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Case Report

Pleural malignant deciduoid mesothelioma: Case report of a rare variant of epithelioid mesothelioma

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ABSTRACT

Malignant deciduoid mesothelioma (MDM), a rare variant of epithelioid mesothelioma, accounts for less than 5% of mesotheliomas. The term deciduoid mesothelioma was introduced by Nascimento et al. in 1994 to describe a rare variant of epithelioid mesothelioma that bears a morphological resemblance to decidua or decidual-type changes. It has an unknown etiology, and the relation with asbestos exposure remains debatable. The presence of decidual-type cells often makes the diagnosis difficult in the small biopsy specimen, especially in the peritoneal biopsy of young female patients, as they can be misdiagnosed as normal decidualized tissue. The prognosis of deciduoid mesothelioma was considered poor, but now reports suggest that the prognosis depends on the grade of the tumor. Mesothelioma is more commonly seen in older men, but deciduoid mesothelioma most often occurs in young women and is initially thought to occur only in the peritoneum of young women. Later case reports of this rare variant occurring in the pleura, the pericardium, and the tunica vaginalis of older adults are also seen. Herein we report a case of a 60-year-old male patient diagnosed with high-grade malignant deciduoid pleural mesothelioma on small biopsy, treated with four cycles of neoadjuvant chemotherapy (Pemetrexed + Carboplatin + Bevacizumab), with partial favorable response to chemotherapy underwent pleurectomy. Following this, the patient was on maintenance chemotherapy; unfortunately, he had progressive disease with adrenal and lymph node metastasis.

1. Introduction

Pleural malignant deciduoid mesothelioma (PMDM) is a rare variant of pulmonary mesothelioma, representing < 5% of all mesotheliomas. This tumor is commonly seen in the peritoneum of young females and was first described by Talerman et al.[1] in 1985; however, the term deciduoid was first used by Nascimento[2] in 1994 as this rare variant is characterized by cytomorphologic features resembling decidualized tissue. The tumor was initially thought to behave more aggressively than other mesotheliomas, but now reports suggest that the aggressiveness depends upon the grade of the tumor rather than the variant.

2. Case Report

A 61-year-old gentleman with a history of covid infection six months back consulted the pulmonology department in our hospital with a persistent cough and breathlessness. He had no known comorbidities. CBNAAT was not detected in sputum.

His chest x-ray showed multiple opacities in the left lung field on evaluation. A CT Thorax was then done, which showed a diffuse irregular nodular pleural thickening on the left involving the visceral, parietal, diaphragmatic, and mediastinal pleura with mild diffuse heterogeneous

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enhancement. A differential diagnosis of Mesothelioma / Metastasis was considered on radiology.

FDG PET-CT was done to assess the extent of the disease. Metabolically active diffuse nodular pleural thickening involving the whole left lung- costal, mediastinal, and diaphragmatic pleura was identified, which warranted a histopathological correlation to rule in possible primary pleural malignancy (Figure 1).

A true cut biopsy of the pleura was done for a definite diagnosis, which showed an infiltrating neoplasm composed of polygonal and round epithelioid cells with well-defined cell borders, abundant glassy eosinophilic cytoplasm, mildly irregular nuclei, coarse chromatin, and occasional prominent nucleoli (Figure 2). Mitosis, 16-18/10hpf, was seen. A spotty area of necrosis was also seen.

Immunohistochemistry showed a positive staining for Calretinin (Figure 3), WT1, and EMA, and negative staining for CK7, Napsin, TTF1, desmin. Thus light microscopy and immunohistochemistry features were diagnostic of a high-grade malignant epithelioid mesothelioma, deciduoid type.

The patient was given four cycles of Pemetrexed + Carboplatin + Bevacizumab. A repeated CT showed a partial favorable response, following which he underwent left near total pleurectomy, which showed a ypT2Nx stage tumor of similar morphology as in small biopsy. The patient was continued with a maintenance chemotherapy regimen of Pemetrexed+Bevacizumab. But unfortunately, a repeat FDG Whole-body PET/CT scan showed progressive disease, with findings of metabolically active residual/ recurrent pleural thickening involving left mediastinal and diaphragmatic pleura. Mildly metabolic subcentimetric stable mediastinal lymph nodes, adrenal and mediastinal lymph node metastasis describe the aggressive nature of a high-grade Malignant deciduoid mesothelioma. The patient is currently on maintenance chemotherapy.

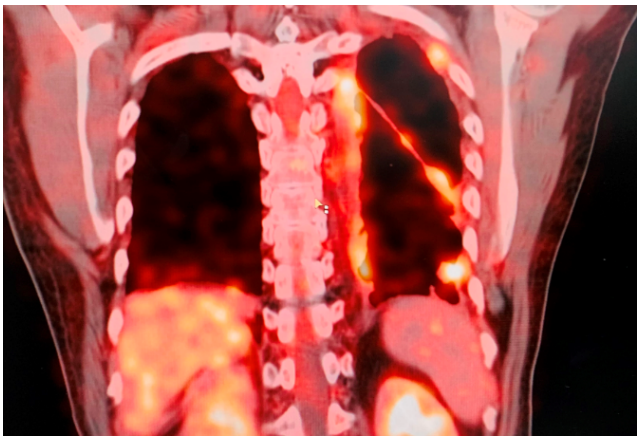

Fig. 1: FDG PET-CT showed a metabolically active diffuse nodular pleural thickening involving the whole left lung- costal, mediastinal, and diaphragmatic pleura.

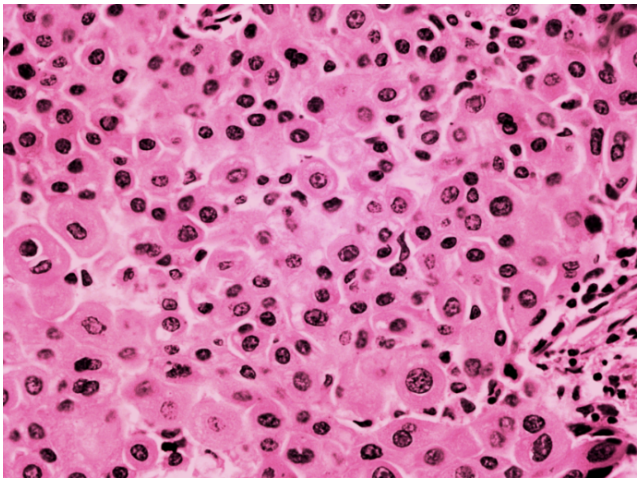

Fig. 2: Hematoxylin and eosin stained section in 40x objective showed polygonal and round epithelioid cells with well-defined cell borders, abundant glassy eosinophilic cytoplasm, mildly irregular nuclei, coarse chromatin, and occasional prominent nucleoli.

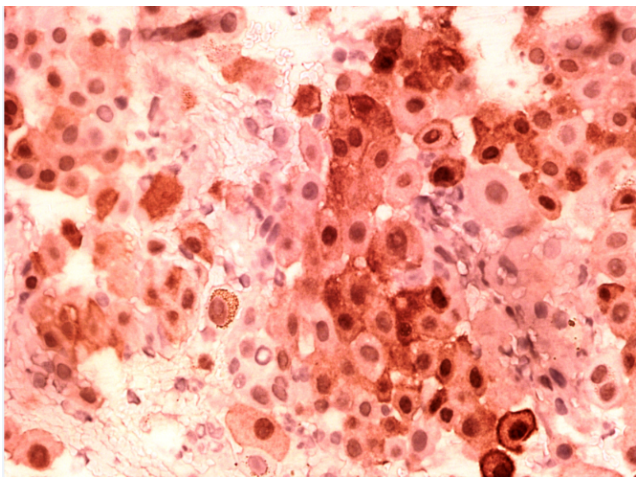

Fig. 3: Immunohistochemistry showed a positive cytoplasmic and nuclear staining for Calretinin.

3. Discussion

Malignant Mesothelioma (MM) is a rare, rapidly progressing, aggressive tumor of the pleura, peritoneum, tunica vaginalis, and pericardium. The first mention of a possible chest wall tumor was made in 1767 by Joseph Lieutaud, the founder of pathologic anatomy, in a study of 3,000 autopsies where he found two cases of “pleural tumors.” In 1819, René-Théophile-Hyacinthe Laennec, the French physician, suggested these tumors’ origin from the pleural cells. In 1908 Miller and Wynn first mentioned Peritoneal mesothelioma.[3] In 1985, Talerman et al.[1]. reported the first case of deciduoid mesothelioma in the peritoneum of a 13-year-old girl who had initially been

diagnosed with diffuse pseudotumoral deciduosis. The long latent period between onset and symptoms and the common/ nonspecific clinical presentation often delay the diagnosis. The histological examination and immunohistochemical analysis can only make a definite diagnosis.

Histologically, WHO has classified mesotheliomas into three major variants: epithelioid, sarcomatoid, and mixed or biphasic. Epithelioid mesothelioma commonly presents as a tubulopapillary or solid pattern but can also show wide morphological appearances, including adenoid cystic,signet-ring, rhabdoid, oncocytic, clear cell, small cell, glomeruloid, pleomorphic, and deciduoid.[4,5]

3.1. Microscopy

Deciduoid mesothelioma is characterized by large, polygonal, or round cells with a distinct cell border arranged diffusely in solid nests and trabecular patterns. The cells have abundant eosinophilic cytoplasm large and vesicular nuclei with coarse chromatin and occcasional prominent nucleoli. It can also have foamy cells with clear cytoplasm and stroma with mucinous or myxoid areas. Nuclear pleomorphism, nuclear pseudo inclusions, multinucleated cells can also be seen. Mitotic index is variable. High mitotic activity and atypical mitoses are also observed[4–6].

3.2. Immunohistochemistry

Differentiation between malignant mesothelioma and reactive mesothelial proliferation is crucial in small biopsies due to limited surrounding stromal tissue. Immunohistochemistry of decicduoid variant is similar to other malignant mesotheliomas with the positivity of WT1, calretinin, CK5/6, pan CK, EMA, D2-40, mesothelin, GLUT-1. Foci of frank stromal invasion and positivity for EMA and p53 favored mesothelioma, whereas desmin immunoreactivity is more common in the reactive mesothelium. However, recent publications suggest a limited diagnostic value of older mesothelioma markers, including p53, desmin, EMA, and GLUT-1. Novel IHC markers BAP1 (BRCA-associated protein), MTAP, 5-hmC (5-hydroxymethylcytosine), EZH2 (enhancer of zeste homolog 2) are thought to be reliable. Nuclear BAP-1 loss is 100% specific for all mesothelioma, including biphasic mesothelioma, to rule out the reactive mesothelial proliferation.[7,8]

3.3. Electron microscopy

On ultrastructural examination, PMDM cells show characteristic mesothelial microvilli. Electron microscopy also reveals many intermediate filaments, which give a glassy eosinophilic appearance to the cytoplasm on hematoxylin-eosin slides. It also demonstrates the cytoplasmic nature of the nuclear pseudo inclusions.[4–7]

3.4. Cytogenetics

Molecular genetic analysis has revealed several key genetic alterations responsible for the development and progression of malignant mesothelioma. The cyclin-dependent kinase inhibitor 2A/alternative reading frame (CDKN2A/ARF), neurofibromatosis type 2 (NF2), and BRCA1-associated protein-1 (BAP1) genes are the most frequently mutated tumor suppressor genes detected in Malignant mesothelioma cells; the alterations of the latter two are relatively characteristic.[9] A recent article has also identified specific genetic abnormalities in pleural deciduoid mesotheliomas, the most frequent being chromosomal gains at 1p, 12q, 17, 8q, 19, and 20 and losses at 13q, 6q, and 9p.[10] Dominak et al.[11] reported the first case of translocation with two balanced translocations: t(1p;12q) and t(16p;16p).

3.5. Treatment

Malignant mesothelioma is highly refractory to conventional therapies even with a combination of aggressive surgical intervention and multimodality strategies, with cure remaining elusive. Treatment of patients with PMDM is the same as treatment of other malignant mesothelioma and is cytoreductive surgery combined with chemotherapy. Cisplatin and pemetrexed are considered first-line. Extensive debulking surgery with intraoperative chemotherapy is associated with better survival, especially patients without lymph node metastases. Radiation therapy is also considered part of trimodal therapy and plays a significant role in decreasing the chance of recurrence.[5–7]

3.6. Prognosis

The prognosis of malignant mesothelioma is mainly dependent on the grade and stage of the disease rather than the histological type. Some publications suggest that the nuclear grade predicts survival in epithelioid malignant pleural mesothelioma and that the necrosis can be a factor that further stratifies overall survival.[12]

4. Conclusions

We highlight the characteristic features of a particular variant of epithelioid mesothelioma. Pathologists should be aware of this rare malignancy, which can be high-grade and aggressive, as in our case. They should not be confused with other neoplastic and non-neoplastic conditions, especially on peritoneal biopsies of younger women, where it can be easily misdiagnosed as peritoneal deciduosis. In addition, more institutional-based studies on large sample sizes should be conducted to assess the prognostic factors, and an alternative scoring system based on histomorphological features should also be proposed to evaluate the aggressiveness of this tumor.

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None.

7. Conflict of Interest

None.

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