Abstract:
Vulvar melanoma is a very rare condition accounting for less than 1% of all gynecological malignancies with usual presentation at late sixties. There are little prospective data and no randomized studies to guide management due to the rarity of the disease. Overall the prognosis of vulvar melanoma is poor, five-year survivals is 20 for patients presenting with regional lymph node involvement. We present a case of 28 years old young women with malignant melanoma of vulvawho underwent radical vulvectomy with partial vaginectomy and urethrectomy and bilateral inguinal block dissection. She expired after 2 years following recurrence.

Keyword: melanoma, vulvectomy, vaginectomy, urethrectomy

CASE REPORT:
A 28 year old woman P3L3/ Married since – 10 yrs presented with the c/o Growth protruding per vulva for past 2 months which was not associated with pain/bleeding, gradual in onset and progressing in nature. History of burning mictuiriton for 1 month, History of excessive white discharge per vaginum for 1 month – foul smelling and associated with itching and History of loss of appetite and weight was present. There was no history of menstrual disturbances. On examination, the general condition of the patient was fair. Local Examination - pigmented nodule of size 2x2 cm was present in right labia. Another nodule of size 2x1 cm in the left labia with associated leukoplakia vulva seen (Fig 1).

Speculum examination - cervix appeared healthy. Per vaginal examination – lower 1/3rd of vagina was also encroached.
Cervix pointing downwards; uterus- antverted, bulky, mobile, fornices free.
(fig 1 vulvar growth)

INVESTIGATIONS:

Biopsy from the growth was suggestive of malignant melanoma. USG abdomen done showed normal uterus and ovaries. No evidence of ascites/pleural effusion. CT (Abdomen and pelvis) – Vagina appears thickened. No significant lymphadenopathy. MANAGEMENT: Surgical oncologist opinion obtained and she was posted for radical vulvectomy. On 24.9.10 Radical vulvectomy with partial vaginectomy and urethrectomy with left ilioinguinal block dissection and right inguinal block dissection done since the growth has infiltrated lower 1/3 of vagina and urethra. Under Spinal anaesthesia by bilateral lazy ‘S’ incision over ilioinguinal region. Right inguinal block dissection done. Left side one node was enlarged clinically. Left inguinal block dissection done. Left iliac block dissection done through extraperitoneal approach. Radical vulvectomy proceeded. Partial vaginectomy done. Urethrectomy done. Neovagina created. Suprapubic catheterisation done (fig 2).
(fig 2 suprapubic catheterisation)

Patient developed lymphoedema (fig 3) of right lower limb on 14th postoperative day which was managed conservatively.
(fig 3 postoperative lymphoedema)

HISTOPATHOLOGY REPORT:

MICROSCOPY: (fig 4)
Section studied show stratified squamous epithelium and underlying tumour tissue. The tumour cells are arranged in sheets and strands and they have round to oval pleomorphic nuclei with scanty cytoplasm. Areas of pigmentation and tumour giant cells are seen. The intervening stroma shows dense lymphocytic infiltrate, areas of haemorrhage and necrosis. The tumour is seen infiltrating the surgical margin.

Right inguinal nodes: 8 lymphnodes showed metastatic deposits.
Left inguinal nodes: 8 lymphnodes showed metastatic deposits. Left obturator node: 2 lymphnodes showed metastatic deposit. Left iliac nodes: 5 nodes showed metastatic deposits.

Impression: Malignant melanoma (According to FIGO staging, she is a case of malignant melanoma of vulva Stage IV).
(fig 4 slide showing tumour cells arranged in sheets and strands with areas of pigmentation)
FOLLOW UP: Patient advised to review once in every month for SPC catheter change. Patient developed local recurrence and admitted for further management. Wide local excision with flap reconstruction was done. Inspite of these treatments patient expired on 13.8.12.

DISCUSSION:
Vulvar melanoma is a very rare condition accounting for less than 1% of all gynaecological malignancies with usual presentation at late sixties. Lesion that originates from melanocytes of skin. Common symptoms at presentation are presence of a mass and/or bleeding, pruritus and pain. The commonest histological subtypes are mucosal lentiginous melanoma or nodular melanoma. The regional lymph nodes are involved in up to one third of patients at presentation, and up to 25% of patients present with distant disease.

Up to one third of vulvar melanomas arise on the labia majora and are characteristically flat, pigmented lesions, while more centrally-based lesions, which frequently involve the labia minora and clitoris, are characteristically nodular, and up to one third may be amelanotic. Both the Clark and the Breslow systems are found to correlate with prognosis in patients with cutaneous melanoma.

CLARKES LEVEL OF INVASION
Level I – all the tumour cells are above the basement membrane
Level II – Extension into the papillary dermis
Level III – interface between the papillary and reticular dermis
Level IV – tumour cells reaching the reticular dermis
Level V – tumour cells invade the subcutaneous fat

BRESLOW STAGING (by measuring the thickness of the lesion at the centre of it)
Stage I – 0.75 mm or less
Stage II – 0.76 to 1.50 mm
Stage III – 1.51 to 3 mm
Stage IV – > 3.00 mm

TREATMENT: Surgery remains the mainstay of treatment for vulval melanoma. There are little prospective data and no randomised studies to guide management due to the rarity of the disease. Superficial lesions, particularly those in a favourable position, may be treated effectively by wide local excision. Unfortunately central lesions involving clitoris and urethra, which are often thicker, may require extensive / exenterative procedures to obtain complete excision. For lesions that are 1 to 4 mm thick, vulvectomy with a 2-cm margin is recommended and a 1-cm margin if lesion thickness is less than 1 mm. The incidence of occult inguinal lymphnodes is less than 5 percent for thin melanomas (<1 mm) and greater than 70 percent for thick lesions (>4 mm). Hence sentinel node biopsy, may be reasonable for patients who have tumour thickness between 1 and 4 mm. Patients with lesions >4 mm thick should undergo an elective lymphadenectomy. recent trials have suggested that adjuvant therapy with high-dose adjuvant interferon-alpha may be of benefit in preventing recurrence in certain patients with cutaneous melanoma involving other body surfaces.

However in case of melanoma of vulva, no trials have yet evaluated the benefit of adjuvant therapy in these women. The role of adjuvant radiotherapy is unknown but maybe considered where resection margins are less than optimal. Vulvar melanomas carry a poor prognosis and show a tendency to recur locally and develop distant metastases through hematogenous dissemination to lungs, liver or brain. Women diagnosed with vulvar melanoma have a median survival of 61 months.
PROGNOSTIC FACTORS: Includes tumor thickness, ulceration and nodal status. In several reports amelanosis and age are also related to outcome.
Overall the prognosis of vulvar melanoma is poor, with 50% of patients surviving five years. Five-year survivals may be as high as 70% for patients with thin lesions (<1mm) but less than 20% for patients presenting with regional lymph node involvement.

REFERENCES: