

A Case of Primary Malignant Melanoma of Ovary

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ABSTRACT

Primary ovarian melanoma is considered arising from mature cystic teratoma which is extremely rare condition. As best of our knowledge till date only 49 cases has been reported worldwide. Most cases are associated with disseminated diseases and poor prognosis. We present here a case of a 45 yr lady in perimenopausal age group, came with c/o heavy menstrual bleeding since 1 month and irregular menses since 1 year. The confirm diagnosis was made on the basis immunohistochemistry. Primary ovarian malignant melanoma is a definite entity and is associated with poor prognosis. Differential diagnosis is difficult for pathologist as it has to be differentiated from metastatic disease. The mainstay treatment of this disease is complete surgical debulking, however chemotherapy and immunotherapy has some role but patient outcome is unpredictable.

Key words: Exploratory laparotomy, immunohistochemistry, melanin bleach staining, Malignant Melanoma of ovary, ovarian tumour

INTRODUCTION

The ovary is a frequent site of secondary spread from extra-ovarian malignancies. Approximately 6–7% of the patients presenting with suspected ovarian neoplasm will prove to suffer from metastatic disease to the ovary. Ovarian involvement by metastatic malignant melanoma is relatively uncommon and it is rare for melanoma to present clinically as an ovarian mass. Solitary metastatic malignant melanoma to the ovary may be confused with primary ovarian carcinoma. We present this case report of metastatic ovarian malignant melanoma simulating primary ovarian cancer.

CASE REPORT

A 46-year-old female, P2 L2, housewife, residing at Sudumbre, Maval, came to gynecology outpatient department with c/o heavy menstrual bleeding for 1 month with irregular menses for 1 year. No c/o pain in abdomen/dysmenorrhea/lump in abdomen. On menstrual history, her last menstrual period was 1 month ago and she was bleeding continuously for 1 month. Her cycles were irregular for 1 year with moderate flow. On examination, there were no ascites and the abdomen was soft. On bimanual pelvic examination revealed, uterus was bulky, non-tender. Moreover, the left fornix was full. All the routine investigations are within normal limit. All the tumor markers were negative including beta-human chorionic gonadotropin, alpha-fetoprotein, lactate dehydrogenase, and carcinoembryonic antigen except cancer antigen 125 which was 303.5. Her pelvic ultrasound revealed a complex, well-circumscribed cyst of 7 cm × 6 cm seen in the left adnexa region with thick wall with soft tissue intensity. There were hemorrhagic products seen on gradient recalled

Access this article online	
Website: www.mmj.net.in	Quick Response Code 
DOI: 10.15713/ins.mmj.42	

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echo. The patient underwent exploratory laparotomy with frozen section. There was a large mass of 7 cm × 6 cm arising from the left ovary separate from uterus. The mass was capsulated and of firm to hard in consistency. Right adnexa normal. The uterus was 8 weeks size with no ascites and no omental deposits. There were no palpable lymph nodes. Tissues sent for frozen section which was inconclusive. Hence, total abdominal hysterectomy with bilateral salphingo-oophorectomy along with pelvic lymph node dissection was done. The pathology report macroscopically a single ovarian cystic mass of 7 cm × 6 cm in diameter with thickened wall, unilocular, few necrotic areas, with hemorrhagic fluid seen inside. On microscopy, the ovarian cyst shows thick-walled hyalinized fibrous tissue with cyst which is lined by pigment laden macrophages. The cyst wall slide showed collections of histiocytes containing golden brown pigments, diffuse infiltration by lymphocytes, and plasma cells with lymphoid follicle [Figure 1]. On immunohistochemistry, Melan A, HMB 45, Vimentin, and S100 were positive. This pattern of immunoexpression confirmed a diagnosis of malignant melanoma and since no evidence of a teratoma in the ovary was ever found, the diagnosis of metastatic malignant melanoma was made. The patient made a good recovery post-operative. After the histological diagnosis, she was clinically reassessed by a dermatologist for a full body check which showed no evidence of cutaneous melanoma. She was then reviewed by an oncophysician for further treatment.

DISCUSSION

Malignant melanomas of the ovary are neoplasms of neuroectodermal origin. Malignant melanomas involving the ovary are more frequently metastatic than primary in nature. Primary melanomas of the ovary are thought to arise on malignant transformation of a mature teratoma, which occurred in only 0.2-2%. Of these, the most common are squamous cell carcinomas. Less commonly adenocarcinoma, fibrosarcoma, carcinoid tumor. Hence, the possibility of a secondary involvement of the ovary from a tumor arising on another place must always be excluded from the study.^[1] The mean age at diagnosis of malignant melanoma metastases to the ovary is 38 years.^[2] Because it meets the criteria laid down by Cronje and Woodruff – (1) no other primary tumor, (2) unilateral ovarian tumor with the associated teratoid element,

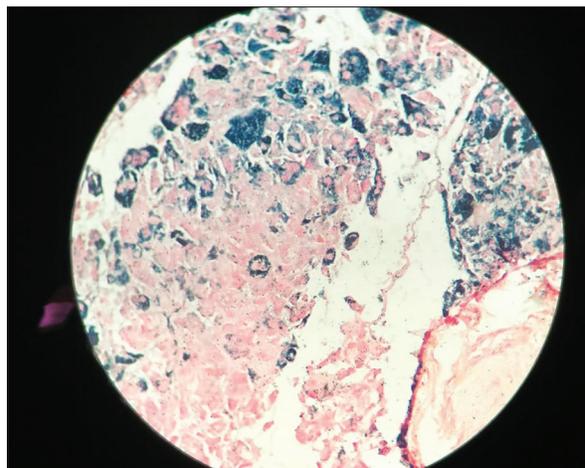


Figure 1: Histopathology: A case of primary malignant melanoma of ovary

(3) good correlation of patient age and symptom, and (4) demonstration of melanocytic junctional activity which is not mandatory for diagnosis.^[3]

The first case of primary ovarian malignant melanoma was found in 1901 by Andrews. Till date, only 49 cases have been reported worldwide. The patient age was 19-86 years (median 49.1).^[4] The presenting symptoms are due predominantly to an enlarged ovary causing abdominal distension and pain. The diagnosis of the disease is challenging for many reasons. As the diagnosis of melanoma in the ovary are made via immunohistopathological examination postoperatively (In one case described by Moselhi *et al.*, the diagnosis was established preoperatively through ascitic fluid cytology).^[5] Tumor markers are not discriminatory and ultrasonography and computed tomography scan were unable to characterize the disease. The pathological diagnosis is also difficult since the morphology of the lesion is often nonspecific. Diagnostic difficulties also arise histologically as the tumors do not have a consistent appearance and they can be mistaken for germ cell or sex cord-stromal tumors or granulosa cell tumor. Hence, definite diagnosis relies on immunohistochemistry. S-100 is expressed in nucleus and the cytoplasm (most sensitive marker) presents in 95% of cases. HMB-45 and Melan A are expressed in the cytoplasm.^[6]

The current recommendation in the limited literature available for the treatment of metastatic malignant melanoma to the ovary remains surgical which includes adequate tumor debulking and evaluation for local, regional, and distant spread. In perimenopausal or postmenopausal women, total abdominal hysterectomy with bilateral salpingo-

oophorectomy remains appropriate. In reproductive age, unilateral salpingo-oophorectomy is currently the treatment of choice if the other ovary appears normal. The adjuvant treatment using chemotherapy or immunotherapy has not yet been established.^[7] Chemotherapy (with different regimens consisting of dacarbazine, cisplatin, and taxol) could be considered as adjuvant treatment, but the patient outcome is unpredictable.^[7] Survival is poor despite aggressive surgical debulking with or without adjuvant therapy. The 5-year survival rate is reported as 11% with metastatic disease.^[6]

The overall prognosis is worse with more than 50% mortality rate in 18 months.^[8] The pattern of spread is to adjacent structures through lymphatic and hematogenous routes (lung, liver, and bone). The optimum treatment for this cancer remains to be defined, but a review of literature suggests a primary role for surgical resection of all tumors.^[9]

CONCLUSION

This case represents the diverse behavior of malignant melanoma. It highlights the difficulty in diagnosing a malignant melanoma. It also highlights the importance of immunohistochemical analysis for diagnostic purposes and the role of adequate surgical debulking to achieve optimal patient prognosis. More researches are required to establish methods for diagnosis preoperatively and definitive post-operative medical management to improve the survival of a fatal disease.

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How to cite: Ghunage SS, Pawar M, Bagade P. A case of primary malignant melanoma of ovary. *MIMER Med J* 2019;3(1):20-22.

Source of Support: Nil. **Conflicts of Interest:** None declared.

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