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Malignant transformation in mature cystic teratoma: A case series

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Abstract
Background: Mature cystic teratoma is the most common ovarian neoplasm of adolescent and reproductive age group. Squamous cell carcinomas account for most common type of somatic transformation followed by adenocarcinoma.
Case Summary: We present a case series of 4 patients with 4 different types of malignant transformation in dermoid and their management at our Institute. The types include Mucinous Adenocarcinoma, Anaplastic Ependymoma, and Rhabdomyosarcoma besides Squamous Cell Carcinoma.
Conclusion Malignant degeneration is rare and continues to be evasive in literature, posing detection and management difficulties. A large dermoid of >10cm in a post-menopausal lady, with a solid enhancing component should arouse suspicion of MT-MCT, especially with deranged serum markers. Overall, treatment must be dictated by the transformed malignant histology. Metastatic tumors must be differentiated from MT-MCTs. It can be safely said that there are no standard rules for these entities in terms of clinico-pathological behaviour, management or prognosis. What needs to be done is to keep a high index of suspicion, as to avoid violation of oncological principles.

Keywords: Ovarian dermoid cyst, malignant transformation, squamous cell carcinoma, mucinous adenocarcinoma, rhabdomyosarcoma, anaplastic ependymoma

Introduction
Mature cystic teratoma (MCT)is the most common (30-50%) ovarian neoplasm of adolescent and reproductive age group, accounting for 60% of all benign tumors [1]. They are derived from the primordial germ cell and malignant transformation of Mature cystic teratoma (MT-MCT) can occur in 0.2-2% cases. [2, 3] Squamous cell carcinomas (SCC) account for more than 80% of these secondary tumors, followed by adenocarcinoma [4]. As it is impossible and possibly irrelevant to distinguish between de-novo co-existence in MCT and MT-MCT, these terms are used interchangeably in literature; and restricted to cases without immature teratoma.
The phenomenon malignant transformation in dermoid cyst was first reported by Johannes Sultetus in 1659 [5]. Since, then there are have been multiple case series and retrospective studies with mixed conclusions. We present here a bouquet of 4 cases with 4 different histotypes of MT-MCT at Department of Gynaecological Oncology, Kidwai Memorial Institute of Oncology, Bengaluru. The cases collected were between 2017-2021.

Case 1: Squamous Cell Carcinoma (SCC) in Dermoid
A 58-year-old post-menopausal lady presented with pain abdomen and CECT showing a 15 x 15 cm abdomino-pelvic calcified solid-cystic mass; CA19.9 was 2,067 U/ml, rest all tumor markers were normal. Frozen section was SCC in mature teratoma. She underwent Total abdominal hysterectomy with bilateral salpingoopherectomy with total omentectomy and retroperitoneal lymph node dissection. Despite Stage IA of SCC in teratoma, the tumor board favoured adjuvant chemotherapy. The patient, however, defaulted, and presented 9 months later with a huge stony hard mass infiltrating the anterior abdominal wall and bladder. The *Figure 1* shows the preoperative image of abdominal wall recurrence. Following poor response to 2 cycles of carboplatin and paclitaxel, she underwent tumor debulking with complete tumor resection and abdominoplasty shown in *figure 2.* She received 6 cycles of Gemcitabine and Docetaxel as did not respond to neoadjuvant cycles of carboplatin and paclitaxel. She was disease free for 10 months, as in the last follow up 3months back.

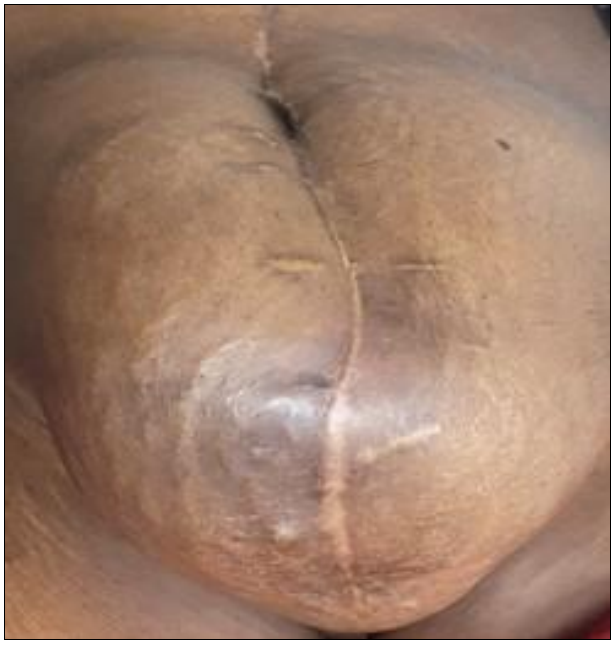

Fig 1: Preoperative picture of abdominal wall recurrence

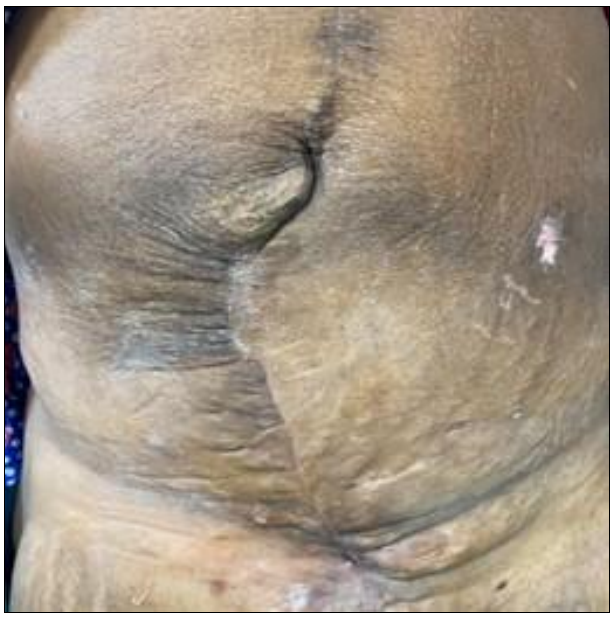

Fig 2: Post operative picture showing healed abdominoplasty scar

Case 2: Mucinous Adenocarcinoma (MAC) in Dermoid

A 15-year-old girl evaluated for irregular menstrual cycles of 4 month's duration. Her CA125 was 250 U/ml and CA19.9 was more than 1000 U/ml. CECT showed a solid-cystic lesion of 24*15*11cm. Frozen section was reported as immature teratoma. She underwent right salpingoopherectomy, total omentectomy and bilateral pelvic lymph node dissection. Final histopathology was reported as "Mucinous carcinoma in teratoma" with stage 1A. As per the decision of the Multi-Disciplinary Tumour board, she was administered 6 cycles of paclitaxel and carboplatin (TC). She continues to be disease free for 5 years.

Case 3: Anaplastic Ependymoma in Dermoid

A 23yr lady presented with bilateral solid-cystic masses (left ovary-18x12cm and right ovary-8x6cm), moderate ascites and peritoneal deposits; only CA125 was raised upto 118 U/ml. Patient was planned for exploratory laparotomy after negative metastatic work up and proper consent of non-fertility preserving surgery. Frozen Section was reported as "metastatic poorly differentiated carcinoma". She underwent total abdominal hysterectomy, bilateral salpingo-oophorectomy, total omentectomy and partial pelvic peritonectomies were done. Final histopathology report was Anaplastic ependymoma of WHO grade III in an ovarian teratoma; Stage IIIC. Immunohistochemistry markers were positive for GFAP, EMA, S100 positive and was Synaptophysin negative. Post operative stay was uneventful. She was planned for 6 cycles of CAP (cyclophosphamide, Adriamycin and cisplatin) in multidisciplinary board meeting but did not come for follow up. A telephonic conversation confirmed that patient had recurrence 6 months later, took treatment at the local hospital but expired on treatment. No further details could be gathered.

Case 4: Rhabdomyosarcoma (RMS) in Dermoid

An 18-year-old girl with secondary amenorrhoea of one year duration, was detected to have a 22*20*10 cm unilateral cystic mass with enhancing solid area and septae; CA125 was 376 U/ml, rest all was normal. Laparotomy revealed ascites with pre operative capsule rupture. Frozen section was reported as "sex cord stromal tumor". She underwent adenexectomy and infracolic omentectomy. Final hitopathology revealed "Mature cystic teratoma with rhabdomyosarcoma" of stage 1C3 with IHC positive for Desmin, Myo D1, Myogenin; negative for Pan CK; and Ki67 high of 70%. She was planned for 6 to 8 cycles VAdC(Vincristine, Adriamycin Cyclophosphamide), but patient refused to take chemotherapy and presented after 3 months with gross ascites, dyspnoea, and renal failure. After stabilizing the patient, first cycle of VAdC regime was administered. Patient deteriorated and died despite intensive care admission.

Discussion

MCTs are the most frequent subtypes of ovarian germ cell tumours. It is reported that the incidence of malignant transformation of MCTs is 0.2%-2%, accounting for 2.9% of all malignant OGCTs.[2,3] Any component of MCTs can progress to malignancy, but the most common malignant transformation type is squamous cell carcinoma (SCC) from the ectoderm SCCs accounting for nearly 80% of MT-MCTs followed by adenocarcinoma; carcinoids come in the third place, while others include adeno-squamous carcinoma, sarcomas, melanomas, meningioma, malignant germ cell neoplasms, basal cell carcinoma, neuroectodermal tumor, thyroid cancer, transitional cell tumor, signet ring cell carcinoma, oligodendroglioma, and sebaceous carcinoma, undifferentiated carcinoma, borderline tumor, melanoma, small cell carcinoma.[6-8] The squamous epithelium, which is commonly present in dermoid cysts, is considered particularly susceptible to neoplastic changes due to its high turnover rate. Traditional concept has been that MTs are observed 10-15 years after first detection of MCTs, suggesting role of mutagens in the peritoneal cavity. Mutations in key oncogenes and tumor suppressor genes, including TP53 and PIK3CA, have been implicated in the malignant progression of these lesions.[9] Although multiple studies have shown the association of SCC in MCT with high-risk HPV, no therapeutic or prognostic implications are yet to be derived from this information.[10]

High suspicion of malignant transformation be raised if occurring if older age group, this seems true for SCC, in our case series. Cases with >10cm in size, raised tumor markers (CA125, CEA,CA19.9 or SCCAg) and irregular solid areas in pre operative imaging with enhancing solid component [4]. This was similar in our case series as all the 4 patients had >10cm size

solid-cystic tumor, with at least one raised tumor markers. Dermoid can be bilateral in 10% of cases. [5]In our case series, one out of 4 was bilateral.

Considering the rarity of MT-MCTs, information is sparse on preoperative and intra-operative prediction, extent of cytoreduction, adjuvant therapy and prognosis. Laparotomy is preferred over laparoscopy to avoid spillage. The general consensus is that complete staging and adjuvant chemotherapy are ideal even in stage IA tumours. Need for lymph node dissection remains debatable, considering the direct / peritoneal spreading nature. Platinum-based chemotherapy is found to improve overall survival. Safety and feasibility of fertility sparing surgery in women with SCC in MCT <45 years of age with apparent stage IA/IC disease has been successfully studied in few studies.[10]

The diagnostic accuracy of frozen for MT-MCTs varies with expertise, with reported sensitivities as high as 100%.[6] Although in our case series only SCC in dermoid could be diagnosed in frozen. Frozen section also relevant in the setting of a planned fertility sparing in young patients as the other 3 cases. Rokitansky's protuberance at the junction between the teratoma and normal ovarian tissue has greatest cellular variety and should be examined carefully to exclude malignant components.[11]

Survival in SCC is as high as 93% for Stage 1 tumours, while it is dismally low (~10-30%) for advanced disease; patients who received chemotherapy in the postoperative period performed better than those who did not. Literature does not support the role of adjuvant radiotherapy in SCCs, despite the generally radio sensitive nature of the histotype [4].

There are multiple case reports of SCC in a dermoid recurrence. It can contain high number o mutations, hence response in patients with either PD-L1-positive or TMB-high status may be expected. Recently, Immune checkpoint inhibitors like Pembrolizumab, Nivolumab and Sintilimab has been successfully tried [12, 13].

Mucinous adenocarcinomas (gastro-intestinal type) arising from MCTs is rare, with a reported incidence of 7%-44% of all the malignant transformation in MCT [14, 15]. It stains positive for CK20, CDX2 and MUC2. They usually present in early stage, and have a better prognosis than other MTs; the same does not apply to the occasional patient with MCT rupture and disseminated disease.[15] Some authors have preferred 5 FU based FOLFOX as adjuvant therapy for Mucinous adenocarcinoma.[16]

Neuroectodermal differentiation is exceedingly rare, with only a few cases of gliomas and primitive neuroectodermal tumors reported. Ependymoma transformation within an ovarian teratoma is exceptionally rare, with anaplastic variants posing a significant clinical challenge due to their aggressive nature. Anaplastic ependymoma is a subtype of grade 3 neuroectodermal tumours, the latter classified a subtype of MT-MCTs by the WHO in 2020. [17] The authors could identify only 8 such cases in recent literature. Most of these cases are seen in the younger age group (range: 17 - 35 years). Reports shows performance of fertility sparing surgery with no significant effect on prognosis. Histologically, ependymomas demonstrate perivascular pseudorosettes, which were present in the histopathological appearance of our third case. IHC is negative for cytokeratin 7 (CK 7), positive for glial fibrillary acidic protein (GFAP), nestin and S100, this was similar to our third case [18-20].

Embryonal rhabdomyosarcoma within a dermoid tumor likely arises from the mesodermal component, representing aberrant differentiation of immature mesenchymal elements. We identified around 4 prior reported cases of RMS in MCT. Proposed theories parallel that of origin of ovarian carcinosarcomas; association with DICER1 mutations have been noted in pure RMS (non-MCT). The behaviour of extra ovarian RMS is less aggressive than primary ovarian RMS. VAC(Vincristine, Ifosfamide and Adriamycin) appear to be the preferred regimes. Histologically, it is characterized by primitive small round cells with rhabdomyoblastic differentiation, confirmed by immunohistochemical markers such as desmin, myogenin, and MyoD1 and negative for Pan CK with a high Ki67 [21, 22].

Conclusion

Malignant degeneration is rare and continues to be evasive in literature, posing detection and management difficulties. A large dermoid of >10cm in a post-menopausal lady, with a solid enhancing component should arouse suspicion of MT-MCT, especially with deranged serum markers. Overall, treatment must be dictated by the transformed malignant histology. Metastatic tumors must be differentiated from MT-MCTs. It can be safely said that there are no standard rules for these entities in terms of clinico-pathological behaviour, management or prognosis. What needs to be done is to keep a high index of suspicion, as to avoid violation of oncological principles.

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Ethics and Consent

Proper written consent has been taken by the patients.

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