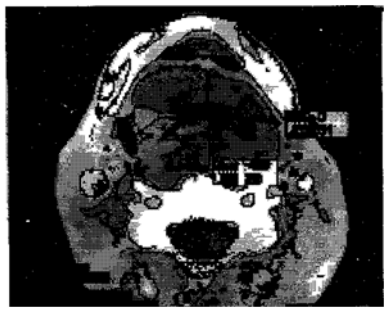

Fig II Axial CT scan showing nearly complete obliteration of hypopharynx with a heterogeneously enhanceing mass lesion (with focal hypodense areas suggestive of necrosis)

Fig III Photomicrograph showing intersecting fascicles of spindle cells with mild anisonucleosis and few mitotic figures (H & E 100 x), suggesting histological diagnosis of leiomyosarcoma

Fig IV Photomicrograph showing subepithelial infilteration by intersecting fascicles of spindle cells and focal myxoid degeneration in stroma (H & E 100x)

mass completely filling the oropharynx and upper and lower limits of the mass could not be assessed because of the lack of any space to mobilize the mass. There was no evidence of clinically significant neck nodes. X-ray chest was normal.

Keeping in view the possibility of respiratory obstruction, urgent tracheostomy was done. CT scan was done to assess the anatomic extent of the mass lesion. It showed a heterogeneously enhancing mass lesion with focal hypodense areas (suggestive of necrosis) in nasopharynx, oropharynx and extending downwards in to the hypopharynx (Fig.I, II). It was pedunculated with attachment to the posterior pharyngeal wall in the oropharynx.

Examination under general anesthesia showed submucosal large polypoidal mass attached to the posterior pharyngeal wall in the oropharynx. On palpation, it was soft to firm in consistency, extending higher up in the nasopharynx, completely occluding it and lower down in the hypopharynx.

Operative removal was done by a transoral approach. The tumor was excised by widely dividing its pedicle and removed through the oral cavity. The defect in the oropharyngeal wall was repaired by mobilizing oropharyngeal mucosa. It was smooth surfaced 9 x5 x7 cm in size and soft to firm in consistency. The cut surface was grayish white, variegated with focal areas of myxoid degeneration.

The sections from the oropharyngeal mass were lined by stratified epithelium and displayed a highly cellular tumor in the subepithelial zone composed of spindle cells arranged in whorls, sweeping fascicles and sheets with mild anisonucleosis and few mitotic figures (Fig.III). Areas of myxoid degeneration were especially seen in the subepithelial zones (Fig.IV). It was reported as leiomyosarcoma. Postoperatively patient had remarkable recovery. Tracheostomy was closed 12 hours after surgery and patient was discharged on fifth postoperative day in satisfactory condition. The patient was asymptomatic with no clinical or radiological evidence of recurrence or residual disease or distant metastasis at 6 months and 1 year follow up.

DISCUSSION

Leiomyosarcoma can occur at any site in the body where smooth muscles are present. They are quite common in the genital tract and the gastrointestinal tract. Rarely, they are found in the head & neck. Majority of head & neck leiomyosarcoma have been described in the scalp, superficial soft tissue and oral cavity[3-7]. Other reported sites include nose, trachea, neck veins, external auditory canal and maxillary sinus. Allen CM et al[1] discussed three cases of metastatic leiomyosarcoma to the oral region. Lalwani AK et al[6] reported a case of paranasal leiomyosarcoma after cyclophosphamide and irradiation in a patient with a history of destructive nasoantral process.

Leiomyosarcoma occurring in the superficial tissue of the head & neck probably arises from the walls of the blood vessels and erector pili musculature of the skin[3] Smooth muscle tumors in the deeper regions may arise from the smooth muscle with in the tunica media of a blood vessel or possibly by aberrant mesenchymal differentiation.

CT scan is of great help in such cases to evaluate the extent of mass and its relationship with vital structures. Imaging of muscle tumors of the head & neck shows a nonspecific mass that may have associated bone destruction or erosion[2]. Muscle tumors are nearly isodense to muscle on plain CT scan and isointense to muscle on T_1 weighted. MRI.T_2 weighted MRI demonstrates homogenous to heterogeneous moderate high signal.[10]

Histological differentiation of leiomyosarcoma and leiomyoma depends on cell size, pleomorphism and increased mitotic figures. Increased cellularity, high mitotic activity and cellular atypia are suggestive of malignancy[9]. The diagnosis in our case was based on histopathology. The use of flow cytometry for identifying deoxyribonucleic acid ploidy can also assist in predicting tumor behavior. No such trial was done in our case.

Complete excision combined with neck dissection for overt nodal metastasis is the recommended treatment of choice[3]. However some authors have noted that these tumors are well circumscribed[7] (as in our case) and wide local excision remains the preferred treatment when technically feasable[4]. Considering the paucity of number of such cases in head & neck available in the literature it is difficult to design definite treatment protocol of this tumor. However Grant CS et al[4] experiences with gastric leiomyosarcoma suggests that wide local excision is the suitable treatment option for such patients. The patient in our case had one year of clinical & radiological disease free follow up. Raj Sindhwani et al[8] treated a case of epitheloid leiomyosarcoma of larynx with surgery and postoperative radiotherapy. We feel that if onco-surgical clearance is inadequate and to cover any microinvasion toward the prevertebral fascia and vertebrae, patients may have some benefit with radiotherapy and/or chemotherapy.

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Address for Correspondence :
Dr. Isha Tyagi
Associate Professor (Neurotology)
Department of Neurosurgery
Sanjay Gandhi Post Graduate Institute of Medical Sciences,
Raebareli Road, Lucknow – 226014, India