An Unusual Case of Primary Malignant Melanoma of Uterine Cervix

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ABSTRACT
Primary malignant melanoma of uterine cervix is a rare and aggressive neoplasm. Approximately 3%-7% of malignant melanomas in women develop within genital tract. Majority occurs in vulva or vagina, but cervix is a rare site. Age range for cervical melanoma is from 19 to 83 years with peak incidence between 60 to 70 years. Malignant melanoma presents with vaginal bleeding or discharge and appears as exophytic, polypoid, pigmented or colorless cervical mass. Diagnosis is by histopathology but should be confirmed by immune-histochemical staining with S100 protein and HMB45. It is also useful in distinguishing amelanotic melanoma from anaplastic carcinoma, high grade lymphoma and sarcoma. Primary melanoma of cervix with prominent spindle cell component should be distinguished from leiomyosarcoma and many benign melanotic lesions. Primary cervical melanoma must be differentiated from secondary metastasis of melanoma from other sites in the body. Prognosis of primary cervical melanoma is generally poor, because diagnosis is usually made at an advanced stage and tumor is highly aggressive and shows local recurrence and widespread metastases. There is no consensus on optimal management of primary malignant melanoma of cervix, because of its rarity. Cervical melanoma is incurable in totality even with the currently available therapies and hence needs to be diagnosed early. Recently the morphological features of primary cervical melanoma on pap smear have been described, raising the hope of early diagnosis. We report for its rarity a case of malignant melanoma of the cervix in a 60 year old female who presented with white discharge per vagina.

Keywords: Malignant melanoma, primary, uterine cervix

INTRODUCTION
In 1959, Cid reported the presence of melanocytes in the cervical epithelium of 3.5% of women. Since then, about 78 cases of primary malignant melanoma of the uterine cervix have been described in the literature. Primary malignant melanoma of the cervix constitutes a rare disease (neoplasm) representing less than 2% of cases of malignant melanoma of the genital tract. Often due to its amelanotic presentation the diagnosis may be missed or delayed. Most of the patients present in advanced stage of the disease and respond poorly to therapy. Diagnosis is confirmed by immune-histochemical method and by exclusion of primary melanoma at other sites. We report the clinical and histological feature of primary malignant melanoma of the cervix in a 60 year old female for its rarity.

CASE REPORT
History: A 60 year old post menopausal woman presented to the gynaecology department with a history of white discharge per vagina since 2 months. Examination: On per speculum examination a 4*5 centimeter bluish black colored mass was seen arising from the anterior lip of the cervix. Clinical diagnosis: Endometriosis or endometrial polyp. Investigations: Punch biopsy of the cervix was done and the specimen was submitted for histopathological examination which revealed a diffusely infiltrative malignant neoplasm composed of highly pleomorphic, round, polygonal to spindle shaped cells with atypical large irregular nuclei and prominent nucleoli. Intra and extra cellular fine brown granules of pigment which were Masson Fontana positive were present. Junctional activity was seen in the epithelium. Immuno-histochemical staining for HMB45 was also positive. The final diagnosis was pigmented malignant melanoma of the cervix. An extensive search for other melanotic lesions in the skin, uveal tract (ophthalmoscopy) and other mucosal sites was negative. Abdominal ultrasound and chest radiograph were normal. Considering the absence
of malignant melanoma of any other site and presence of junctional activity, a final diagnosis of primary malignant melanoma of cervix was made. The patient was referred to regional cancer institute for further management.

**DISCUSSION**

Malignant melanomas are generally found in areas of skin exposed to the sun but may also be present in non-exposed sites, such as genital tract and oesophagus, among other sites. Primary malignant melanoma of the cervix is very rare. A cervical melanoma may be either melanotic or amelanotic. Diagnosis of amelanotic melanoma may be missed due to the absence of pigment and thereby needs caution. The present case exhibited classic features of melanotic melanoma and hence posed no diagnostic dilemma.

Patients with malignant melanoma of the cervix may range between 19 to 83 years, although the majority of them have occurred between 60 to 70 years. In most cases, vaginal bleeding or discharge is the usual presenting complaint. Some patients may remain asymptomatic. Physical examination usually reveals a polypoidal exophytic mass which may be red, brown, grey, black or blue in color or colorless, in amelanotic melanoma, which constitute up to 55% of cases in the cervix. Recently the morphological features of primary cervical malignant melanoma in pap smear have been reported as bizarre and abnormal cells containing pigment, raising the hope for an early diagnosis.

Malignant melanoma in the uterine cervix which may be melanotic or amelanotic is composed of cells with varying degree of pleomorphic and prominent eosinophilic nucleoli. In the melanocytic type, dark brown intracellular pigments which stain positive with Masson’s Fontana stain are seen. In the absence of pigment, the differential diagnosis includes anaplastic carcinoma, poorly differentiated squamous cell carcinoma, adenocarcinoma, rhabdomyosarcoma, stromal sarcoma and high grade lymphoma.

Melanoma cells are negative for epithelial markers—cytokeratin and EMA, and smooth muscle markers. They are positive for S100 protein (more sensitive) and HMB45 (more specific). Primary melanoma of cervix must be differentiated from metastatic melanoma from other sites of the body including skin and eye. Norris and Taylor have suggested the following criteria to diagnose primary malignant melanoma of the cervix (a) presence of melanin in the normal cervical epithelium (b) absence of melanoma in another site of the body (c) presence of junctional activity in the cervical epithelium near the lesion (d) if metastasis
is found, it should be according to the cervical carcinoma pattern.

As cervix is an unusual site for malignant melanoma, the international federation of gynecology and obstetrics (FIGO) staging system for cervical cancer is used rather than the Clark and Breslow scales, because the FIGO staging system correlates better with prognosis. There is no consensus on optimal management of primary malignant melanoma of cervix, because of its rarity. Radical hysterectomy with pelvic and para-aortic lymphadenectomy is the most common procedure which may be followed by radiotherapy or chemotherapy.

Melanoma of the cervix is a rare and aggressive neoplasm. The prognosis of primary malignant melanoma is generally poor because diagnosis is usually made at an advanced stage and the tumour is highly aggressive in both local recurrence and widespread metastases. Average survival reported in the world literature of these patients ranges from 6 months to 14 years.

CONCLUSION

Though primary malignant melanoma of the cervix is a rare disease, it should be considered in the differential diagnosis of cervical malignancies. The diagnosis should be confirmed using special stains and immunohistochemistry. No consensus has been established regarding treatment of primary malignant melanoma of the cervix. Prognosis of primary cervical melanoma is generally poor, because diagnosis is usually made at an advanced stage. Hence early diagnosis of cervical melanoma is essential.

REFERENCES