DISCUSSION:

Liposarcoma is a malignant neoplasm of adipocytic origin and considered to be the most common soft tissue sarcoma accounting for 17% to 30% of all soft tissue malignancies in adults. Its most common site of occurrence is thigh, followed by retroperitoneum and inguinal region [1, 2]. Liposarcomas of head and neck region are rare, comprising around 3% of all sarcomas at this location and are seen primarily in adults with a mean age of 57 years [3, 4]. It usually presents as a soft, slow growing, painless, ill-defined mass that appears yellowish in color and soft to firm in consistency. In the oral cavity, the lesion is even rarer, representing just 0.3% of all sarcomas, [5]. On gross examination, well-differentiated liposarcomas are usually well-circumscribed and tend to be uniformly yellow. The variable presence of fibrous tissue correlates with gray-white areas that can predominate in sclerosing and spindle cell subtype. The histologic hallmark of adipocytic well differentiated liposarcoma is the presence of mature adipocytic proliferation, exhibiting striking variation in cell size with at least focal nuclear atypia in fat cells or stromal spindle cells. Lipoblasts, if present, are considered to be diagnostic, however, they might be absent. Thus, Extensive sampling is critical because the most important diagnostic pointer can be very focal and therefore easily overlooked.

On immunohistochemistry, most adipocytic tumors express S100 protein which may play a minor role in highlighting lipoblast. MDM2 and CDK4 are becoming increasingly popular confirmatory markers because they are essentially never overexpressed in benign lipomas. MDM2 can cross react with macrophages, and multinucleated giant cells as most often seen in fat necrosis. However, these markers are usually negative in a typical spindle cell lipomatous tumor therefore they can be used to confirm the diagnosis. Variable staining for CD34, S100 and desmin has been reported in a typical spindle cell lipomatous tumor. Loss of nuclear expression of Rb gene is detected in approximately 60% of the cases.

The main differential diagnosis of well-differentiated liposarcoma is benign lipoma. The diagnostic clue favoring liposarcoma is varying size adipocytes and cytologic atypia. Lipoblasts are not necessary for diagnosis, may be difficult to find or may be even absent in WDLS. Another diagnostic pitfall is fat necrosis or atrophy which show varying size adipocytes. However absence of cytologic atypia favors atrophy/ necrosis over liposarcoma. Another important lesion that can be mistaken for true adipocytic neoplasm is massive localized lymphedema. This lesion affects mostly obese women and affect proximal medial aspect of extremities. Histologically it is composed of mature adipocytes demarcated by expanded connective tissue septa. This widened septa can simulate fibrous band of ALT/ WDLPS. However atypia is absent. Another differential is Lipomatous hemangiopericytoma, which is a variant of solitary fibrous tumor containing an adipocytic component and hemangiopericytoma like vascular pattern. CD34 and STAT6 positivity helps to differentiate it from WDLS.

Rarity of this tumor at this location with paucity of literature, evokes an interest in this case. This case highlights significance of histomorphology with immunohistochemistry to differentiate this neoplasm from its close mimics.