

# Deciduoid Mesothelioma of Peritoneum Masquerading as Peritoneal Carcinomatosis.

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## Research Article

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# Abstract

Mesothelioma is an insidious neoplasm that develops from mesothelial cells. About 80 % of mesotheliomas originate in the pleural cavity. Other sites where it has been reported are the peritoneal cavity, tunica vaginalis, and the pericardium. A 45-year-old female presented with complaints of abdominal distention and pain for three months. Physical examination revealed signs of wasting of the appendicular and axial skeleton muscles, loss of subcutaneous fat, and hollowing of the eye sockets. There was pitting edema in the bilateral lower limbs. Per abdomen, examination revealed abdominal distension with umbilicus in the midline. On palpation, gross ascites was present, and no organomegaly, definitive mass, or lump was palpable. On percussion, the dull note was heard all over the abdomen, and fluid thrill was appreciated. The ascitic fluid examination revealed the presence of atypical cells. Omentectomy was done and sent for histopathological examination.

The specimen of omentectomy was in multiple fragments and measured 17x16x3cm. Few of the fragments were nodular, soft to firm. On serial slicing, the cut section was gray-white with areas of necrosis. Microscopic examination showed sheets of malignant cells. These tumor cells were immunoreactive to EMA, Cytokeratin, Vimentin, Calretinin, WT-1, and D2-40 and immunonegative to Desmin (highlighting only the entrapped reactive mesothelial cells), Inhibin, BerEP4, TTF-1, CD 68, Napsin, ER, CEA, CDX2, PR, PAX-8, and SALL4. Ki 67 labelling index was 15%. The features were of Malignant Mesothelioma, Deciduoid variant. Deciduoid mesothelioma is a rare subtype with a poor prognosis. So, the mesothelioma should be distinguished from deciduositis.

## Introduction

Talerman et al. and Nascimento et al. in 1985 and 1994 first described the malignant deciduoid mesothelioma (MDM) in the peritoneum of a young female, and later it was also reported in pleura of elderly individuals. <sup>[1, 2, 3]</sup> Malignant deciduoid mesothelioma is a rare subtype of epithelioid mesothelioma which can mimic an exuberant ectopic decidual reaction.<sup>[2]</sup> To date, less than 40 cases have been reported in Pubmed indexed English literature. We herein report a case of MDM in a 45-year-old female who was misdiagnosed with Peritoneal Carcinomatosis.

## Case Report

A 45-year-old female presented with complaints of abdominal distention, constipation, decreased appetite, and pain for three months. There was a significant weight loss of approximately 15 kg in the past three months. There was no family history of malignancy, tuberculosis, and history of asbestos exposure. The general condition of the patient was low, with a performance status of ECOG 3. The patient had clinical signs of malignant cachexia, like wasting of the appendicular and axial skeleton muscles, loss of subcutaneous fat, and hollowing of the eye sockets. There was pitting edema in the bilateral lower limbs. Per abdomen, examination revealed distension of abdomen with umbilicus in the midline. No visible peristalsis or dilated veins were seen all over the abdomen. Hernial sites were normal. The gross ascites

was present on palpation, and no organomegaly, definitive mass, or lump was palpable. On percussion, the dull note was heard all over the abdomen, and the fluid thrill was appreciated. On auscultation, normal bowel sounds were heard. The ascitic tap was done, and ascitic fluid was sent for cytological examination, which revealed atypical cells. The omentectomy was done and sent for histopathological examination.

Omentectomy specimen was received in multiple fragments, and it measured 17x16x3cm [Fig. 1a]. Few of the fragments appeared nodular, soft to firm. On serial slicing, the cut section was grey, white with focal areas of necrosis. On microscopic examination, the multiple sections examined showed diffusely infiltrating tumor cells arranged in sheets. The cells were polygonal, mild to moderately pleomorphic, with round vesicular nuclei, conspicuous nucleoli, and a moderate amount of eosinophilic glassy to clear cytoplasm with sharp cellular outlines. Unremarkable, flat cuboidal mesothelial lining in strands and tubules were identified, entrapped between the malignant cells [Fig. 1b]. Few multinucleated and binucleate cells [Fig. 1c], along with large areas of palisading necrosis, were present [Fig. 1d], No granuloma was seen, and ZN stain for acid-fast bacilli was negative. PAS and PAS-D highlighted the presence of neutral mucin [Fig. 1e, 1f]. On immunohistochemistry, the tumor cells were immunoreactive for CK5/6, EMA, D2-40 [Fig. 1g, h, i] Calretinin, WT-1, and Vimentin [Fig. 2a, b, c] The tumor cells were negative for Desmin (highlighting only the entrapped reactive mesothelial cells), MOC31, CD 68 [Fig. 2d, e, f] BerEP4, CEA [Fig. 2g, h], inhibin, TTF-1, Napsin, ER, CDX2, PAX-8, and SALL4. The Ki 67 labelling index was 15% [Fig. 2i]. The features were of Malignant Deciduoid Mesothelioma.

## Discussion

Nascimento et al. described deciduoid peritoneal mesothelioma as a rare subtype which is unrelated etiologically to asbestos exposure.<sup>[2]</sup> Deciduoid peritoneal mesothelioma has been reported in elderly patients in pleura, pericardium, and the spermatic cord in association with asbestos exposure.<sup>[4, 5]</sup> The four major histological subtypes of mesothelioma are epithelioid, sarcomatoid, desmoplastic, and biphasic. Deciduoid mesothelioma is a rare subtype of epithelioid mesothelioma, in which a various range of histopathological patterns is seen.<sup>[6, 7]</sup> The deciduosis is frequently seen in the setting of pregnancy, premenarchal and postmenopausal status, which manifest as small discrete nodules or excrescences peritoneal surface, and the scarce case may present as pseudo tumoral lesions. Decidual cells strongly express CD10, inhibin, estrogen, and progesterone receptors and lack pan-cytokeratin and calretinin expression.<sup>[4, 8]</sup> The involvement of the peritoneal cavity and ascites contradicts the diagnosis of deciduosis.<sup>[9]</sup> Microscopically, in benign deciduosis, the cells are present among the fibro collagenous tissue, and cells have round nuclei, smaller and dark clumped chromatin, and moderate to plenty amount of cytoplasm. The mesothelioma has vesicular nuclei, fine chromatin, prominent nucleolus, and occasional mitotic figures.<sup>[1, 3, 4]</sup> Ultrastructurally, the deciduoid mesotheliomas can reveal long, branching microvilli, intracytoplasmic lumina, and filaments, which results in glassy cytoplasm in light microscopy. The presence of intracytoplasmic glycogen can give rise to cytoplasm clearing, as reported by Orosez et al.<sup>[10, 11]</sup> The WHO and International Mesothelioma Panel recommend using at least two

markers, carcinoma, and mesothelioma, in addition to a pan-cytokeratin. [7, 12] An immunohistochemical panel is essential for accurate diagnosis and differential diagnosis. The immunohistochemical markers panel depends on the histologic type of mesothelioma, the tumour's location, and the type of tumor being considered in the differential diagnosis. [12] A single marker is not diagnostic of mesothelioma. The positive immunohistochemical panel includes calretinin, cytokeratin 5/6, WT-1, D2-40, and vimentin. The tumor cells are negative to MOC 31, CEA, and BerEP4. [7] PAX8 is a very sensitive and specific marker for peritoneal Mullerian tumors and is negative to reactive and malignant mesothelial cells. [13] The mean survival time reported in the previously published cases was less than eight months. However, our patient survived for three months. [1, 2] Fluorescence in situ hybridization helps identify the band aberrations on 1p, 12q, 17, 8q, 19, and 20, these chromosomes as these are mostly chromosomal gains that help to distinguish the reactive mesothelial cells and mesothelioma. Dominak et al has reported 2 balanced translocations: t(1p;12q) and t(16p;16p). [15] A combination chemotherapy regimen of an antifolate (pemetrexed and raltitrexed) and a platinum-based (cisplatin) agent has shown a median survival of more than a year. Because of the peritoneal confinement of MM and the low occurrence of metastasis, perioperative intraperitoneal chemotherapy, and cytoreductive surgery has been considered as a curative treatment option with an overall 5-year survival rate of 29–63%. [5, 14] MDM behaves more aggressively than conventional epithelioid mesotheliomas. However, in many cases, this has not been proved to have a worse outcome. However, when it is showing the atypical mitotic figures and high mitotic rate, it may show a very aggressive clinical course. Hence, grading and subtyping may influence the patient treatment and outcome. [14]

## Conclusion

Malignant deciduoid mesothelioma must be considered in the differential diagnosis of peritoneal mass, and it must be distinguished from deciduosis. The differential of mesotheliomas varies with histological type and tumor location. The features of atypia, atypical mitosis, and the wide immunohistochemical panel help to clinch the diagnosis.

## Declarations

**i. Funding:** Nil

**ii. Conflicts of interest/Competing interests:** NIL

**iii. Ethics approval:** Waived

**iv. Consent to participate:** Consent taken

**v. Consent for publication:** Consent taken

**vi. Availability of data and material (data transparency):** No new data generated, or the article describes entirely theoretical research. Data sharing not applicable to this article as no datasets were generated or analysed during the current study.

**vii. Code availability (software application or custom code):** NA

**viii. Authors' contributions**

	JYOTSNA NARESH BHARTI	JEEWAN RAM VISHNOI
Planning	√	
Conduct		√
Reporting	√	
Concepts & Design		√
Data acquisition		√
Data Interpretation	√	
Manuscript preparation, editing & review	√	

**Declaration of patient consent:** The authors certify that they have obtained all appropriate patient consent forms. The patient has given her consent for their images and other clinical information to be reported in the form. The patients understand that their names and initials will not be published, and due efforts will be made to conceal their identity.

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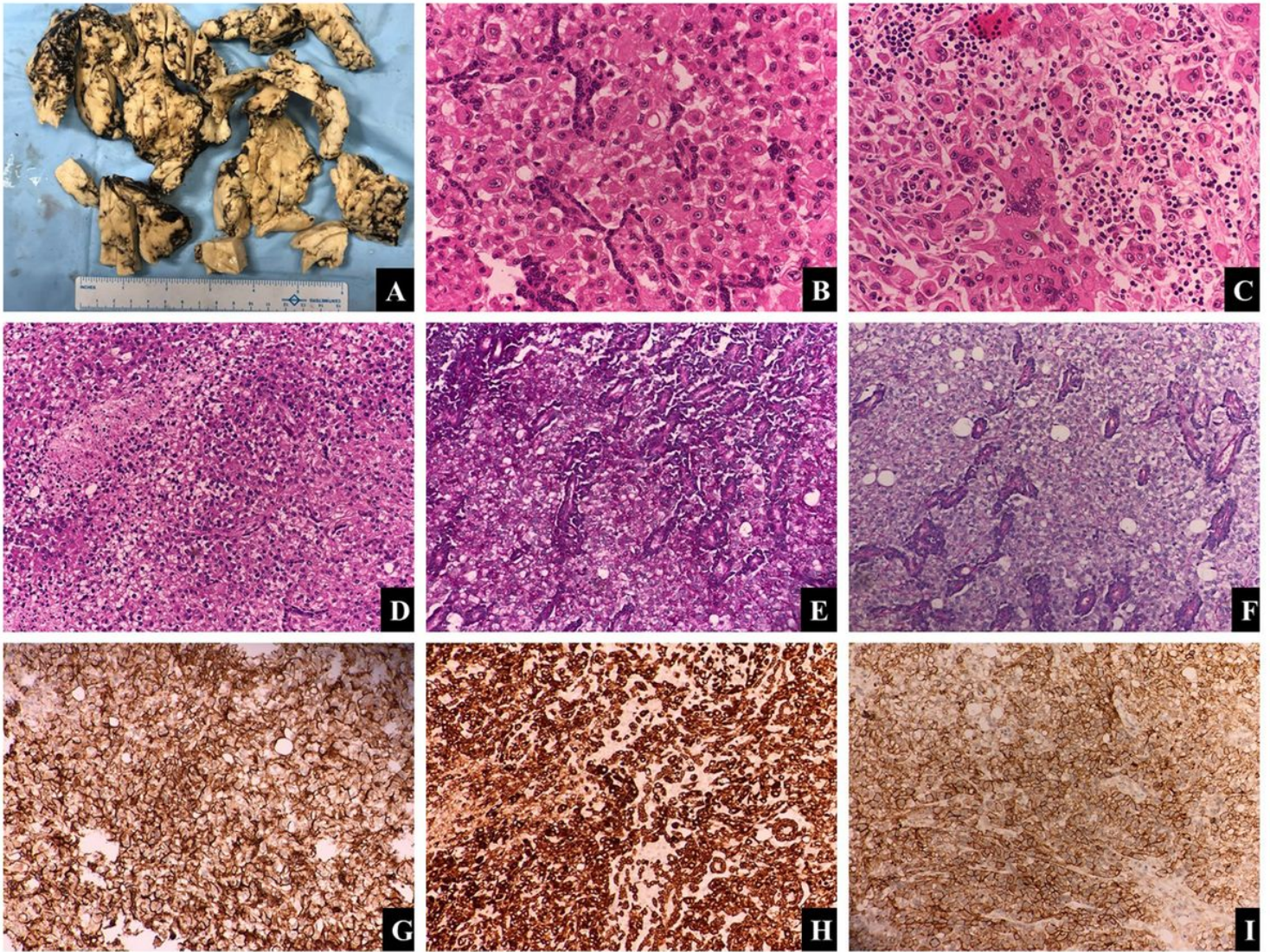
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## Figures

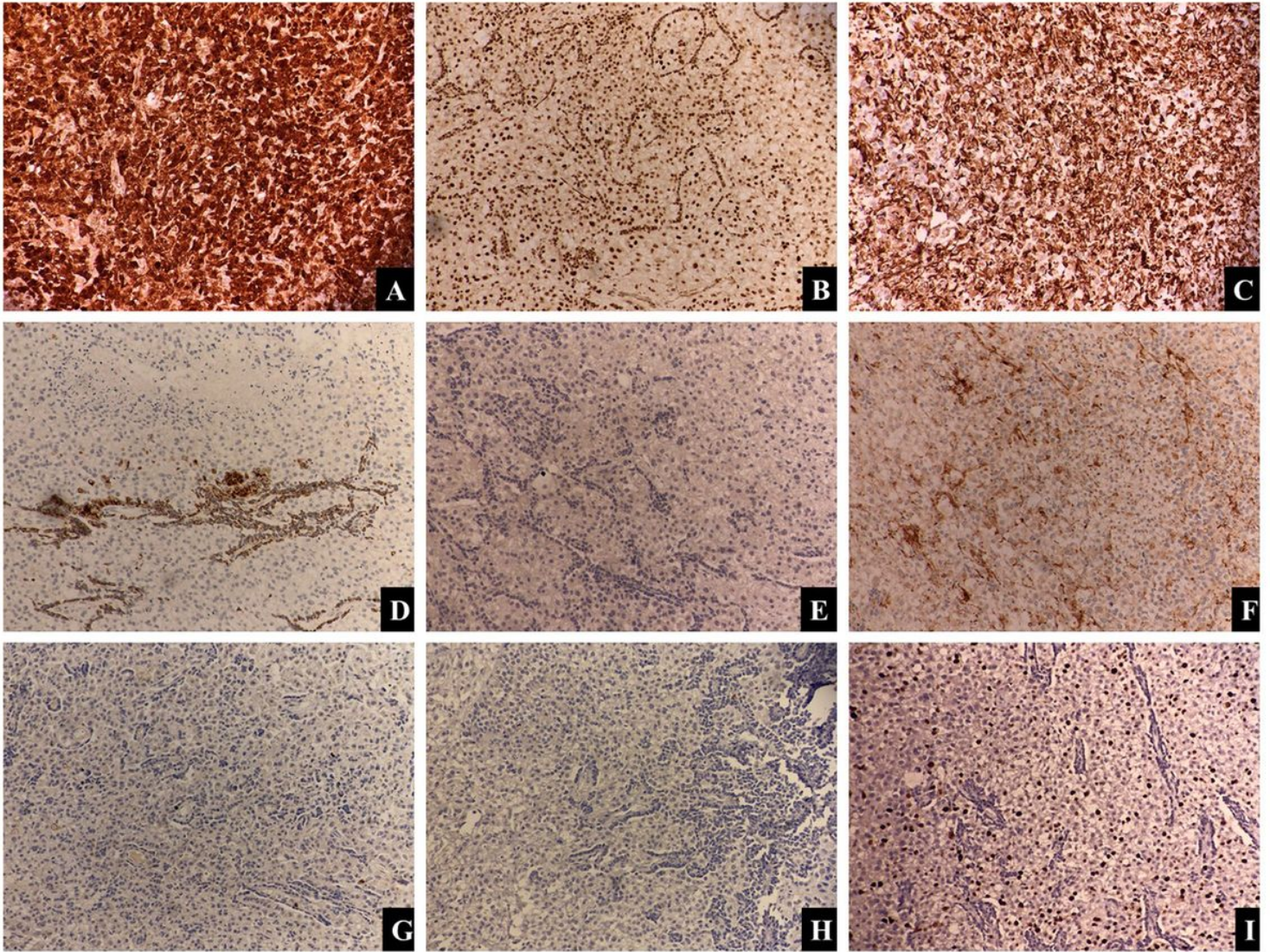




**Figure 1**

[Fig 1a] Omentectomy specimen in multiple fragments. [Fig 1b] Sheets of malignant cells with entrapped mesothelial cells. [HE 40X] [Fig 1c] Few multinucleated and binucleate cells. [HE 20X] [Fig 1d] Large areas of palisading necrosis. [HE 20X] [Fig1e, f] PAS and PAS-D highlighted the presence of neutral mucin. [HE 20X] [Fig1g, h, i] Tumor cells are immunoreactive to CK5/6, EMA, D2-40 Calretinin, WT-1, and Vimentin. [HE 20X]





**Figure 2**

[Fig 2a, b, c] Tumor cells are immunoreactive to Calretinin, WT-1, and Vimentin. [HE 20X] [Fig 2d, e, f] Tumor cells are negative to Desmin (highlighting only the entrapped reactive mesothelial cells), MOC31, CD 68. [HE 20X] [Fig 2g, h] Tumor cells are negative to BerEP4, CEA. [Fig 2i] Ki 67 labelling index is 15%.