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Case Report

Rare Case: Sebaceous Carcinoma Arising within an Ovarian Dermoid Cyst

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Abstract

This case report discusses a rare find of a sebaceous carcinoma within an ovarian dermoid cyst in a 49-year-old patient. There have been 10 reported cases worldwide as of August 2018. This report discuss not only the case itself but the subsequent discovery of a previously unknown hereditary condition in the patient in question.

Keywords: *Ovarian Dermoid; Sebaceous Carcinoma; Mature Cystic Teratoma; Lynch Syndrome*

Introduction

Sebaceous carcinomas are a rare form of cancers accounting for approximately 0.7% of all skin cancers within Caucasian, Asian and Indian populations. 75% of these are periocular and thus occur in or around the eyelid. They are a result of neoplastic growth of sebaceous glands of which most are located in the head and neck region of the body. Dermoid cysts of the ovary, also known as mature cystic teratomas are a common germ cell tumour which differentiate in an abnormal manner over time. These structures are often found to have tissues and mature structures inside such as hair, bone, cartilage, fat, thyroid tissue and even teeth. Due to their slow growing nature, the majority of dermoid cysts are benign, with only roughly 1-2% exhibiting malignant changes [1]. As of August 2018, there were 10 documented cases of a sebaceous carcinoma arising within a mature cystic teratoma of the ovary in the world [2-11]. Whilst many of these cases have been idiopathic in their origin, it has been suggested that hereditary conditions such as Lynch syndrome and Muir-Torre syndrome play a role in this rare tumour finding.

Case Report

A 49-year-old female presented to her general practitioner complaining of a feeling of fullness in the right pelvis ongoing for several months. On examination, she was found to have a palpable fullness in the pelvis and had a Ca-125 of 28. It was also noted at that time that there was a family history of ovarian cancer and thus an ultrasound of the pelvis was arranged. The ultrasound showed a left adnexal cyst measuring 11.1cm by 11.5cm described as thick walled and containing mural nodules and low-level echoes. The patient was referred to a red flag gynecology clinic following consultant radiologist advice.

The patient was seen at the clinic one week later and further investigations were sought including CEA and Ca19-9 as well as a CT of the chest, abdomen and pelvis. CT showed a left adnexal mass of mixed contents with no evidence of metastatic disease. The mass measured 11 cm by 10cm by 11cm in size and found to contain soft tissue, cystic, calcified and fat density components. It was planned to discuss the patient at a multi-disciplinary meeting following CT result. A total laparoscopic hysterectomy and bilateral salpingo-oophorectomy was arranged for the following month and specimens were sent to histopathology for evaluation. A ruptured capsule was noted on histopathological examination leading to a query over spillage of tumour contents. Following histopathology, as detailed below, the patient was referred to medical oncology. Due to the rarity of this tumour, it was not recommended by the oncology team for any adjuvant therapy due to uncertain benefit. Follow up CT scans were suggested every 3 months for the 1st year to be reviewed thereafter. It was also found upon consultation that there was a strong family history of various cancers including paternal bladder cancer, uterine and bowel cancer of a paternal aunt and bowel cancer of another paternal aunt. This discovery was particularly significant in the case.

A CT of the chest, abdomen and pelvis was carried out 6 weeks postoperatively and was concluded to have normal postoperative appearances with no evidence of residual or metastatic disease.

Pathological Findings

The initial pathologist's report commented that the sampled tissue consisted of almost entirely of a sebaceous carcinoma with a focus of normal thyroid tissue and fat necrosis. The left ovary was described as being 10cm in maximum dimension with a ruptured capsule and was filled with solid sebaceous material. A diagnosis of sebaceous carcinoma arising within a background of a teratoma was reached with a provisional FIGO stage of 1C applied at that time. Following the report, it was decided to discuss the case at MDM, at which point it was asked to be reviewed by the professor of histopathology of Northern Ireland due to the rare nature of the case. The left ovary was noted to show a little unremarkable thyroid tissue together with fat necrosis as well as carcinoma in several of the sections present with large areas of necrosis. The carcinoma was composed in areas of cells with scant cytoplasm but predominantly showing abundant foamy cytoplasm.

Furthermore, the tumour cells showed loss of normal nuclear staining pattern for the mismatch repair proteins MSH2 and MSH6, with retention of staining in peritumoral lymphocytes. The involvement of the mismatch repair pathway to carcinogenesis in such a young patient along with a strong family history was highly suggestive of Lynch syndrome (also known as HNPCC – hereditary non-polyposis colorectal cancer).

Following review by her gynaecologist and medical oncologist, the patient was referred to a geneticist and bloods were taken to test for a mismatch repair gene abnormality, namely within MSH2 and MSH6.

Discussion

Dermoid cysts, also called mature cystic teratomas, are one of many ovarian teratomas. Others include immature teratomas and mesodermal teratomas such as ovarian carcinoid tumours or struma ovarii. Whilst mesodermal teratomas are made up of one tissue type, both mature and immature teratomas are compiled of multiple germ cell layers (ectoderm, mesoderm and endoderm). The major difference between these is that immature teratomas are more likely to be malignant in nature. In the setting of the ovary, approximately 95% of teratomas are of the mature cystic type and are usually benign due to the slow growth noted over a prolonged period of time.

Dermoid cysts develop from germ cells which proliferate in an abnormal manner leading to the development of mature tissues within. Ectoderm can be present in the form of skin and sebaceous glands. Mesoderm can give rise to muscles, cartilage, bone and urogenital structures. Endoderm can lead to development of lung and gastrointestinal tissue. A retrospective study of 501 patients over the course of 34 years between 1964 and 1998 conducted by [1], found that malignant transformation was found in 1.4% of mature cystic teratomas. Whilst another study of 283 cases first published in 1996 by R T Wu et al., 1996, found there to be malignant transformation in 0.7% of those cases, all with a pathology of squamous cell carcinoma. Overall, it is deduced from various studies that malignant transformation of these cysts is less than 2% and is often a squamous cell carcinoma derived from squamous epithelium. Thus, this particular case joins a small group of only 10 previously documented cases of sebaceous carcinoma within a dermoid cyst as of 2018.

According to the last case reported by Lima RB et al in 2018, 3 of the 4 features mentioned which are frequently seen in cases of malignant transformation are present. These are the patient being over 45 years old, a cyst diameter greater than 10cm and a fast growth rate of the dermoid cyst. The patient in question had an ultrasound of the pelvis less than 3 years previously and at that time there was no ovarian cyst present.

While most cases are idiopathic, there is a significant association with Muir-Torre syndrome. This is a rare phenotypic variant of Lynch syndrome with autosomal dominant inheritance and characterized clinically by the presence of at least one sebaceous skin tumor and at least one visceral malignancy.

Lynch syndrome (hereditary non-polyposis colorectal cancer) is an inherited, autosomal Dominant, cancer predisposition syndrome that increase the lifetime risk of colorectal (up to 68%), endometrial (up to 62%), ovarian (up to 39%) cancer at young age with slight increase in GIT, urinary tract and Brain cancer and it is caused by a variant in one of the mismatch repair genes (MLH1, MSH2, MSH6 orPMS2).

Therefore, it is important to consider the broader impact of a diagnosis of sebaceous carcinoma and refer for genetic testing In the presenting case, the subsequent diagnosis of Lynch syndrome based on her family history of disease and the proven DNA mismatch repair within MSH2 and MSH6 is significant. There is now awareness of the need for prevention of other cancers including colorectal, endometrial, gastric and ovarian cancers, not only in the patient herself but also in her child who has a 50% chance of inheriting the genetic mutation.

Consent

Written informed consent was obtained from the patient for the publication of this case report, a copy of the written consent is available for review upon request.

Conflict of Interest

The authors have no relevant financial relationships or conflicts of interest to report.

Author Contribution

All authors contributed to the literature search, all authors critically reviewed, edited and approved the final manuscript for publication.

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