Embryonal Rhabdomyosarcoma of the Uterus Corpus : An unusual presentation of a rare disease

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Introduction

- Rhabdomyosarcoma of the uterus corpus is a rare tumour that predominantly affects adolescent and young adults [McCluggage 2002, *Int J Gynecol Cancer*, 12(1):128-32]
- It can be classified into three main groups
  - Embryonal (most favourable type)
  - Alveolar
  - Pleomorphic
- To date, less than 60 pure uterine rhabdomyosarcomas have been reported in the literature [Garrett 2012, *J Clin Oncol*, 31(4):48-50]

Objective

- The purpose of this case report is to highlight the challenges in identifying and managing rhabdomyosarcoma of the uterus

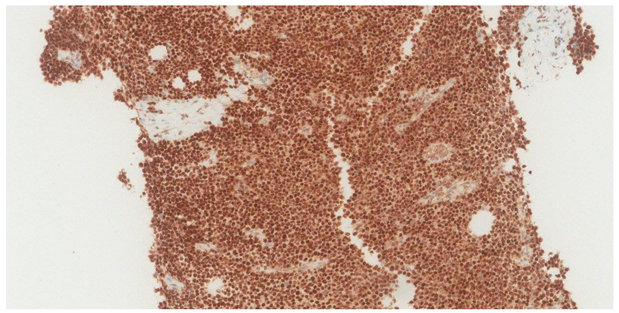

Image 1. Immunohistochemistry of paraffin-embedded human rhabdomyosarcoma tissue slide stained with MyoD1.

Case Description

We report the unusual case of a 34 year old south asian woman who presented with a three week history of abdominal ascites and loss of appetite. Tumour markers were sent, and they revealed a raised CA125 at 263 but normal CA19-9, CEA and CA153. CT abdomen and pelvis showed peritoneal carcinomatosis with uterine mass, pelvic lymphadenopathy, ascites, pleural effusions, and probable pleural metastatic deposits. The differential diagnosis was possibly uterine, ovarian or primary peritoneal malignancy with a separate uterine pathology or possibly infective causes (e.g. tuberculosis). Cytology of ascitic fluid supported an inflammatory and reactive picture rather than neoplastic while test for mycobacterium and acid fast bacilli were both negative. However, her ascites rapidly refilled and her symptoms acutely worsened. Further investigation including an ultrasound guided biopsy of her omental cake was performed which demonstrated an atypical malignancy. After a panel of immunohistochemical markers such as MyoD1 (**Image 1**) and fluorescence in situ hybridisation the final histology report concluded that the appearances were of embryonal rhabdomyosarcoma. Following her diagnosis, she underwent three cycles of radical (curative) chemotherapy at the local cancer unit. This was unfortunately complicated by the re-accumulation of ascites and pleural effusions despite repeated ascitic drainage and pleural aspirations. Pleural fluid cytology that was sent was consistent with metastatic rhabdomyosarcoma. She continues to undergo further chemotherapy treatment with support from the palliative care team albeit with a poor prognosis.

Discussion

- Rhabdomyosarcoma may originate at any site and the uterus is rarely the primary site
- Clinical symptoms are often nonspecific and may present as abdominal distension and investigative findings maybe of uterine mass as in this case
- It can appear at any age making it diagnostically challenging
- The commonest site of distant metastases for embryonal rhabdomyosarcoma are lungs, liver, bones and brain
- Review of the literature revealed treatment options included surgical excision, radiotherapy and chemotherapy
- In most reports on uterine rhabdomyosarcoma, the main determinant factor of disease prognosis appears to be the severity of disease at initial presentation [Garrett 2012, *J Clin Oncol*, 31(4):48-50]

Conclusion

- Pure rhabdomyosarcoma of the uterus is a rare tumour
- Uterine rhabdomyosarcoma in adults have a higher tendency to develop distant metastases or recurrence as in this case [Yamada 2016, *Diagn Pathol*, 11:3]
- Early identification and radical surgical treatment is pertinent to improve survival outcomes