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Primary lesser sac myxoid liposarcoma: A case report

S. Navin Noushad^{a,*}, R. Rajaraman^b, Subbiah Shanmugam^c^a Surgical Oncology, Govt. Royapettah Hospital, Chennai, India^b Dept. of Surgical Oncology, Govt. Royapettah Hospital, Chennai, India^c Dept. of Surgical Oncology Govt. Royapettah Hospital, Chennai, India

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ABSTRACT

INTRODUCTION: Lesser sac pathological entities are uncommon. Most of these are tumors and are generally misdiagnosed as retroperitoneal lesions.**CASE REPORT:** A 62 year old male with past history of treated hypopharyngeal cancer presented with progressive abdominal distension. Physical examination revealed a midline intra abdominal mass in the epigastrium and umbilical region. Radiological investigations were suggestive of a retroperitoneal tumor, an image guided biopsy was reported as atypical lipoma. Surgical exploration confirmed a large multi lobulated tumor arising primarily from the lesser sac, post operative histopathological examination confirmed a myxoid liposarcoma.**DISCUSSION:** Primary lesser sac tumors are rare, a literature review of primary lesser sac tumors with particular reference to myxoid liposarcoma is presented.**CONCLUSION:** Primary lesser sac liposarcomas are rare neoplasms. The myxoid variant is unique for its peculiar biological behavior, in its sensitivity to chemotherapy and radiotherapy and for the presence of specific cytogenetic marker.© 2016 The Author(s). Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

1. Introduction

Lipomas represent 50% of all benign soft tissue tumors. Liposarcoma comprises of 15% of adult sarcomas and predominantly affect the extremities [1]. About 10–20% of sarcomas arise from the retroperitoneum and includes a heterogeneous list of histological types [2]. The most common site for abdominal liposarcoma is the retroperitoneum. Primary lesser sac sarcomas are rare and lesions are often misdiagnosed as originating from the retroperitoneum [3]. We report this case for the rare site of origin and unusual histology.

2. Case report

A 62 year old male presented with progressive distention of the abdomen. He had completed multimodal treatment with 60 Gy of external beam radiotherapy and 4 cycles of cisplatin and 5 FU chemotherapy for locally advanced hypopharyngeal cancer 11 months back. On examination he had a 20 × 10 cm midline intra abdominal mass occupying the epigastrium, umbilicus and upper hypogastrium. A USG abdomen revealed a large solid retroperitoneal mass displacing bowel loops and left kidney with bilateral

hydrocele. MRI/CT scan showed a large 30 × 23 × 13 cm lobulated mass in the retroperitoneum with predominant intermediate signals and multifocal areas of fat intensity on T1 images (Fig. 1). A core biopsy done before referral to our unit was reported as atypical lipoma. Further evaluation with CT chest and testicular tumor markers were normal. An upper GI endoscopy revealed a normal gastric mucosa with extrinsic compression along the greater curvature.

A provisional diagnosis for retroperitoneal atypical lipoma/liposarcoma was made and surgical exploration done by a midline laparotomy incision. A large well encapsulated, multi lobulated mass arising posterior to stomach but anterior to pancreas and displacing left kidney, spleen and bowels was encountered (Fig. 1). A diagnosis of lesser sac tumor was made and complete excision of the mass was done. Post operative histopathological examination confirmed liposarcoma–myxoid variant (Fig. 2). IHC studies showed the tumor to be S100 and vimentin positive (Fig. 3) but desmin negative. The patient had an uneventful post operative period and was discharged.

3. Discussion

Primary lesser sac pathological processes are rare. A dynamic list of infective, inflammatory and neoplastic lesions affecting the lesser sac has been reported in literature. Space-occupying lesions of the lesser sac include pancreatic pseudocysts or abscesses, lymphadenopathy along the lesser curvature of the stomach, and

* Corresponding author:

E-mail addresses: navinnoushad2004@gmail.com (S.N. Noushad), rajaanu@rediffmail.com (R. Rajaraman), subbiahshanmugam67@gmail.com (S. Shanmugam).

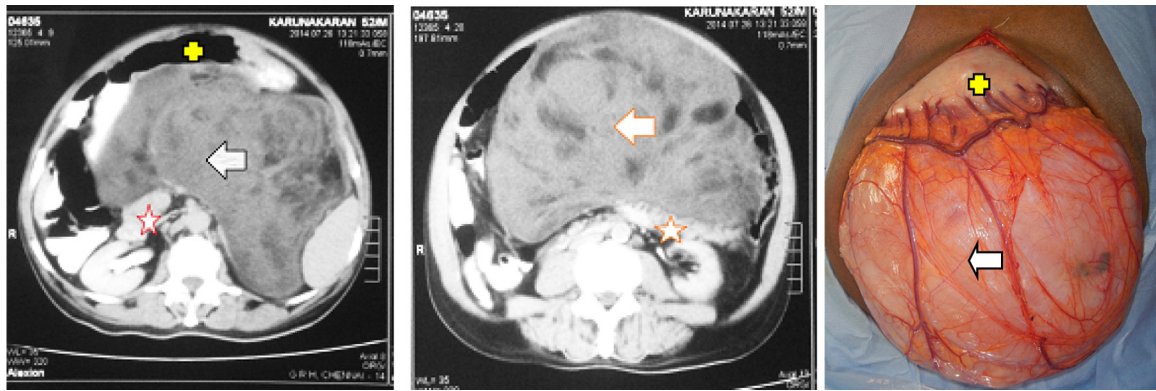


Fig. 1. CT scan & intra operative photograph showing tumor (arrow) posterior to stomach (cross) but anterior to pancreas (star), kidneys and vessels.

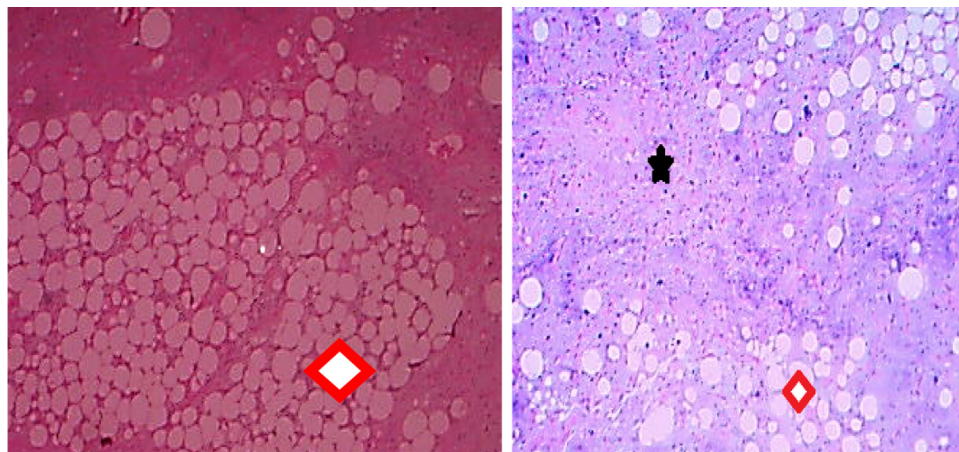


Fig. 2. Showing bimorphic pattern – lipid areas (red diamond) & myxoid regions (black star).

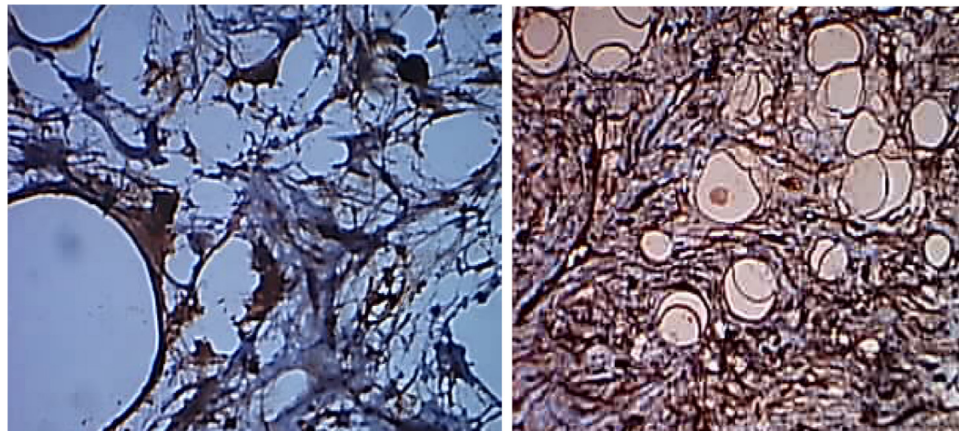


Fig. 3. IHC studies showing S-100 (left) & Vimentin (right) positivity.

neoplasms. Pathologic lymph node enlargement is commonly the result of metastatic spread from gastric or esophageal carcinoma or occasionally tuberculosis. Primary tumors of the lesser sac are rare and include benign tumors (lymphangioma, neurogenic tumor, teratoma) and malignant neoplasms (liposarcoma, GIST). Most tumors involving the lesser sac represent secondary spread from intrinsic tumors of the stomach, liver or pancreas [4].

Primary lesser sac liposarcoma is scarcely reported though retroperitoneal liposarcoma is a common tumor [3]. Liposarcomas are tumors of predominately adult men primarily affecting the

extremities followed by the retroperitoneum. Liposarcoma occurs in 3 main histomorphological forms each with unique biological behavior, a well-differentiated, myxoid/round cell and pleomorphic types. Some tumors have a combination of morphologic types and are classified as combined or mixed-type liposarcomas. The anatomical distribution of liposarcoma is related to the histologic type with well differentiated, myxoid/round-cell liposarcomas and pleomorphic variants having a striking predilection for the limbs, while dedifferentiated liposarcoma occurs predominantly in the retroperitoneum. The risk of distant metastasis and

survival are influenced by histological type with well differentiated, myxoid variants with a better outcome than pleomorphic and round cell types [1,2].

Myxoid liposarcoma accounts for approximately 40% of all liposarcomas and usually prefers deep extremity soft tissues (66%) but rarely may involve retroperitoneum or subcutaneous tissue. Histological features include paucicellular myxoid stroma, arborising capillary network (chicken wire vessels), a bimorphic cell population of signet ring lipoblast and uniform round cells representing primitive non lipogenic mesenchymal cells. Occasional microcyst enclosing eosinophil material may be present. Lesions with a greater than 5% of round cell population have 5 year survival rate of 50% and are deemed high grade while pure myxoid lesions and low grade lesions have a 90% 5 year survival [5].

Cytogenetically a specific translocation t(12;16) (q13-14;p11) is found in about 90% of myxoid liposarcomas, including tumors with a mixture of myxoid and round cell components. In one-third of the cases this translocation is the sole cytogenetic anomaly, the other secondary aberration, trisomy 8 is seen in 6–8% of the cases. An alternative unbalanced translocation t(12;22)(q13;q12) has been identified in about 5% of the tumors [6]. The molecular genetic consequences of the t(12;16) is the formation of a fusion transcript involving FUS in 16p11 and CHOP in 12q13. The role of the encoding chimeric protein in pathogenesis or tumor biology remains unknown [7]. This cytogenetic alteration is highly specific for myxoid liposarcoma and its detection by FISH/RT-PCR has been proposed to confirm diagnosis when routine histology is inconclusive [8].

Primary lesser sac tumors are often misdiagnosed clinically as arising from the retroperitoneum and radiographic imaging is the key to diagnosis. Imaging with USG is usually non diagnostic due to superimposed bowel gas. CT scan is the imaging modality of choice with MRI adding modest additional information. It permits detection of anatomical origin of the tumor, offers a cue about histology, assessment of operability and metastatic spread [9]. Specific signs like the beak sign, embedded organ sign and the Phantom organ sign may aid to differentiate from primary retroperitoneal lesions [9]. A CT imaging guided treatment policy obviating the requirement for open biopsy has been suggested [10].

Myxoid liposarcomas are unique in that they tend to metastasize to unusual soft tissue and bone locations with synchronous or metachronous deposits in retroperitoneum and axilla even in the absence of pulmonary metastasis. Surgery is the treatment of choice. Adverse prognostic factors for local recurrence and survival remain undefined [5]. These tumors in contrast to other soft tissue sarcomas exhibit extraordinary high response to radiotherapy and substantial sensitivity to trabectedin and ifosfamide chemotherapy [5]. In the series by wang et al. the risk of local recurrence after neoadjuvant or adjuvant radiotherapy was 8% at 5 years [11]. Guadagnolo et al. from the MD Anderson Cancer Center demonstrated a local recurrence of 3% in 127 patients treated with surgery and neoadjuvant and/or adjuvant radiotherapy [12]. The strongest argument for the effect of radiotherapy providing local control comes from the large Canadian multicenter study consisting of 415 cases, in which the addition of radiotherapy significantly prevented local relapse (18% vs. 4%) and also reduced by sevenfold the 5-year local recurrence rate in patients with positive margins [13]. Chemotherapy sensitivity of myxoid liposarcomas has been confirmed retrospectively by several authors (katz et al., patel et al. and Grosso et al.) and prospectively by Gronchi et al. in a phase II trial [14–17]. The response rate as assessed by choi criteria varied between 43 and 86% in these studies with none reporting progression while on chemotherapy [17]. However the role of adjuvant therapy for abdominal myxoid liposarcoma is unclear due to rarity of these tumors at these sites.

4. Conclusion

Primary lesser sac liposarcomas are rare neoplasms. The myxoid variant is unique for its peculiar biological behavior, in its sensitivity to chemotherapy and radiotherapy and for the presence of specific cytogenetic marker. Caution is advised regarding the use of adjuvant therapy for abdominal myxoid liposarcomas as all available data is derived from experience with extremity tumors.

Conflict of interest

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Ethical approval

None required.

Consent

Authors confirm informed consent was obtained from patient to publish this report.

Author contribution

All authors contributed equally to data collection, literature search, manuscript writing, review and final approval.

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