DUAL PATHOLOGY- RETROPHARYNGEAL LIPOSARCOMA AND PERIPANCREATIC DIFFUSE LARGE B CELL LYMPHOMA

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ABSTRACT

Liposarcomas in the head and neck region are rare and are extremely infrequent in the retropharyngeal space. Diffuse large B cell lymphoma (DLBCL) is the most common histological subtype of Non Hodgkin’s lymphoma, forty percent of which present with primary extranodal lesions. Gastrointestinal tract is the most common extranodal site for DLCBL. The following case report describes a dual pathology of retropharyngeal well differentiated liposarcoma and peripancreatic diffuse large B cell lymphoma in a 70 year old male patient.

Key words: Retropharyngeal liposarcoma, peripancreatic lymphoma, dual pathology.

INTRODUCTION:

Liposarcoma is one of the common sarcomas of adulthood and the head and neck region constitutes approximately three percent of all locations. Retropharyngeal liposarcomas are rare and only nine prior cases have been reported. There are five histopathological types, each with differing prognosis. The main treatment remains surgical wide excision1.

Diffuse large B cell lymphoma (DLBCL) is the most common histological variant of Non Hodgkin’s lymphoma (NHL) (around twenty five percent). It has male predominance and the incidence increases with age, the mean age of presentation being 64 yrs. Gastrointestinal tract is the most common site for primary extra nodal lymphoma2.

Occurrence of this dual pathology in a patient has not yet been reported in the English medical literature.

CASE REPORT

A seventy one year old male patient, a known diabetic and hypertensive, presented with complaints of progressive pain abdomen since 1 year, mass in the abdomen since 6 months and progressive dysphagia to solids since 3 months, more to solids than liquids. Pain abdomen was intermittent, predominantly in the epigastric region, aggravated on taking food.

He had been evaluated elsewhere and a flexible endoscopy had revealed a posterior pharyngeal wall swelling, displacing the larynx anteriorly. It had also revealed hiatus hernia, grade III to IV reflux esophagitis, florid duodenal ulcer with stigmata of recent active bleeding.

Examination of the neck revealed shift of the external laryngeal and tracheal framework to the right. Rigid telescope of larynx showed a smooth bulge in the left side of the posterior pharyngeal wall, the inferior extent of which could not be made out. Bilateral vocal cords were mobile.

Computed tomography with contrast revealed a...
well encapsulated mass, displacing the common carotid artery and the internal jugular vein posterolaterally (Fig.1). Superiorly it was extending from the level of C1 vertebra and inferiorly up to superior border of C6 vertebra. Posteriorly it was extending up to anterior border of vertebral body, laterally up to submandibular space.

He was planned for expandable direct laryngoscopy and biopsy under general anesthesia, but the procedure was abandoned as intubation could not be done. Computed tomography guided fine needle aspiration cytology was done instead.

He was then lost to follow up for 5 months. When he was subsequently reviewed, he had undergone direct laryngoscopy, biopsy and emergency tracheostomy at a local hospital. The biopsy had revealed no evidence of malignancy. However, the patient continued to have dysphagia and was readmitted for further evaluation. At this time, a repeat rigid telescope of the larynx revealed progression in size of the bulge arising from the left side of the posterior pharyngeal wall, which was touching the epiglottis and obscuring view of the larynx.

Under general anesthesia, using a transcervical approach, the mass was dissected and removed in toto (Fig.2).

Histopathological examination revealed lobules of mature adipocytes in varying stages of differentiation, some being spindle shaped and pleomorphic, with enlarged hyper chromatic nuclei, some with irregular nuclear membrane, moderate to scanty vacuolated and eosinophilic cytoplasm, few binucleate and bizarre forms, interspersed amongst large univacuolated adipocytes and few rhabdomyoblast-like large ovoid cells with abundant eosinophilic granular cytoplasm and sclerotic, focally myxoid stroma containing few
proliferating vascular channels, chronic mononuclear inflammatory cell infiltrate forming focal aggregates and mast cells, leading to a diagnosis of well differentiated liposarcoma (sclerosing type with focal myxoid areas) with foci of rhabdomyoblastic differentiation. Immunohistochemistry revealed the tumor cells with rhabdomyoblastic differentiation to be desmin positive (Fig. 3).

Abdominal examination revealed soft, non-distended abdomen, with tenderness in the epigastrium. A firm, non-tender, intraperitoneal mass around 10x15cm, was felt in right lumbar region with smooth surface. The mass was not moving with respiration. Divarication of recti was present.

Upper gastrointestinal endoscopy revealed extrinsic compression in the region of pylorus and first part of duodenum, with mucosal involvement. Multiple biopsies were taken from duodenum and pylorus.

Histopathological examination revealed section from peritoneal deposits showing malignant cells arranged in clusters, acinar pattern, small sheets and individually lying, characterised by large cells with abundant cytoplasm, few with vacuolation, pleomorphic irregular nuclei with prominent nucleoli, surrounded by fibro-collagenous stroma with focal myxoid changes, areas of calcification and thickened blood vessels, suggestive of Non-Hodgkin lymphoma - diffuse large B cell lymphoma (DLBL). Immunohistochemistry confirmed the diagnosis since the tumor cells were CD20 positive, CD3 negative (Fig. 4).
Fig. 4a Showing malignant lymphoid cells arranged in clusters, acinar pattern with pleomorphic nuclei with nucleoli and fibro-collagenous stroma. IHC : CD20 + ve, CD3 - ve.

He was planned for workup for Rituximab based chemotherapy.

The patient and his relatives were explained about the nature of the disease and further treatment. They were not willing for any further management.

He expired about 6 months later after several bouts of hematemesis.

DISCUSSION

Liposarcomas are the second most common soft tissue sarcomas of adult life after malignant fibrous histiocytosis with mean age of fifty, with slight male predilection. They account for around fifteen percent of all soft tissues neoplasms. Their incidence in the head and neck region is approximately 3 to 5.6 percent, and occurs more often in the larynx than in pharynx3,4.

Diffuse large B cell lymphoma (DLBCL) is the most common histological subtype of Non Hodgkin’s lymphoma (NHL) around thirty three percent. About forty percent of DLBCL with primary extranodal lesions, the gastrointestinal tract being the most frequent site, with stomach being most often involved2,5.

Liposarcomas usually present as painless enlarging masses. When they arise in the retropharyngeal space they are difficult to detect early. They could present with dysphagia, foreign body sensation in the throat if tumors are large. Examination findings include bilateral neck swelling and a reduced antero-posterior diameter of the pharynx, although these do not appear unless the tumor has reached a large size6. Our patient presented with progressive dysphagia to solid.

The gastrointestinal DLBCL typically present with a variety of symptoms including watery diarrhea and severe weight loss2. Our patient presented with pain and mass per abdomen and progressive dysphagia.

Liposarcoma is the malignant counterpart of lipoma and most frequently arises in the deep soft tissues of the retroperitoneum and the proximal extremities. The most recent World Health Organization classification of soft tissue tumors recognizes five categories of liposarcomas: well differentiated, myxoid, round cell, pleomorphic and dedifferentiated1. In our patient the liposarcoma was of a well differentiated type(sclerosing type with focal myxoid areas ) with foci of rhabdomyoblastic dedifferentiation6. Rhabdomyoblastic differentiation is very rare with only few cases being reported in literature.

For retropharyngeal liposarcomas, computed tomography and magnetic resonance imaging aid in establishing a diagnosis. Well differentiated liposarcomas are often diagnosed in CT and MRI, with a largely lipomatous mass (more than 75% of the mass) and the non lipomatous part in thick septae and focal nodules. Myxoid and pleomorphic variants are easier to diagnose by CT/MRI, as compared to well differentiated counterpart.

Imaging plays an important role in diagnosis and staging of pancreatic masses. This is particularly true for lymphomas, as treatment and prognosis are significantly different from pancreatic adenocarcinoma. Computed tomography is the commonly used imaging modality2.

Due to the deep location of retropharyngeal tumors, a core biopsy or incisional biopsy usually obtains a small tissue and hence the frequent misdiagnoses.
Ultrasonography guided or computed tomography guided fine needle aspiration cytology can help to distinguish pancreatic lymphoma and adenocarcinoma. Definite diagnosis of primary pancreatic lymphoma without a tissue diagnosis is difficult since the clinical signs are very similar to pancreatic ductal adenocarcinoma.

The principal management approach for liposarcoma is wide surgical excision, however, in the head and neck, the lesion is usually close to vital neurovascular structures, so the extent of surgical excision is restricted to avoid severe complications and the use of adjuvant radiotherapy is on the rise. Irrespective of the presenting symptoms, the mainstay of DLBCL treatment is chemotherapy with or without radiotherapy (CHOP regimen with or without Rituximab).

The major prognostic factor for liposarcoma is the histological subtype. Well differentiated liposarcoma is the most common subtype. It is locally recurring, but rarely metastasizing, and it has improved prognosis as compared to the other subtypes. Myxoid is similar to the well differentiated subtype, and are unlikely to metastasize and have a favorable prognosis. However, there are more locally aggressive forms, having high local recurrence rate and have worse prognoses. Late detection, misdiagnosis, deep anatomical sites and the associated difficulties of surgery add to worse prognosis.

**CONCLUSION**

A dual pathology of retropharyngeal liposarcoma and peripancreatic DLBCL has not been reported in the medical literature.

Rhabdomyoblastic differentiation of diffuse large B cell lymphoma is very rare with only few cases being reported in literature.

Transcervical approach was adequate for the retropharyngeal mass excision.

Due to short follow up period, the prognostic implication of the rhabdomyoblastic differentiation and wide surgical excision could not be assessed.

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**REFERENCES**


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