



### **Case Report:**

## **A Rare Case of Metastatic Intra-abdominal Melanoma Following Exenteration of Right Eye for Primary Choroidal Melanoma**

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**Abstract:** The most common primary intraocular malignant tumour is choroidal melanoma. Reported incidence of intraocular melanoma is less than 1 per 100,000. Liver is the most frequent site of metastasis, and involvement of the other sites generally occurs in association with liver metastasis. The present case had an unusual presentation of, rapidly refilling massive ascites and right pleural effusion, following exenteration of the right eye 4 years earlier for primary choroid melanoma. To the best of our knowledge this is the first case of intra abdominal melanotic rapidly refilling ascites following primary choroidal melanoma masquerading as an ovarian tumour. We have discussed the atypical presentation, difficulties in arriving at a diagnosis preoperatively, intraoperative dilemmas and the post operative management.

**Keywords:** Primary choroidal melanoma; Melanotic ascites; Debulking surgery.

### **Introduction**

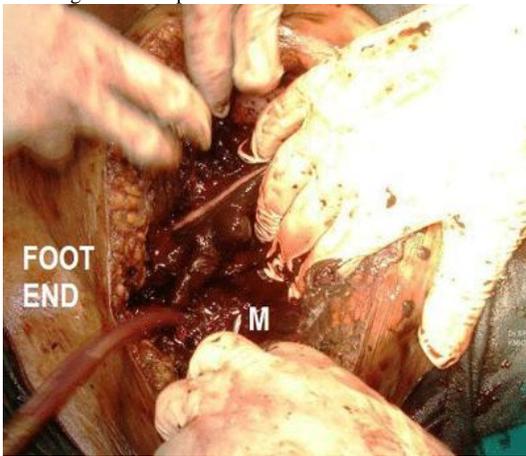
Malignant melanoma may involve choroids, ciliary body, retina or conjunctiva.(1) Choroidal melanomas are commonly asymptomatic and are often discovered during routine ophthalmic examination however; in some cases it may produce symptoms like loss of vision, photopsias and visual field defects.(2) Metastatic ovarian melanoma following cutaneous melanoma presenting as an acute abdomen due to intra abdominal rupture(3) and presenting as solid ovarian mass with massive ascites has been reported with dismal outcome.(4,5) The present case is unique, first it has followed

a primary choroidal melanoma, where there is absence of lymphatic drainage in the choroid. Dissemination (haematogenous?) into the abdominal cavity presenting as rapidly refilling ascites and right pleural effusion, but not involving the gastrointestinal tract, genital tract or the solid viscerae is worth noting.

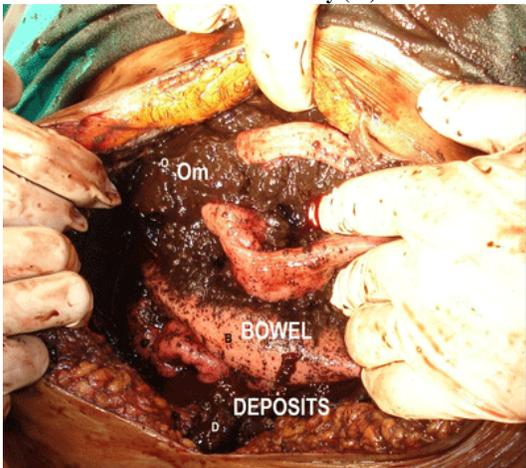
### **Case Report:**

A 54 year old woman presented with history of distension, pain abdomen, and breathlessness for the past 15 days. She had a past history of right orbital exenteration for primary choroidal melanoma four years back. Emergency paracentesis and pleurocentesis relieved her symptoms dramatically. Per speculum examination revealed normal cervix and vagina. Per vaginal and rectal examination detected a multiparous size uterus and an indistinct left adnexal mass probably measured 6x8x6 cms. Bilateral parametrium was supple and rectal mucosa was free. Provisional diagnosis of advanced ovarian malignancy was made. Her haemoglobin, white blood, platelet counts, biochemistry, electrocardiogram, echocardiography were normal. Serum CA-125 was 738.7 IU/ml and Carcinoembryonic antigen(CEA) was 0.523 U/ml. Chest x-ray was suggestive of right sided pleural effusion. Pleural and peritoneal fluid cytology and cell block studies revealed atypical cell clusters. Ultrasound examination of the abdomen and pelvis following paracentesis did not reveal any abnormality in the upper abdomen, other than basal pleural effusion. Uterus measured 5.7x2.3x3.3 cms; endometrial

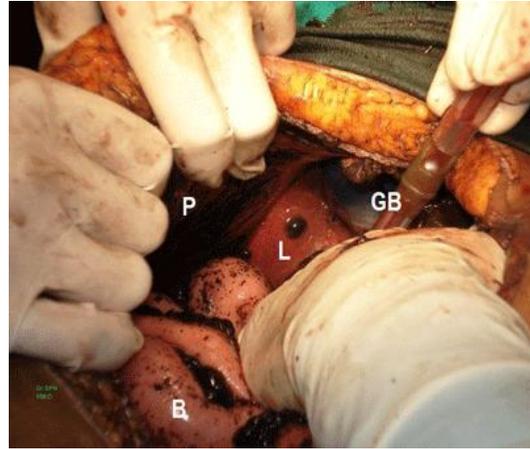
thickness of 2.7 mm. Adnexal echogenicity was noted on the left side measuring 5.2x5.7 x5 cms, right ovary was atrophic. Mild to moderate ascites was noted. During laparotomy it was surprising to find bulky dark brownish ,black pigmented deposits inside the abdominal cavity easily separating from the, peritoneal surfaces of bowel, mesentry, peritoneum, liver, gall bladder, stomach ,spleen ,undersurface of diaphragm. The omentum was studded with black deposits. Uterus and bilateral ovaries were atrophic and appeared normal. On table frozen section of omental biopsy was consistent with malignant melanoma- metastatic to omentum (Figure 1,2,3). Then, a decision for palliative surgery was taken and hence a panysterectomy and peritoneal lavage with warm normal saline was performed. She had a stormy postoperative period. With supportive therapy involving transfusion of blood and blood products, inotropic agents for cardiac support, transparentral nutrition to supplement the loss of proteins through the black coloured ascitic fluid she recovered and was discharged on day 14. She refused any further treatment postoperatively and expired after 4 weeks following a sudden episode of breathlessness.



**Figure 1: Blackish brown melanin pigment smearing the intrabdominal cavity.(M)**



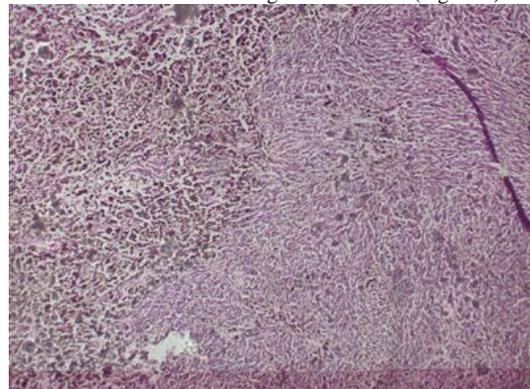
**Figure 2: Omentum (Om) studded with melanin deposits, loops of bowel (B) visible after clearing the melanin deposits, deposits of melanin (D) in the pelvis covering the atrophic uterus and ovaries.**



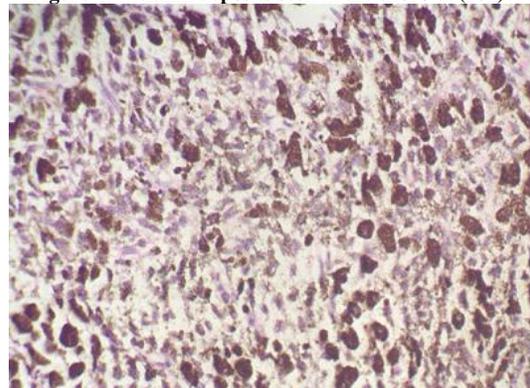
**Figure 3: Superficial deposits on the L-liver, P-parietal peritoneum, GB- gall bladder and B-bowel.**

**Pathology and microscopy:**

Total abdominal hysterectomy and bilateral salpingo-oophorectomy and omental biopsy specimen: Uterus measured 5.2x2.1x3.5 centimeters, cervix, endometrium, myometrium, both fallopian tubes and both ovaries were unremarkable. Sections of omentum showed clusters of melanin pigments, consistent with metastatic malignant melanoma (Figure-5)



**Figure 4: Melanin deposits seen in the choroid. (10x)**



**Figure 5: Melanin deposits seen in the omentum.(40x)**

**Discussion:**

Choroidal melanoma arise from uveal melanocytes residing in the uveal stroma and originating from the neural crest.(8) Choroidal melanomas are quite different from their cutaneous counterparts with regards to presentation, metastatic pattern and treatment. Lymphatic spread has not been demonstrated, consistent with the absence of lymphatics in the choroid. The

incidence of intraocular melanoma is less than 1 per 100,000.(9) Spread to the liver is more frequent, while metastases to other sites- lung, heart, gastrointestinal tract, lymph nodes, pancreas, skin, central nervous system, bones, spleen, adrenal, kidneys, ovaries, thyroid, contralateral choroids, breast generally occur in association with liver metastases.(10)

It is interesting to note the acute presentation of the present case mimicking an ovarian tumor, with high serum CA-125 levels. However, she needed an emergency exploration to relieve her symptoms and for a definitive diagnosis. Our dilemma was clarified only when the intra-operative appearance of bulky, blackish brownish deposits were seen in the abdominal cavity and further ,on table frozen section of omental biopsy revealed metastatic melanoma. Similiar experiences have been reported by Habek.D et al (3) and Abe Y et al.(4)

As there was no gross parenchymal involvement of the solid intra abdominal organs, bowel and its mesentery, internal genital organs, no palpable para aortic or pelvic lymphnodes, decision for only a palliative total abdominal hysterectomy and bilateral salphingo-oophorectomy with omental biopsy and peritoneal lavage to wash out the bulky blackish brownish deposits all over the peritoneal cavity was performed.

Authors have reported the role of palliative and complete resections when there is involvement of bowel and solid organs and have had improved overall survival in metastatic melanoma.(6,7) However, other authors have also reported the dismal performance and high morbidity and mortality associated with ovarian metastasis and further dissemination (4,5), as in the present case . Due to rarity of these presentations each patient needs to be assessed for the type of surgical intervention and follow up. A cost effective adjuvant therapy needs evaluation in this clinical scenario!

Intra-abdominal malignant melanoma secondary to a primary in the choroid, following exenteration of the orbital contents mimicking as an advanced malignant ovarian tumour is a very rare clinical entity. It needs to be considered in the differential diagnosis. They have very poor prognosis. The adjuvant therapy in a low resource setting needs to be researched.

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