Case Report | Nepal Journal of Health Sciences
Submitted:10/03/2022
Accepted:08/06/2022
Published:25/06/2022

Gliomatosis Peritonei associated with Ovarian Immature Teratoma - A rare entity

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ABSTRACT

Gliomatosis peritonei is a rare entity associated with ovarian teratoma, both mature and immature. We present a case of a 24-year-old female diagnosed with immature ovarian teratoma, grade 2 on histopathological examination. The presence of mature glial tissue on the omentum tissue established the diagnosis of gliomatosis peritonei. It is a harmless condition with a favorable prognosis. However, regular follow-up is required as it is associated with the risk of recurrence and malignant transformation. Adequate sampling and examination of multiple biopsies for the presence of an immature neural element is important to rule out metastatic involvement.

Keywords: Gliomatosis peritonei; immature teratoma; prognosis.

INTRODUCTION

Immature teratomas are defined as teratoma consisting of a variable amount of primitive neuroectodermal components. Gliomatosis peritonei is a rare entity especially associated with immature teratomas. It is characterized by peritoneal and omental implants composed of mature glial tissue. Although a harmless condition, it is often associated with recurrence in cases of incomplete surgical excision and rarely with malignant transformation. Considering its rarity, one needs to be aware of this entity and meticulously examine the sections with extensive histological sampling to look for an immature neural component to exclude metastatic immature teratoma.

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Citation
Maharjan D, Gaire D, Dhakal S, Tiwari SB. Gliomatosis Peritonei associated with Ovarian Immature Teratoma - A rare entity. Nepal J Health Sci. 2022 Jan-Jun;2(1):114-6

CASE REPORT

We present a case of a 24- year -old female who had been experiencing abdominal distension and discomfort for the past five months. On abdominal examination, a huge non-tender mass was palpable corresponding to the size of the uterus at 24 weeks of gestation. Serum alpha feto protein (AFP) was 45u/l. Other tumor markers such as lactate dehydrogenase (LDH), carcinoembryonic antigen (CEA) and beta human chorionic gonadotropin (HCG) were within the normal limit.

USG showed a massive pelvic cystic mass with septations suggestive of ovarian neoplasm. CT scan revealed acyst measuring 20.6 cm X 19.9 cm X 8.8 cm over the left adnexa.

She underwent staging laparotomy with left salpingo-oophorectomy with appendectomy and omentectomy along with pelvic lymph node dissection.

We received the histopathological specimen consisting of an ovarian cystmeasuring 13x12x 7 cm, cut section of which showed a multiloculated

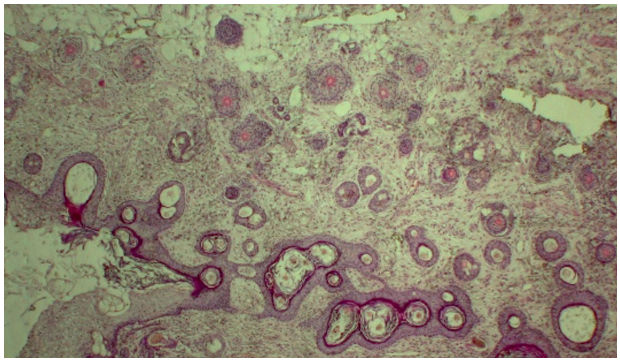

Figure 1: Mature component showing keratinized stratified squamous epithelial lining and skin adnexal structures. HE stains 200X

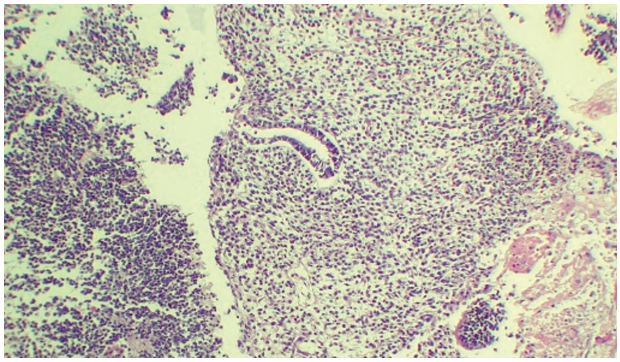

Figure 2: Immature components are composed of small cells arranged in solid sheets and tubules. HE stains 200X

cyst filled with clear yellow fluid with irregular solid areas. The solid areas consisted of bony areas and hair tuft. The attached fallopian tube was grossly unremarkable. Received specimen labeled as omentum consisted of fibrofatty tissue measuring 8x4x3.5 cm with no omental deposits noted on gross examination.

On microscopic examination,cyst lined by keratinized stratified squamous epithelium was noted with underlying adnexal structures such as pilosebaceous unit, eccrine glands, adipose tissue as well as pancreatic acini (Figure 1). Some of the cysts were lined by simple mucinous epithelium. Mesenchymal components consisting of cartilage, bone, smooth muscles and adipose tissue were noted. Extensive areas of mature glial tissue with scattered ganglion cells were seen with some areas showing increasing cellularity and few mitotic figures (Ki-67= 3%).

The immature component consisted of more than one low power field but less than three low power fields. It was composed of small cells arranged in solid sheets, tubules and rosettes. These cells had a high N: C ratio, hyperchromatic nuclei with inconspicuous nucleoli and scant cytoplasm (Figure 2). Occasional mitotic figures were also noted.

Extensive sampling was done from the ovarian mass to look for the components of other germ cell tumors such as yolk sac tumor or embryonal carcinoma but they were not present.

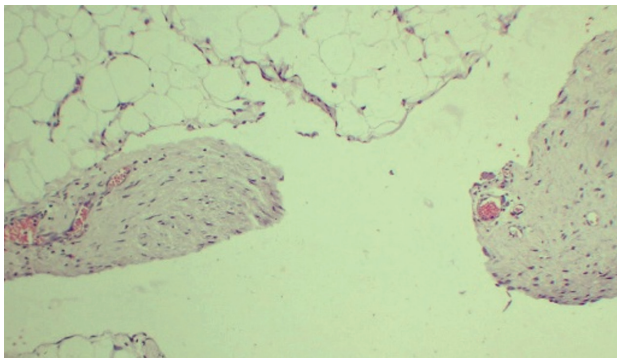

Figure 3: Gliomatosisperitonei showing nodules of mature glial tissue and adipose tissue of the omentum. HE stains 200x

Sections from the omentum showed adipose tissue infiltrated by mature glial tissue corresponding to the diagnosis of gliomatosis peritonei (Figure 3). Immature neural components were not seen even on extensive sampling.Pelvic lymph node dissection showed three reactive lymph nodes. There was no evidence of nodal gliomatosis as well as metastatic immature teratoma.

Hence, following the histopathological examination, diagnosis of pure immature teratoma, grade 2 with gliomatosis peritonei was made. The patient had an uneventful postoperative recovery and was referred to the cancer center and has undergone three cycles of chemotherapy.

DISCUSSION

Immature teratoma constitutes about a third of malignant germ cell tumor.[1] According to WHO,

immature teratomas are defined as teratomas with a variable amount of primitive embryonal-type tissues, typically neuroectodermal tissue arranged in tubules and rosettes. The prognosis of immature teratoma depends on staging and its grade. Grading depends on the amount of neuroectodermal tissue present. In our case, primitive neuroepithelial tissue was present in more than one low power field but less than three low power fields making it an immature teratoma of grade 2 as per the three-tiered grading system.

Immature teratomas can have peritoneal and omental involvement in the form of implants composed of mature glial tissue. This is a rare entity and is termed gliomatosis peritonei. Cases of immature teratoma with nodal gliomatosis have also been reported with no change in overall prognosis.[2,3] Rarely, gliomatosis peritonei can coexist with endometriosis.[2,3]There are several theories on the origin of gliomatosis peritonei but the metaplastic theory is the most accepted one which explains it as a metaplastic response to neoplastic stimuli.[4]

Immature ovarian teratoma with gliomatosis peritonei has a favorable prognosis.[3]The fate of these implants includes either fibrosis with disappearance or persistence without any morphological changes. Rarely, they can undergo malignant transformation.[5]

Immature teratoma of grade 1 that is limited within the ovary can be managed with surgery alone but grade 2 or grade 3 immature teratoma requires additional chemotherapy. Complete resection is difficultin cases of extensive peritoneal gliomatosis causing residual peritoneal disease. This results in the risk of recurrence and malignant transformation. Hence, a long-term follow-up is necessary for such patients.[5]

The unfavorable outcome in cases of immature teratoma with gliomatosis peritonei has been noted to be associated with a lack of extensive histological sampling.[6] Thus, adequate sampling of the specimen and multiple biopsies are required to exclude immature glial tissue or teratoma elements. The presence of immature neural tissue confirms the metastatic involvement and influencesthe treatment and prognosis.[5]

SUMMARY

Gliomatosis peritonei is a rare entity associated with ovarian immature teratoma. It has a favorable prognosis, provided that metastatic peritoneal involvement is ruled out. Patients with the persistent peritoneal disease are at the risk of recurrence and malignant transformation. Hence, regular follow-up with a CT scan is imperative.

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