Myeloid Sarcoma Of Vulva: A Short Update

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Dear Editor,

Myeloid sarcoma comprises of a collection of cells of granulocytic series. It is usually considered as a tell-tale of poor prognosis. Involvement of sites like brain, eye, spinal cord, respiratory tract not only can have a devastating impact on immediate organ function but also can be life threatening. Involvement of rarer sites like mediastinum, pleura, female genital system can be a great masquerade and confusing for the treating clinicians. Here we did a short review on “vulvar myeloid sarcoma”—its presentation, demographic details, treatment and outcome.

There are wide differentials for the vulvar swelling or mass. Most cases fall in the category of inflammatory disorders, benign cysts or congenital malformations. Rarely, gynecologists do encounter uncommon causes of vulvar mass like myeloid sarcoma, dermatofibrosarcoma protuberans, malignant fibrous histiocytoma etc. [1]. Knowledge regarding these rare conditions while approaching a case of vulvar mass will broaden the horizons of differential diagnosis for a treating clinician.

We did a Medline search of following ‘MeSH’ terms like “Vulvar myeloid sarcoma”, “Vulva and AML” and “Vulvar involvement in AML”. In total, we found seven indexed cases and one non-indexed case [2]. We included reports written only in English literature and hence two cases written in other languages (one each in French and Chinese language) were excluded from review. We read

the remaining six cases extensively, details of which are summarized in Table 1.

The reviewed cohort included six patients with a median age of 33 years (range 16–73 years). Interestingly, 50 % cases were reported from the East Asia region (one each from Taiwan, China and Japan). Vulvar sarcoma was the initial presentation in four cases with remaining two presenting as a manifestation of relapse. Cases with Vulvar involvement at initial presentation had either isolated (case no. 4 and 5) or associated medullary (case no. 2 and 6) involvement [2–5]. Cases who relapsed at the vulva (case no. 1 and 3) had undergone HSCT previously for MDS and AML respectively [6]. Most patients had FAB-M5 subtype of AML (case no. 1, 2 and 3). Case reported by Ersahin et al. [3] also had synchronous Paget’s disease and infiltrating ductal adenocarcinoma of right breast. Case reported by Hu et al. [7] had extensive myeloid sarcoma involving bilateral peri-renal spaces, ureters and adnexae.

Patients were treated with either chemotherapy alone (case no. 1 and 2), chemotherapy plus radiotherapy (case no. 3) or chemotherapy plus HSCT (case no. 5). One patient was lost to follow up soon after the diagnosis (case no. 2). After therapy, disease of four patients (case no. 1, 3, 4 and 5) was in remission on follow up at the time of reporting individual cases by the respective authors [2, 4, 6, 7]. Two patients—case no. 1 and 6 died during follow up due to sepsis and disease relapse respectively [3, 7].

As per WHO, a myeloid sarcoma is a tumour mass consisting of myeloid blasts occurring at any extra medullary site. Interestingly, simple infiltration by myeloid blast cells are not considered as myeloid sarcoma unless the tissue architecture is effaced. Common sites of EM involvement are skin, soft tissue and lymph node. We have reported in recent past few rare sites of MS like mediastinum, pleura, spinal cord and orbit [8–11]. Much rarer is

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Table 1 Review of cases of vulvar myeloid sarcoma with the demographic details, treatment given and outcome

S.no.	Age	Author	Province of patient	Year	Time of vulvar involvement	Clinic of presentation	Systemic involvement	Type of AML	Molecular/cytogene-tic abnormalities	Treatment given	Outcome as per documentation in individual articles
1.	45	Hu et al. [7]	Taiwan	2015	Relapse (post 336 months of HSCT for MDS)	Dermatology	After 0.4 months of vulvar relapse	M5	t(1;7) (p22;q36) t(3;21) (q22;q26) loss of chromosome 16	Chemotherapy alone (ARA-C + idarubicin)	Associated with MS involving bilateral perirenal spaces, ureters and adnexae. Patient achieved CR with resolution of vulvar mass Died after 3 months of reinduction due to sepsis
2.	25	He et al. [5]	China	2013	Initial presentation	Gynaecology	Simultaneously	M5	45,X,-X t(8;21) (q22;q22) AML1/ETO	Lost to follow up	Lost to follow up
3.	41	Nazer et al. [6]	Saudi Arabia	2012	Isolated relapse (post 10 months of HSCT for AML)	Gynaecology	No systemic relapse	M5	Not applicable	Reinduction chemotherapy + involved field radiotherapy	Complete resolution of mass
4.	16	Policarpio-Nicolas et al. [4]	USA	2011	Isolated Initial presentation	NA	No bone marrow involvement	Not applicable	Not applicable	Chemotherapy alone	In complete remission
5.	21	Isonishi et al. [2]	Japan	2011	Isolated initial presentation	Gynaecology	No bone marrow involvement	Not applicable	Not applicable	Induction chemotherapy followed by allogeneic HSCT	1st medullary relapse → achieved 2nd CR with salvage chemotherapy and allo-HSCT → 2nd medullary relapse → achieved 3rd CR with salvage chemotherapy and allo-HSCT → 3rd medullary relapse → chemotherapy only → achieved 4th CR
6	73	Ersahin et al. [3]	USA	2007	Initial presentation	Gynaecology	Simultaneously	M2	Not available	Chemotherapy (ARA-C + idarubicin) and tamoxifen	Associated Paget's disease and infiltrating ductal carcinoma right breast. Patient died with 6 months due to disease relapse

the simultaneous involvement of two or more extra medullary sites [7, 8, 10–12]. Female genital tract is also one of the rare site of involvement in acute leukaemia. Case reported by Hu et al. exemplifies the importance of being extremely vigilant while following the previously treated cases of AML. The case had a fatal course with multiple sites of extra medullary relapse compromising organ functions. We recently reported a case of previously treated ALL who had extra medullary relapse after 4 years of treatment completion. Ovary was the site of extra medullary relapse for which she received ALL-REZ BFM 2000 protocol and involved field radiotherapy (24 Gray in 12 fractions). She underwent matched related allogeneic HSCT from elder sibling. At present she is in complete remission at day +60 of post transplantation [13].

Myeloid sarcoma can (1) be isolated (2) precede AML (3) coincide with AML (4) represent blast transformation of MPN's or (5) represent a relapse in a previously treated AML. Studies have shown that t (8; 21) and inversion (16) are the two most common cytogenetic abnormalities detected in cases of AML with granulocytic sarcomas [12]. Commonly, FAB subtype M4 and M5 are mostly associated with extra medullary tissue involvement. In most cases isolated myeloid sarcoma progresses to frank systemic disease with medullary involvement in a short time span of 9–15 months. Hence isolated myeloid sarcoma should also be tackled aggressively with anti-leukemic therapy followed by consolidation with allogeneic HSCT if the appropriate donor is available. Example of case 5 shows that in some cases remission might not be long lasting despite consolidation with HSCT [2].

This review suggests that reporting of such rare cases should be promoted in order to increase the general awareness of atypical yet relevant presentations of hematological malignancies. As depicted in Table 1, most of the times patients with vulvar lesions initially consult either dermatologists or gynecologists. It is of utmost importance not to misdiagnose such cases as benign diseases like sexually transmitted disease related lesions or Bartholin's cyst/abscess [5].

The main purpose of this review is to sensitize clinicians, especially dermatologists or gynecologists regarding this rare presentation. This will prevent us from chasing unnecessary investigations and thereby delaying diagnosis. Also, prompt diagnosis will minimize the disease related complications and hasten the treatment.

Compliance with Ethical Standards

Conflict of interest None.

Ethical Approval Conducted study did not involve any human/animal requiring approval for the same.

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