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Case Report

A rare case of atypical lipoma in retropharyngeal space and review of literature

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ABSTRACT

The term atypical lipoma was introduced to describe the relatively benign course of well-differentiated liposarcomas when they occur in extremities compared to their retroperitoneal counterparts. A 32-year-old lady presented with dysphagia, excessive snoring and difficulty in breathing for three months. On examination, a smooth globular retropharyngeal swelling was seen extending anteriorly till epiglottis obscuring the view of vocal cords. The CECT scan demonstrated features suggestive of a lipomatous lesion with possible sarcomatous change. Excision was done through a transcervical approach and histopathology showed features suggestive of atypical lipoma of retropharyngeal space. As the tumor was removed in toto, adjuvant radiotherapy was not advised. Hence if any swelling in the retropharyngeal space is noted, the probability of soft tissue tumors must be ruled out by a CT/MRI scan before attempting incision and drainage or any other surgical procedure. Postoperative adjuvant radiotherapy is limited to high-grade tumors, incompletely excised tumors and more radiosensitive liposarcoma subtypes such as myxoid sarcomas.

Keywords: Retropharyngeal space, Liposarcoma, Dysphagia, Surgery

INTRODUCTION

Atypical lipomatous tumors are locally aggressive mesenchymal lipogenic tumors that arise commonly in the retroperitoneum, trunk and infrequently in head and neck region. Here we report a rare case of atypical lipoma situated in the retropharyngeal space. The term atypical lipoma was introduced to describe the relatively benign course of well-differentiated liposarcomas when they occur in extremities compared to their retroperitoneal counterparts.[1] Atypical lipomatous tumor and well-differentiated liposarcomas are morphologically and genetically identical with varied nomenclature to avoid both overtreatment and undertreatment.[2] Tumors that are located in the periphery have no risk of metastasis, thus complete resection with negative margins is often curative for them, and the term "Atypical Lipomatous tumor" is more preferred than sarcoma.[2] Only a few cases of retropharyngeal liposarcoma have been reported in the literature.

CASE REPORT

A 32-year-old lady presented to the ENT department with complaints of difficulty in breathing and swallowing for three months, which rapidly progressed in a week. She also complained of change in voice and excessive snoring. On clinical examination, a globular swelling was seen in the oropharynx arising from the posterior pharyngeal wall. Video laryngoscopy showed a diffuse smooth swelling arising from the posterior pharyngeal wall, extending superiorly from the Passavant's ridge and

inferiorly going beyond the arytenoids. The swelling is bulging anteriorly reaching till the free border of the epiglottis and obscuring the view of vocal cords (Figure 1).

Figure 1: A large globular retropharyngeal swelling, anteriorly reaching the epiglottis obscuring the view of vocal cords.

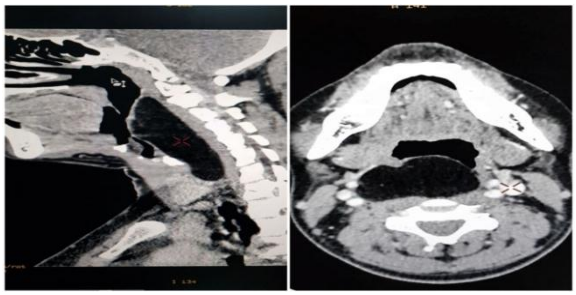

Figure 2: CECT neck showing a large well defined, lobulated, hypodense, fat density lesion with thin enhancing septae involving retropharyngeal space with lateral extension from one carotid to the other carotid artery.

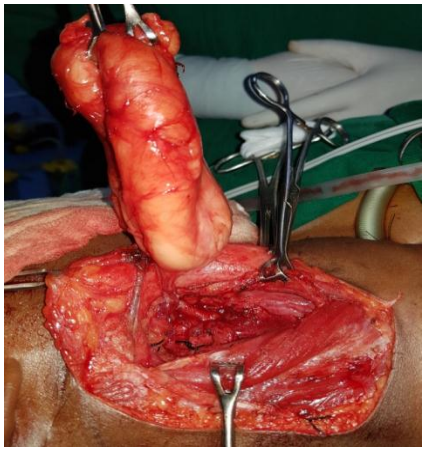

Figure 3: Intraoperative picture showing completely excised tumor by transcervical approach.

The endoscope was gently negotiated in between the epiglottis and the swelling to assess the vocal cord mobility, which was found to be normal. Contrast-enhanced computed tomography (CECT) neck showed a large well defined, lobulated, hypodense, fat density lesion measuring 8.6 × 4.7 × 3.9 cms involving retropharyngeal spaces of oropharynx, hypopharynx and retrolaryngeal spaces of supraglottic, glottic and infraglottic regions causing pharyngeal and supraglottic luminal narrowing with lateral extension from one carotid to the other carotid artery; suggestive of lipomatous lesion. The CECT also showed thin enhancing septae within the lipomatous lesion, suggesting the possibility of liposarcoma (Figure 2). Peroral fine needle aspiration cytology (FNAC) showed features suggestive of an adipocytic lesion. Based on these clinical and radio-pathological findings, a provisional diagnosis of retropharyngeal lipoma was made. Trans-cervical excision of the retropharyngeal lipoma was done after securing the airway with tracheostomy (Figure 3).

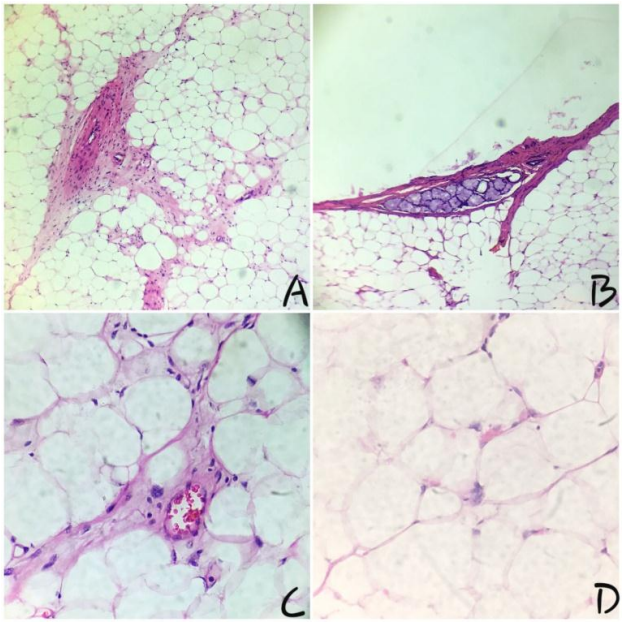

Figure 4: (A) Adipocytes with spindle cells showing fibroblastic and myofibroblastic differentiation (10x); (B) The edge of the tumor showing mucous glands entrapped in the fibrous septa (10x); (C) High-resolution view showing atypical cells with blood vessels lined by endothelial cells. RBCs inside the vessel are seen (40x); (D) High-resolution view showing numerous adipocytes divided by thin fibrous septa (40x).

In the immediate post-op period, the patient developed a non-enhancing retropharyngeal space and cervical hematoma, which resolved in 2 weeks with conservative management. Histopathology examination showed microscopically fat lobules within an encapsulated tumor, adjacent to which few areas of spindle cells with fibroblastic and myofibroblastic differentiation was seen. Edges also showed a few keratinous fibres and a focus of mucous glands with fibrous regions that showed a few giant cells thus suggesting atypical lipoma (well-differentiated liposarcoma) (Figure 4). She was in regular

follow up every month for six months and was recurrence-free.

DISCUSSION

Liposarcomas are rare soft tissue sarcomas that account from 8 to 17% of soft tissue sarcomas. They are commonly seen in the extremities, retroperitoneum, trunk and infrequently in head and neck region with an incidence of 3 to 6%.[3] Cheek, anterior neck, larynx and pyriform sinus are the most common sites (13-14%) while orbit and supraglottic region (7-8 %) are the other head and neck sites where liposarcomas were reported.[4]

Retropharyngeal space is a space bounded anteriorly by the buccopharyngeal fascia, posteriorly by pre-vertebral fascia, extends superiorly from the skull base to mediastinum at the level of T4 vertebrae inferiorly. It's divided into anterior and posterior by alar fascia which extends from skull base to T2 vertebrae. The posterior space is also known as 'danger space' as infections or an abscess within the retropharyngeal space can directly extend into the mediastinum. It is separated from the parapharyngeal space by a thin fascial layer and lies within close proximity to the internal jugular vein, common carotid artery and vagus nerve.

A literature review of retropharyngeal liposarcomas done by Alicandri-Ciufelli et al mentions that dysphagia was the most common presenting symptom (5/9 cases) and the others were dysphonia, neck swelling, snoring and globus pharyngeus.[5] In our case, she presented with a rapid growth of the swelling, which suggested the malignant nature of the tumor.

The differential diagnoses of the retropharyngeal swelling were an abscess, tubercular cold abscess from Pott's spine, cervical vertebra disc prolapse, posterior pharyngeal wall tumors, lipoma, etc. In our case, the patient presented with a smooth retropharyngeal swelling that looked more like an abscess.

Imaging modalities like CT and MRI scans usually helps in differentiating the abscess from other diagnoses. For our patient, CECT scan revealed a large well-defined fat density lesion with thin enhancing septae in the retropharyngeal space, suggesting liposarcomatous lesion and excluding abscess. Murphey et al in his study mentions that well-differentiated liposarcomas are frequently diagnosed in CT or MRI scans, with a largely lipomatous mass (>75% of the lesion) and non-lipomatous components in thick septa or focal nodules.[6] Guo et al also reported a similar case with extensions from the level of the hyoid bone to the superior margin of the mediastinum, where the mass displaced trachea and larynx anteriorly and carotid arteries laterally.[7] In his report just like our case, FNAC showed features that suggested of lipoma, but after excision histopathology diagnosed it as well-differentiated liposarcoma. Azeem et al reported a similar case in the oropharynx but was extending into the oral cavity with the same clinical features and extensions.[8] In another study by Gaskin et al, MRI T1 weighted image showed predominantly high-signal mass with multiple ill-defined, mildly thickened septa and its corresponding contrast-enhanced axial T1-weighted image showed enhancement of these septa.[9] In an article by Mavrogenis et al, CT scan showed a lobulated, well-circumscribed mass with multiple septations and variable contrast enhancement.[10]

Liposarcoma is histologically identified with the presence of lipocytes of various shapes and sizes with nuclear atypia. Enzinger et al classified liposarcomas into well-differentiated, myxoid, pleomorphic and dedifferentiated types.[11] Well-differentiated liposarcoma can be sub-classified into 4 morphological subtypes as lipoma-like, sclerosing, inflammatory and spindle cell subtypes.[12] Lipoma like subtype is the most common, which consists of atypical cells and frequently lipoblasts. Sclerosing type is second most common and contains collagenous fibrous tissue with scattered adipocytes and atypical multinucleated stromal cells, while inflammatory which is rare type has chronic inflammatory cells with occasional lymphoid follicles scattered in a fibro collagenous stroma with sparse multinucleate atypical cells. Spindle cell subtype has histologic similarity to neural neoplasms with whorls or storiform growth pattern and can have areas of traditional lipoma like morphology.[2] In our present case; histopathological examination showed microscopically fat lobules within an encapsulated tumor, adjacent to which few areas of spindle cells with fibroblastic and myofibroblastic differentiation was seen. The tumor edges also showed a few keratinous fibres, foci of mucous glands with fibrous areas and few giant cells suggesting atypical lipoma. JG et al in his study mentioned that under microscopy, the lesion had components of mature adipocytes and lipoblasts with nuclear atypia.[7] Azeem et al in his case report revealed a well-encapsulated tumor consisting predominantly of myxoid stroma with few mature fat cells, lipoblasts and floret cells suggesting myxoid liposarcoma.[8]

In our case, complete excision of the tumor was done through transcervical approach. In a case report by Azeem et al, the excision of the tumor was done per orally, which differs from our case.[8] If the tumor is localized to the oropharynx and with limited extension, peroral excision of the tumor is possible. In a large tumor with greater extensions, transcervical is the preferred approach. The primary modality of treatment for well-differentiated liposarcoma is wide margin excision while high graded liposarcomas may benefit from adjuvant radiation. However, in the head and neck region, wide excision of the lesion is not possible in all cases due to the proximity of the vital structures. McCulloch et al reported that there was an 80% rate of local or distant recurrence in patients with incomplete surgical excision compared with a rate of only 17% when complete excision was accomplished.[3] The necessity of adjuvant

radiotherapy should be decided based on the site, extent and grade of the tumor.[13] In a study done by Eeles et al, adjuvant radiotherapy was found to reduce the local recurrence rate in head and neck sarcomas.[14] Vella et al reported a similar case of retropharyngeal liposarcoma of dedifferentiated type histologically wherein the patient was subjected to adjuvant rotational intensity modulated radiotherapy; volumetric modulated arc therapy after excision and had no recurrence later.[13] In our case, as complete excision was done and histopathology was well-differentiated liposarcoma, the patient was advised only regular follow up. After an extensive literature search, only one case treated by Gundelach et al underwent adjuvant radiotherapy for well-differentiated retropharyngeal liposarcoma after complete excision and the follow up revealed no evidence of recurrence.[15] So far, chemotherapy has not been found to be effective for the adequate treatment of soft-tissue liposarcomas. Doxorubicin and dacarbazine with or without cyclophosphamide have proved effective in the treatment of advanced myxoid liposarcomas.[16]

Various studies suggest the histological subtype to be the major prognostic factor for liposarcoma. Well-differentiated liposarcoma tends to recur locally, but rarely metastasizes and has a better prognosis among the other subtypes of liposarcoma.[17] The presence of tumor necrosis and increased age (>45 years) are also said to be associated with poorer prognosis.[18] The 5-year survival rates according to Golledge et al in his study on 76 head and neck liposarcomas were 100 % for well-differentiated, 73% for myxoid, 42% for pleomorphic and 0% for round cell type which has the poorest prognosis.[19]

CONCLUSION

Retropharyngeal liposarcoma is an extremely rare differential diagnosis of any retropharyngeal space swelling. Hence if any swelling in the retropharyngeal space is noted, the probability of soft tissue tumors must be ruled out by a CT/MRI scan before attempting incision and drainage or any other surgical procedure. Per-oral excision of the tumor is limited only to localized tumors, while the preferred approach is always trans-cervical. Postoperative adjuvant radiotherapy is limited to high-grade tumors, incompletely excised tumors and more radiosensitive liposarcoma subtypes such as myxoid sarcomas.

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