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Case Report

Primary Ewing's sarcoma of vulva: a rare entity and a review of literature

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ABSTRACT

Ewing sarcoma family of tumour (ESFT) is a group of bone and soft-tissue neoplasm which has variable neuroectodermal differentiation. In one end Ewing sarcoma (ES) is poorly differentiated, on the other end primitive neuroectodermal tumour (PNET) shows a clear evidence of neural differentiation. Extraosseous Ewing sarcoma (EES) commonly arises in soft tissues of trunk or extremities with a few reported rare sites include oro-naso-laryngo pharynx, neck, pleura, retroperitoneal space, mediastinal space and genital tract. Till now 30 cases of EES of vulva and vagina have been reported in the literature. We report a case of ES vulva in a 35-year-old patient. Her all routine markers were found to be normal, and magnetic resonance imaging (MRI) showed a vulval mass with high vascularity without infiltration to the surrounding structures. Ultrasonography (USG) guided biopsy(histopathology) revealed to be a small round cell tumour with necrosis and clusters of cells around blood vessels. Immunohistochemical staining demonstrated that the cells were positive for membranous CD99 and vimentin with nuclear positivity for NKX2.2, and negative for synaptophysin, chromogranin A, CD 56, Ki 67-30%, whereas all other IHC markers to rule out differentials of small round cell tumour were negative. Based on histopathological examination (HPE) and immunohistochemistry (IHC) panel diagnosis of ES was confirmed. After ruling out for the metastatic lesions,3 cycles of neo adjuvant chemotherapy with VAC were given and planned for surgical resection. After that she is on regular follow up with adjuvant chemotherapy. EES is a rare very rapid growing aggressive tumour requiring IHC, HPE and molecular genetics for exact timely diagnosis and multimodality treatment for better prognosis and survival.

Keywords: Ewing's, Round cell, EWS-FLI translocation

INTRODUCTION

Ewing sarcoma family of tumour (ESFT) is a category of bone and soft-tissue sarcoma which shows different percentage of neuroectodermal component. In one end Ewing sarcoma (ES) is poorly differentiated, whereas the counterpart, primitive neuroectodermal tumour (PNET) has clearcut neural differentiation. Both varieties of tumour demonstrate the cell surface markers of the gene products MIC2 and CD99, considered highly sensitive immunohistochemical (IHC) hallmark for ES. Prevalence of ES is second most among all (15%) sarcomas in paediatric age group. Virtually These malignancy group shows genetic translocation of the long arm of chromosome 11 and 22. The EWSR1 and FLI1 gene may cause overactivity of insulin-like growth factor 1 playing central role in tumorigenesis.[1]

This category of tumours are highly sensitive to chemotherapy. As per data, very few cases of extraosseous Ewing sarcoma (EES) have previously been documented in the medical research. In this report we present a case of primary Ewing sarcoma of vulva (extending to perineum).

CASE REPORT

A 35-year-old lady presented to OPD of Acharya Harihar Post Graduate Institute of Cancer (AHPGIC), Cuttack with a painful vulval mass for last 6 months. She underwent evaluation of the mass outside 5 months back. On examination a 10×8 cm size vulval swelling seen on right side. The skin overlying the mass was intact, no ulceration and no pigmentation was noted. On investigation, her magnetic resonance imaging (MRI) showed a mass on right side of vulva with strong vascularity without infiltrating surrounding structures displacing the vagina to the left. She was put up for ultrasonography (USG) guided biopsy from the mass. Histopathological report showed equal distribution of small round cells with scanty eosinophilic cytoplasm with indistinct cytoplasmic membrane, and round nuclei with hyperchromatia. The nucleoli were inconspicuous with finely stippled chromatin. Geographical pattern of necrosis and clumps of round cells around blood vessels were seen. Round cell tumour favouring neuroendocrine neoplasm of vulva was diagnosed. Immunohistochemical staining revealed positivity for membranous CD99 and vimentin with nuclear positivity for NKX2.2, and negative for synaptophysin, chromogranin A, CD 56, Ki 67-30%. Thus, histopathological examination (HPE) and immunohistochemistry (IHC) together were compatible with the diagnosis of ES/PNET (Figure 3). Positron emission tomography computed tomography (PET CT) showed a metabolically active soft tissue mass in right vulva of 3.5×7×10 cm with SUV max 10.1, pushing vagina to the left without any local or distant metastasis. The patient was started on neoadjuvant chemotherapy with vincristine, adriamycin, cyclophosphamide/ifosfamide and etoposide (VAC/IE). After finishing her 3rd cycle (VAC) she was referred to our institute for better management. On examination at OPD we found a firm to hard tender smooth mass of approximately 8×8 cm size in right vulva displacing the vagina to the left without any induration or skin ulceration (Figure 1). The mass was not fixed to the underlying structures. Rests all gynaecological and systemic examination was normal. No palpable regional lymph node was found. MRI report showed lobulated well defined heterogeneously enhancing mass lesion of size 6.7×5.5×7.1 cm seen at right vulva abutting and indenting vaginal wall and external sphincter without sign of infiltration. No extension to true pelvis was seen (Figure 1).

Surgery was planned after discussion with institutional tumour board.

Intra operative findings

A 6 cm longitudinal incision given over the right vulva, a firm solid whitish lobulated (8×6 cm) mass was excised and the dead space was obliterated with proper haemostasis, following which skin margin was closed (Figure 2).

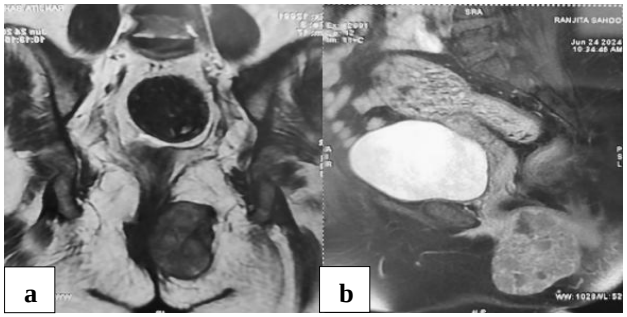

Figure 1 (a and b): MRI findings of the right vulval mass.

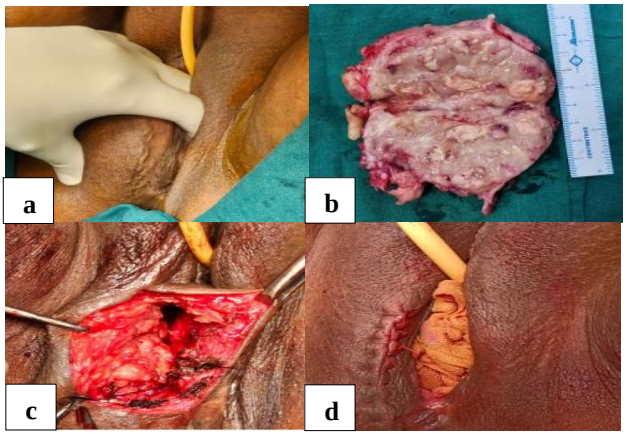

Figure 2: Pre-operative intraoperative post-operative findings and cut section of the mass (a) pre-op mass, (b) cut section, (c) intra op, and (d) after primary repair.

The final histopathology report showed grade 2 ES with tumour differentiation score 3, mitotic rate score 1, tumour necrosis score 1, LVSI -not identified, all margins were negative (Figure 3). Post-operative period was uneventful and she got discharged on seventh day. Now she is on regular follow up with adjuvant chemotherapy. On cytogenetics study EWSR1/FLI1 gene fusion was seen positive.

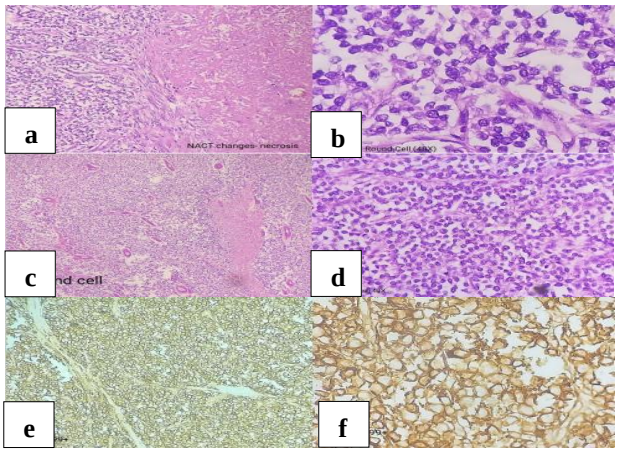

Figure 3 (a-f): Cluster of round cells with necrosis and IHC showing CD99 positive.

DISCUSSION

All members of the ES family of tumors are treated following the same line of management as sarcoma. Literature review found 34 additional cases of primary vulvar ES; fifteen from the Asian continent, nine from North America, seven from European, two African, and one Australian continent.[2] Median age at diagnosis was 21.5 years with a range from 3 to 65 years, with our case representing at reproductive age group. Maximum no presented with a painless mass with median size 5 cm (range 1–20 cm). Thirty-two cases were positive for CD99 staining, genetic rearrangement was confirmed by FISH (30%) or by RT-PCR (15%). Almost 30% of patients were seen at an advanced stage at diagnosis. Pelvic lymph nodes (15%) and lungs (15%) were M.C metastatic sites. 82% were treated with surgical resection and chemotherapy (76%), with lesser receiving no radiation. After a mean follow-up of 17 months, 44% of patients had local and distant site recurrence. 53% of patients were doing well. The prognosis of EES is less favourable compared with the skeletal subtype, although factors affecting prognosis seem to be similar in both subtypes. Notably, the 5-year overall survival rate is superior for localized EES compared with localized skeletal ES.[3] In our case, the swelling was tender with fast growth velocity, like most of the cases. USG-guided biopsy and IHC proved it to be a neuroendocrine tumour of the vulva. A positive immunostaining of CD99/MIC2 and an intranuclear FLI1 suggest ES/PNET. All other markers rule out a rhabdomyosarcoma, Merkel cell carcinoma and small cell carcinoma. [t(11;22) (q24;q12) (EWS-FLI1 gene rearrangement)] is seen in this case, which is a specific genetic marker for it. EES is more aggressive with poor outcomes than its bony counterpart, with a five-year disease-free survival of around 75% for skeletal ES, while 38% for the extra-skeletal counterpart. Since the present case had no distant metastasis and the mass was well circumscribed, surgical resection could be done successfully after three cycles of NACT without any residual disease. She completed adjuvant chemotherapy and has been doing well for the last 10 months. Current practice for management of ES includes resection of the tumour followed by adjuvant chemotherapy, whenever possible. The role of radiotherapy is not well established yet for the reported cases.

CONCLUSION

EES is a very rare, rapid growing neoplasm of an aggressive pattern mainly seen in young individuals. Immunohistochemistry and molecular study, along with histopathological study, is needed for prompt diagnosis of Ewing's Sarcoma. Early diagnosis, vigorous treatment and stringent follow-up is of utmost necessity for better management and overall survival. Some promising markers like CA-125 can play a pivotal role in prognosis and follow-up, which can be substantiated with more studies.

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REFERENCES

1. Xu M, Liu Y, Zeng S, Wang H, Weng G, Li F. Primary vaginal Ewing sarcoma with uterine fibroid: A case report. Medicine (Baltimore). 2020;99(27):e20859.
2. Cathcart AM, Babb P, Davis J, Emerson J. 30 Primary vulvar Ewing sarcoma: a case report and systematic literature review. Gynecol Oncol Rep. 2022;44(2):S14-5.
3. Abboud A, Masrouha K, Saliba M, Haidar R, Saab R, Khoury N, et al. Extraskeletal Ewing sarcoma: Diagnosis, management and prognosis. Oncol Lett. 2021;21(5):354.

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