Case Report

Vulvar melanoma with multisystemic metastases: A rare case presentation

Shailvi Singhal¹, K S Sunil Reddy¹, Kokila Kathirvel¹, Lokesh Meena¹, Abhishek Kumar Yadav¹, Pankaj Sharma²

From ¹Junior Resident, ²Associate Professor, Department of Radiology, All India Institute of Medical Sciences, Rishikesh, Uttarakhand, India

ABSTRACT

Malignant melanoma is the second most common vulvar malignancy which occurs in elderly females. In advanced stages of the disease, distant metastases can occur to any organ; hence, comprehensive radiological evaluation is necessary. We report a case of a 45-year-old female who presented with progressively increasing growth over the vulva for 6 months. On detailed clinical, pathological, and radiological workup, multiple metastases were found in the brain, liver, pancreas, peritoneum, lungs, and pleura. The final diagnosis of vulvar melanoma with multisystemic metastases was made after the histopathological examination.

Keywords: Malignancy, Melanoma, Metastases, Vulvar melanoma

elanoma is the neoplasm of the melanocytes which originate from the neural crest cells [1]. It most commonly occurs in cutaneous tissue [2] but can also occur in orbit and mucosa. Visceral melanoma is a rare occurrence. Melanoma is also the 19th most common cancer worldwide with an incidence of 2.8–3.1/100,000 population [3]. The most common vulvar malignancy is squamous cell carcinoma. Malignant melanoma is the second most common vulvar malignancy [4]. The most commonly affected site is labia majora and clitoris. Vulvar melanoma is an aggressive tumor with a high propensity for locoregional spread and distant metastases. It usually affects females in their sixth to the eighth decade and presents with growth at the vulva or pigmented macule [1]. They may be pruritic and the overall prognosis of vulvar melanoma is poor. Here, we report a case of a 45-year-old female who presented with vulvar growth with multiple metastases.

CASE REPORT

A 45-year-old female, resident of Uttarakhand presented to the outpatient department with complaints of growth over the vulva for 6 months which was progressively increasing in size. There were no associated complaints of discharge or pruritus. Her menstrual cycles are regular without any dysmenorrhea. She has three living children, born by normal vaginal delivery. The past child's birth was 21 years back. She had no other comorbidity or addiction.

On examination, irregular black-colored growth of size 6×3 cm was seen at the vulva extending over the whole of

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the right side and infiltrating into the vagina. On per vaginal examination, the rectovaginal septum was immobile suggestive of infiltration. Her blood workup revealed microcytic normochromic anemia with hemoglobin of 8 g/dl, total leucocyte count of 12800 cells/mm³, and erythrocyte sedimentation rate of 18. The differential leukocyte count was normal.

Biopsy was taken from the growth and the histopathological examination showed fine granular intracytoplasmic melanin pigment in metastatic malignant vulvar melanoma (Fig. 1). Fineneedle aspiration cytology from the inguinal nodes also suggests metastasis from melanoma. For further workup, contrast-enhanced computed tomography (CECT) chest and abdomen with contrastenhanced magnetic resonance imaging (CE MRI) Brain were done for the patient. CECT showed a large lobulated lesion involving the vulva and vagina with locoregional lymphadenopathy with hepatic, pulmonary, peritoneal, pancreatic, and pleural metastasis (Figs. 2). CE MRI brain showed multiple T1 hyperintense lesions involving bilateral cerebral and right cerebellar hemispheres (Fig. 3). The lesions were iso to hypointense on T2WI with perilesional edema. On diffusion-weighted imaging, the restriction was seen with blooming artifact on GRE images. Lesions were showing enhancement after post-gadolinium enhancement.

DISCUSSION

Vulvar melanoma is the second most common malignancy of the vulva after squamous cell carcinoma, which is the most common vulvar malignancy. They usually occur in the age group of the sixth to the eighth decade. Various predisposing factors

Correspondence to: Shailvi Singhal, Room Number - 806, Building Number 83, All India Institute of Medical Sciences, Rishikesh - 249 201, Uttarakhand, India. E-mail: shailvi15@gmail.com

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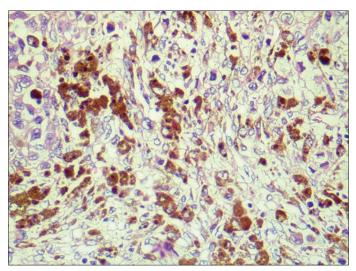


Figure 1: Histopathological examination showing fine granular intracytoplasmic melanin pigment in metastaic malignant vulvar melanoma

are human papillomavirus infection, previous irradiation, any ulceration, diabetes mellitus, and an immunocompromised state [4]. The patient is either asymptomatic in the early stage of the disease or may present with pigmented macule.

The incidence of melanoma is increasing in the world which is attributed to an increase in exposure to ultraviolet radiation. The primary site of melanoma most commonly is the skin, other sites can be the eye or mucosa. Visceral melanomas are rare. Melanoma cells may contain melanin or may be amelanotic. The presence of melanin gives the tumor a high signal on T1 weighted MR images, which is very uncommon in other tumors [5]. Melanoma most commonly metastasizes to lymph nodes through the lymphatic spread [2]. However, hematogenous metastasis occurs in advanced tumors to the liver, brain, lung, and soft tissues.

The staging of the tumor depends upon the depth of invasion. Breslow measured the vertical layer from the top of the epidermal layer to the bottom of the invasion. The major determinant of the prognosis of the patient is the thickness involved. If the tumor is ulcerated, thickness is measured from the base of the ulcer to the bottom layer. Clark et al. defined the depth of the invasion based on histological invasion [1].

Imaging modalities, such as ultrasonography (USG), computed tomography (CT), or positron emission tomography (PET-CT), have a very limited role in the early stage of the disease. USG of the regional lymph nodes could be done before surgery to look for the local spread of disease. However, in the advanced stages of the disease, imaging is necessary to look for distant metastases. Imaging involves MRI brain with CECT chest, abdomen or pelvis or PET-CT. PET-CT has the benefit of whole-body coverage; however, spatial resolution is poor. MRI brain has a very high sensitivity for the detection of brain metastasis [2]. Brain metastasis occurs late in the course of the disease and has a poor prognosis as compared to other visceral metastasis. Median survival after brain metastasis is just 3–4 months [1]. Brain metastasis occurs at the grey-white matter junction and is usually multiple. They exhibit hyperintense

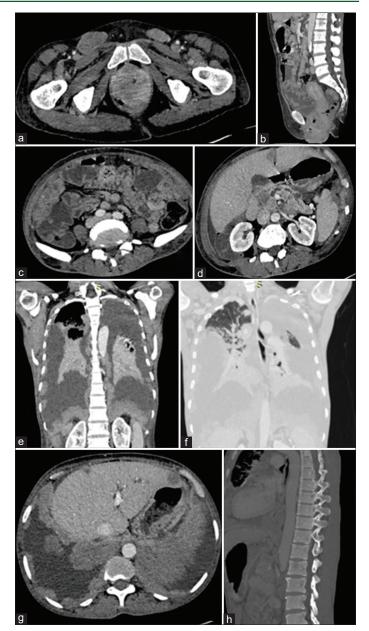


Figure 2: Axial and sagittal contrast-enhanced computed tomography abdomen and pelvis images show large heterogeneously enhancing mass involving vulva and vagina (a and b), nodular enhancing omental deposits (c), multiple pancreatic metastatic lesions compressing main pancreatic duct (d), Multiple metastatic nodular deposits in lung and pleural cavity with bilateral pleural effusion (e-g), heterogeneously enhancing liver metastasis (g), multiple lytic lesions in T9 and T10 vertebral body (h)

signals on T1WI. However, amelanotic melanoma exhibits iso to hypointense signal on T1WI.

Intrathoracic metastasis is common second only to lymph nodal metastasis. Pulmonary metastases from melanoma can be solitary; however, usually, they are multiple and round [5]. The liver is the most common site of visceral metastases in patients with melanoma. CECT abdomen is the modality of choice for the detection of visceral metastases.

Wide local excision of the primary tumor is the mainstay of the treatment for early-stage disease [6]. Radiation therapy can be used for the treatment of the patient with the locoregional disease in whom surgery is contraindicated. For brain metastases,

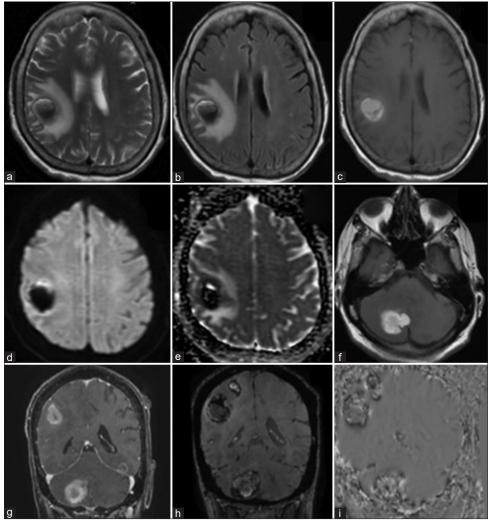


Figure 3: Multiple well defined (at least 10) rounded lesions are seen in the right frontal, parietal, left temporal lobes and right cerebellar hemisphere (a-i). They are showing heterogeneously hyperintense signal on T1/T2/FLAIR images (a-c and f) with few hypointense areas and surrounding perilesional edema. On diffusion-weighted imaging peripheral patchy area of diffusion restriction is seen showing low value on corresponding ADC maps (d and e). On susceptibility weighted imaging blooming is seen with mixed signal on filter phase images (h and i). On post-contrast images (g) few of the lesions are showing heterogeneous post-contrast enhancement

stereotactic radiosurgery can be performed. Ul Ain and Rao reported a case of 70-year-old female with vulvar melanoma with metastatic lymphadenopathy [7]. Li *et al.* reported a case of 42 years old female who presented with vulvar itching and pain and was diagnosed to have vulvar melanoma. The patient was managed with wide local excision, however, 16 months postoperatively, the patient developed metastases to the lungs, liver, breast, and peritoneum [8].

CONCLUSION

Vulvar melanoma is the second most common vulvar malignancy. It is essential to know the imaging features of melanoma. As melanoma can spread to any organ, imaging plays a crucial role in the search for metastasis in the advanced stages of the disease.

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