

# A Diagnostic Curiosity: Mature Cystic Teratoma Of The Ovary With Pneumatosis Cystoides - Like Appearance: Case Report And Review Of Literature

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## Abstract

Multicystic appearance in a mature teratoma is a common feature of a common tumour but little information is available on the mechanism of their constitution. Various authors have named this multicystic appearance differently and have given several hypotheses for their occurrence. The present case report describes a case of mature cystic teratoma in a 38 year old female with multiple cystic spaces resembling those seen in pneumatosis cystoides intestinalis, with a review of cases of mature cystic teratoma with multicystic appearance described in the past.

## INTRODUCTION

Mature teratomas are cystic or, rarely solid tumors that contain various mature tissues derived from one or more of the embryonic germ layers: the ectoderm, mesoderm, and endoderm. Multicystic appearance in a mature teratoma is a common but rarely described feature. In the past only 21 cases have been reported with such an appearance.<sup>234</sup>

## CASE REPORT

A 38 year old female presented with history of lower abdominal pain during menstruation of 6 months duration. She was evaluated and clinically diagnosed to have an intramural fibroid and an endometriotic cyst in the right ovary. Subsequently, she was listed for total abdominal hysterectomy with right sided salpingoophorectomy. Post operatively, sebaceous material was aspirated from the 5x6 cm right sided ovarian cyst.

## PATHOLOGICAL FINDINGS

A 5cm right ovarian cyst with attached fallopian tube and a separately sent uterus with cervix were received in formalin. Cut section of the ovarian cyst showed yellowish pultaceous material along with few hair and areas of hemorrhage. Few cystic spaces filled with yellow fluid were identified. The uterus showed an intramural fibroid and the fallopian tube was unremarkable.

Microscopic examination of the ovarian cyst showed a cyst

lined by stratified squamous epithelium with underlying dermal appendages and congested blood vessels. Multiple cystic spaces of varying sizes lined by foam cells and multinucleate foreign body type giant cells were seen.

A diagnosis of mature cystic teratoma with Pneumatosis cystoides-like appearance was made on histopathological examination.

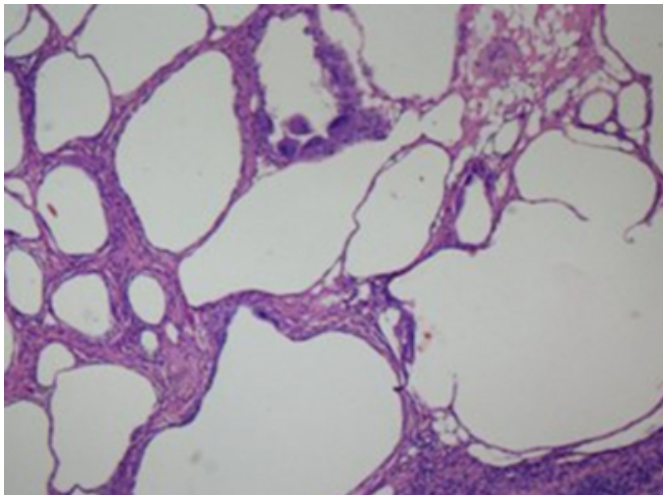
## Figure 1

Fig 1: Gross picture of mature cystic teratoma with multicystic appearance



**Figure 2**

Fig 2: Multiple cysts of varying size seen surrounded by foamy histiocytes & foreign body giant cells (H&E x100)



**DISCUSSION**

Mature cystic teratoma (dermoid cysts), accounts for approximately 25% of all ovarian tumors. They are unilateral in 88% of the cases and present with symptoms related to the mass lesion. They contain a varied mixture of ectodermal, mesodermal and endodermal structures distributed in an organized fashion. <sup>1</sup> The first description of pneumatosis cystoides-like appearance in a mature cystic teratoma was given by Maudsley and Zakhour in a 29 year old primigravida in 1988. <sup>2</sup> Subsequently, two case studies with 13 and 7 cases respectively have been reported. <sup>3,4</sup> These multicystic structures have been variably called as sieve-like areas and Pneumatosis cystoides-like appearances by various authors. <sup>2,3,4</sup> The multicystic structures can occur in any age group, but they are generally found in tumors from older patients. <sup>5</sup> Maudsley and Zakhour suggested these cysts to be gas cysts, as on routine staining and immunohistochemistry neither cyst contents, nor endothelial lining were identified. <sup>2</sup>

Later, Rubin and Papadaki studied the histological, immunohistochemical and ultrastructural features of this multicystic appearance in 13 cases with mature cystic teratoma. On histological examination, the cysts were found to be lined partly or completely by macrophages and foreign body type giant cells. Special stains with oil red-O showed strong staining of the cyst contents and a weaker staining of the macrophage cytoplasm. Immunohistochemical study established the macrophagic and endothelial nature of the lining epithelium because of the cytoplasmic positivity for

Mac387 and factor VIII- related antigen. The cystic spaces were found to be lined by predominantly macrophages and in occasional cases by endothelial cells or lipid laden polyhedral cells, on ultrastructural examination. The authors concluded that these multicystic structures might evolve due to the focal destruction of the lining epithelium followed by infiltration of the oleous material, which leads to a granulomatous reaction leaving an almost acellular multicystic sieve-like structure without any lining predominantly. <sup>3</sup>

Canzonieri et al also described similar findings in the seven cases studied by them. On immunohistochemical analysis, the macrophagic and endothelial nature of the lining cells was confirmed by positivity for KP1, KIM 6 and FVIII-related antigen. The authors suggested that the sieve like areas may be due to a granulomatous reaction by extravasated sebum and lipids or due to dilatation of lymphatic vessels. <sup>4</sup>

Various other authors also describe the multicystic structures as a reactive phenomenon due to hair, sebum or degenerated sebaceous glands leading to a granulomatous response.

In the present case, the cystic structures lined by macrophages and multinucleated foreign body type giant cells may also be a granulomatous response to the pultaceous material as evidenced in the past studies.

Mature cystic teratoma with Pneumatosis cystoides-like appearance is a diagnostic curiosity. Thus, we have presented here, a rarely reported but commonly found feature of a common tumour.

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