Quiz Case

A huge vulval cyst with iliac lymph node enlargement – A unique presentation of a rare tumor

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A 32-year-old female presented with abdominal pain, right labial mass, and right iliac lymph node enlargement for 4 years, to the gynecology department. On examination, a fluctuant, polypoid mass involving the right labium majus was noted and clinical diagnosis of vulval cyst was made. Her MRI findings revealed a well-defined 10.0 × 10.0 × 5.0 cm right labial lesion with heterogeneous signal intensity. It revealed hyperintense signal with areas of hypointensity on T2 and predominantly hypointense on T1, without any calcifications. Another altered signal intensity lesion measuring 5 × 5 cm was seen along the right iliac vessels, suggestive of the right iliac lymphadenopathy. USG-guided fine-needle aspiration (FNA) of the right iliac lymph node was performed and slides were stained with PAP, H&E, and MGG [Figure 1].

**Figure 1:** FNAC of the right iliac lymph node (Arrowhead-plexiform vasculature, Arrow- lipoblast), PAP, x100 (original).

**Q1 – WHAT IS YOUR INTERPRETATION?**

- a- Angiomyxoma
- b- Myxofibrosarcoma
- c- Myxoid dermatofibrosarcoma protuberans
- d- Myxoid liposarcoma.

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Question 1 answer – d-Myxoid liposarcoma.

BRIEF DISCUSSION

Smears made from FNA of the right iliac lymph node showed tissue fragments with myxoid matrix and plexiform vascular network [Figure 1: Arrowhead] surrounded by round to ovoid cells with mild atypia admixed with univacuolated to multivacuolated lipoblasts with scalloped nuclei [Figure 1: Arrow]. A diagnosis of low-grade myxoid liposarcoma (MLS) was made on FNAC. FNAC of the iliac lymph node was followed by removal of the right labial mass which was sent to pathology department for histopathological evaluation. The lesion was a well encapsulated, grayish-white to grayish-brown, lobular mass measuring 8.5 × 5.0 × 4.0 cm [Figure 2].

Gross evaluation revealed solid, multilobulated cut surface with grayish-white, fleshy and gelatinous areas with foci of hemorrhage [Figure 3]. Representative sections were submitted for microscopic evaluation. H&E stained sections from labial mass showed nodular lesion with hypocellular and hypercellular areas. Hypocellular areas showed uniform round to oval-shaped cells admixed with variable number of univacuolated to multivacuolated lipoblasts embedded in prominent myxoid stroma, rich in arborizing chicken wire capillary vasculature [Figure 4].

Extensive grossing was done and then slides were stained with H&E. Sections from labial mass showed nodular lesion with hypocellular and hypercellular areas. Hypocellular areas show uniform round to oval-shaped cells admixed with variable number of univacuolated to multivacuolated lipoblasts embedded in prominent myxoid stroma, rich in arborizing chicken wire capillary vasculature [Figure 4]. Hypercellular areas showed predominantly sheets of round to ovoid cells (approximately 25%) with high nuclear cytoplasmic ratio and absent to scant amount of cytoplasm, favoring a diagnosis of high-grade MLS [Figure 5].

ADDITIONAL QUIZ QUESTIONS

Q2 – Plexiform blood vessels are frequently seen in all except
  a. MLS
  b. Superficial angiomyxoma
  c. Lipoblastoma
  d. Myxofibrosarcoma.

Q3 – Unimultivacuolated cells with scalloping of nuclei are the feature of
  a. Foamy macrophage
  b. Pseudolipoblast
  c. Lipoblast
  d. Adipocyte.

Q4 – Most common translocation involving MLS is?
  a. t(12;16)(q13;p11)
  b. t(7;16)(q33;p11)
  c. t(17;22)(q21.3;q13.1)
  d. t(11;12)(q23;q15).
ANSWERS TO ADDITIONAL QUESTIONS

Q2 (d); Q3 (c); Q4 (a).

Q2 (d) – Plexiform vessels are thin branching vessels usually seen in MLS, superficial angiomyxoma, and lipoblastoma. Myxofibrosarcomas, meanwhile, show thick walled, curvilinear vessels with perivascular alignment of tumor cells.

Q3 (c) – Criteria for diagnostic lipoblast include – hyperchromatic, indented or sharply scalloped nucleus with lipid-rich droplets in cytoplasm, and an appropriate histological background. Multivacuolated cells distended with hyaluronic acid (pseudolipoblast) can be seen in myxofibrosarcoma.

Q4 (a) – The entire range of myxoid/round cell liposarcoma is genetically tied to recurrent rearrangement of DDIT3 that partners with FUS in >95% of cases with a resulting FUS-DDIT3 fusion [t(12;16) (q13;p11)] or partners with EWSR1 in the remaining cases with a resulting EWSR1-DDIT3 fusion [t(12;22) (q13;q12)]. Identification of either the FUS-DDIT3 or EWSR1-DDIT3 transcript is considered both highly sensitive and specific for myxoid/round cell liposarcoma, allowing its distinction from morphologically similar neoplasms.

BRIEF REVIEW OF TOPIC

Vulvar sarcomas account for only 1–3% of all vulvar malignancies and the most frequent primary vulvar sarcoma is leiomyosarcoma.1,2 MLS of vulva is very rare and only seven cases are reported in literature till date, to the best of our knowledge [Table 1].3–8 MLS is the second most common subtype of liposarcoma harboring translocation (12;16) (q13;p11) which creates a chimeric gene FUS-DDIT3 and it encompasses a spectrum of tumors defined by their degree of lipoblastic differentiation. It is divided into low-grade and high-grade MLS according to the WHO classification proposed in 2013. At one end of the spectrum is low-grade MLS associated with favorable prognosis and, on the other end, is, high-grade MLS, defined as having ≥5% round cell component, considered more aggressive tumor which tends to metastasize.

MLS of vulva can be mistaken clinically as benign because of their rare location and presentation, which can lead to delayed treatment.9,10 Like in our case, a clinical diagnosis of vulval cyst was made and patient presented with iliac lymph node metastasis which was reported as low-grade MLS. However, on histopathology of labial mass, we encountered the round cell component as well, thus rendering it as high-grade MLS. Sometimes in hypercellular variant, stroma is

Figure 5: Hypercellular areas showing predominantly sheets of round to ovoid cells. (H and E, ×100) (original).

Table 1: Clinical profile of seven cases of MLS of vulva (original).

| Study              | Age (years) | Site               | Duration | Clinical diagnosis         | Maximum size (cm) | Management                          | Outcome       |
|--------------------|-------------|--------------------|----------|----------------------------|-------------------|-------------------------------------|---------------|
| Brooks and LiVolsi[3] | 15          | Vulvar perineum, recurred on the left posterior medial thigh | 20 months | Soft-tissue sarcoma         | 18                | Wide excision; local recurrence as round cell/high-grade myxoid liposarcoma 20 months later treated by chemotherapy | DOD*          |
| Donnellan and Moodley[4] | 26          | Left labium majus et minor | 4 years  | Bartholin cyst Lipoma       | 10                | Local excision followed by reexcision | NED+ 9 m     |
| Wu and Tarn[5]       | 45          | Right labium majus | 72 months | Lipoma                      | 7                 | Local excision; reexcision of 6 cm recurrence 16 months later | NED, 28 m    |
| Schoolmeester et al.[6] | 34          | Left vulval mass   | 11 months | Unknown                     | 11.7              | Local excision                      | NED, 28 m    |
| Back et al.[7]       | 33          | Bilateral perineum | 4 months | Unknown                     | 20 and 15         | Wide excision with radiotherapy     | NED, 2 yr    |
| Kwak et al.[8]       | 37          | Bilateral vulval mass | 3 weeks | Unknown                     | 20 and 15         | Wide excision with radiotherapy     | NED, 44 m    |
| Present case         | 32          | Right labium majus | 4 years  | Cyst                        | 8.5               | Wide excision                       | NED, 6 m     |

*DOD: Died of disease, +NED: No evidence of disease
| Differential diagnosis | Age | Location | Gross | Growth pattern | Cellularity | Morphology | LB | Blood vessels | Background | Mitotic activity | IHC | Genetics |
|------------------------|-----|----------|-------|----------------|-------------|------------|----|--------------|------------|----------------|-----|----------|
| Angio myxoma           | Reproductive age (30 years) | Superficial and deep | Polypoidal, partly circumscribed cut surface is homogenously gelatinous | Infiltrative | Low | Spindle and stellate cells with small round hyperchromatic nuclei | -nt | Small thin walled to large hyalinized blood vessels | Myxoid with fine collagen fibrils | Rare | absent | CD34, Vimentin ER, PR | t(12;21)(q15;q21.1) |
| Botryoid RMS           | Children (<10 years) | Mucosa lined hollow organs | Polypoidal with clusters of small sessile or pedunculated nodules | Polypoidal | Mode-rate | Subepithelial condensation of tumor cells (Cambium layer) comprising of primitive small round cells, stellate cells and rhabdomyoblast | -nt | - | Myxoid | Low to moderate | Vimentin, MyoD1, Myogenin | Loss of heterozygosity chromosome 11p15.5 |
| Myxoid DFSP            | Young-middle-aged adults | Superficial | Multinodular cutaneous masses, gray-white cut surface with gelatinous areas | Diffuse infiltrative | Moderate | Uniform spindle cells with plump elongate nuclei arranged in storiform pattern | -nt | Prominent thin-walled vessels | Myxoid | Low to moderate | CD34 | t(17;22)(q21.3;q13.1) (COL1A1-PDGFB) |
| Myxoid leiomyosarcoma  | Middle to older | Deep soft tissue | Well-demarcated cut surface is fleshy white-gray mass with whorled app & foci of gelatinous changes | Ill-defined | Mode-rate | Elongated spindle cells with blunted-ended nuclei arranged in long dissecting fascicles | -nt | Not seen | Myxoid | Low | SMA, CALDESMON | Complex with genetic instability |
| MFS                    | Elderly (60–80 yrs) | Superficial and deep | Multiple gelatinous nodules (superficial), Single mass with infiltrative margin (deep), cut surface is variably gelatinous | Multinodular | Moderate | Plump, spindle or stellate cells having large atypical hyperchromatic nucleus | Pseudolipoblast | Curvilinear, elongated blood vessels with perivascular condensation of tumor cells | Myxoid | High | MSA, SMA | Complex karyotype |
| MLS                    | Young adults | Deep soft tissue | Well-circumscribed, multinodular cut surface is gelatinous to fleshy | Nodular | Mode-rate | Mixture of uniform round-oval cells and bland fusiform cells | +nt | Plexiform, branching | Myxoid | Rare | Vimentin S100 | t(12;16)(q13;p11) (FUS-DDIT3) |

LB: Lipoblast, RMS: Rhabdomyosarcoma, DFSP: Dermatofibrosarcoma protuberans, MFS: Myxofibrosarcoma, MLS: Myxoid liposarcoma, -nt: Absent, +nt: Present
less myxoid and capillary network is less prominent leading to erroneous diagnosis of round cell tumor on FNAC. Hence, there is a need for extensive sampling.

Histologically, MLS can be confused with other myxoid tumors more common in the vulva such as aggressive angiomyxomas, botryoid embryonal rhabdomyosarcoma, myxoid dermatofibrosarcoma protuberans, myxofibrosarcoma, and myxoid leiomyosarcoma [Table 2].

**SUMMARY**

Vulvar MLS is an extremely rare case reported in the literature. The present case marks the seventh reporting of vulval myxoid/round cell liposarcoma and the first one presenting with iliac lymph node metastasis. Both pathologists and clinicians should be aware of the occurrence of this entity in vulval region to ensure the correct diagnosis and appropriate management of the patient with this potentially curable neoplasm.

**COMPETING INTEREST STATEMENT BY ALL AUTHORS**

The authors declare that they have no competing interests.

**AUTHORSHIP STATEMENT BY ALL AUTHORS**

All authors certify that we have actively participated in the conception, design, defining intellectual content and preparation and editing of manuscript.

**ETHICS STATEMENT BY ALL AUTHORS**

This was a retrospective analysis of the data and all the procedures were done after obtaining informed consents from patients.

**LIST OF ABBREVIATIONS** (In alphabetic order)

- DFSP – Dermatofibrosarcoma protuberans
- FNA – Fine needle aspiration
- FNAC – Fine needle aspiration cytology
- LB – Lipoblast
- MLS – Myxoid liposarcoma
- MFS – Myxofibrosarcoma
- RMS – Rhabdomyosarcoma.

**EDITORIAL/PEER-REVIEW STATEMENT**

To ensure the integrity and highest quality of CytoJournal publications, the review process of this manuscript was conducted under a **double-blind model** (the authors are blinded for reviewers and vice versa) through automatic online system.

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