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Case report

Peritoneal deciduoid mesothelioma: a rare variant of epithelioid mesothelioma in a young man

T. Arudselvan, P. G. N. L. Chandrasiri, H. D. Wijesinghe, M.V.C. De Silva

Department of Pathology, Faculty of Medicine, University of Colombo, Sri Lanka

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Abstract

Deciduoid mesothelioma is a rare variant of malignant epithelioid mesothelioma. Diagnosis of deciduoid mesothelioma is very challenging especially in settings with limited resources. It morphologically overlaps with many other malignancies showing epithelioid morphology. We report a case of deciduoid mesothelioma occurring in a previously healthy 35-year-old-man. The clinical, morphological and the immunohistochemical features that aid in its diagnosis are discussed.

Key words: mesothelioma, deciduoid, epithelioid mesothelioma, pathology

Introduction

Mesothelioma is a malignant tumour of the mesothelial lining that occurs in the pleura and less frequently in the peritoneum and pericardium. Mesothelioma is classified morphologically as epithelioid mesothelioma, sarcomatous mesothelioma and biphasic mesothelioma (1). Deciduoid mesothelioma is a rare variant of epithelioid mesothelioma that shows the cytomorphology of decidualized tissue and demonstrates an aggressive clinical course (2). Initially deciduoid mesothelioma was thought to occur mainly in the peritoneum of young females with no history of asbestos exposure (2). However subsequent cases were reported occurring in both peritoneal and pleural cavities of men and women of all ages with or without exposure to asbestos (2,3,4). Morphologically deciduoid mesothelioma overlaps with many malignant and benign conditions. The diagnosis of deciduoid mesothelioma requires evidence of invasion and expression of mesothelial markers with exclusion of other differentials.

Case History

A previously healthy, 35-year-old tile worker presented with history of evening pyrexia of one month associated with dry cough, loss of weight and loss of appetite. The fever was low grade and accompanied with night sweats. His erythrocyte sedimentation rate was persistently high (>100mm in 1st hour). Screening for infective diseases including tuberculosis, anti-nuclear antibodies (ANA) and antineutrophil cytoplasmic antibodies (ANCA) were negative. Ultrasound scan abdomen was normal. The contrast tomography (CT) chest did not reveal any abnormality. He was started on antituberculosis drugs but showed a poor response. The contrast tomography (CT) abdomen and pelvis revealed enlarged upper mesenteric lymph nodes. Endoscopic examination of gastrointestinal tract and histological assessment of serial biopsies were normal. Multiple peritoneal and omental deposits were found on exploratory laparoscopic surgery.

Corresponding author: Dr Thadshajini Arudselvan
Department of Pathology , Faculty of Medicine,
University of Colombo, Sri Lanka
thadsha89@gmail.com

Biopsies from the omental and peritoneal deposits showed sheets and nodules of large polygonal cells infiltrating the fat. The constituent cells had abundant eosinophilic glassy cytoplasm with distinct cell borders and enlarged vesicular nuclei with prominent nucleoli. Multinucleated tumour cells were also present. Mitoses were not prominent. There was no necrosis. A lymphoplasmacytic infiltrate was present (Figure 1). The tumour cells showed diffuse positivity for calretinin, CK 5/6, AE1/AE3, P53 and EMA and focal positivity for CK 7 (Figure 1). The cells were negative for desmin, CK 20, melan A and HepPar 1.

A diagnosis of epithelioid mesothelioma was made based on the morphological and immunohistochemistry findings. The patient was transferred to the National Cancer Institute for further chemotherapy and follow up.

Discussion

Mesothelioma is a malignant neoplasm of mesothelial origin (1). The subtypes of mesothelioma are epithelioid mesothelioma, sarcomatous mesothelioma and biphasic mesothelioma. Deciduoid mesothelioma is a rare type of epithelioid mesothelioma which resembles decidua morphologically. It was first described in 1945 by Talerman et al who described a case of diffuse pseudo-tumoral deciduosis in a 13-year-old girl (5).The features described in this case were compatible with the features of deciduoid mesothelioma.

The term of deciduoid mesothelioma was first used in 1994 (6). Initially it was thought that deciduoid mesothelioma occurred exclusively in the peritoneum of young females with no history of asbestos exposure (6). In the later literature, cases have been reported in the pleura and peritoneum of both males and females of a wider age range both with and without a history of exposure to asbestos (2). According to the literature around 50% of the deciduoid mesothelioma occurs in the peritoneum with a female predominance (7). A few cases have been reported in the pericardial cavity (8). In contrast, classic epithelioid mesothelioma occurs mostly in elderly males with history of asbestos exposure (5,9).

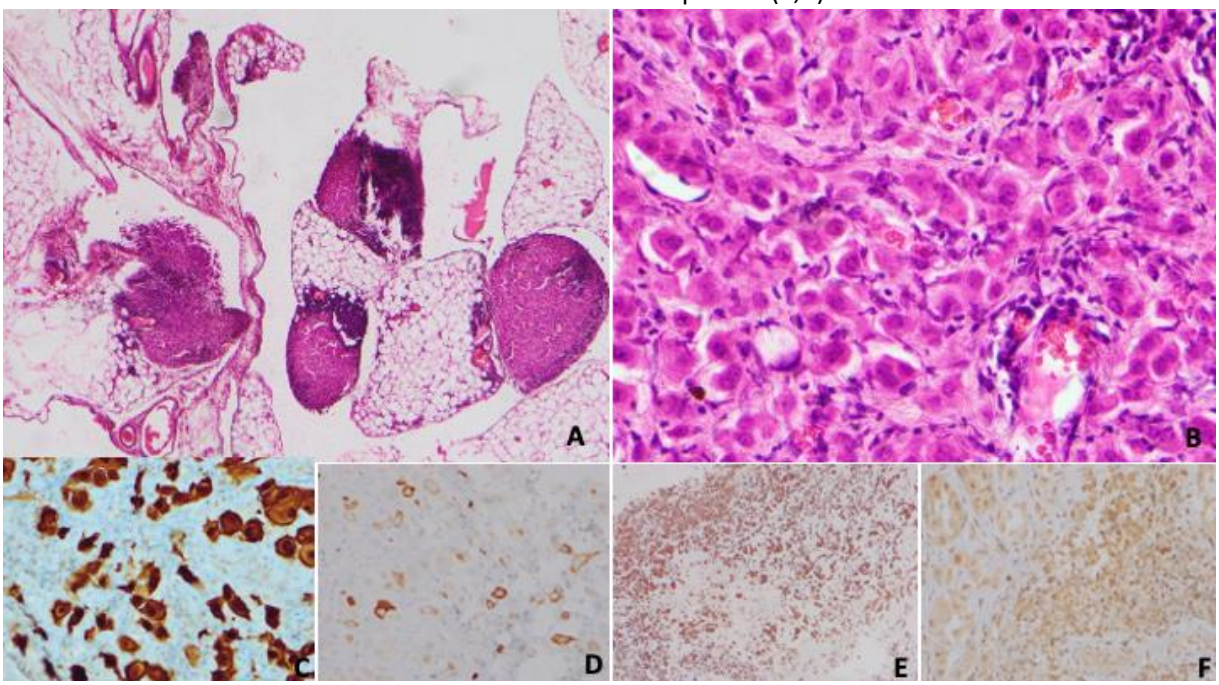

Figure 1: A): Tumour nodules infiltrating fat (Haematoxylin and eosin stain (H&E) ×40). B)The tumour cells had abundant eosinophilic glassy cytoplasm and enlarged vesicular nuclei with prominent nucleoli (H&Ex400). The tumour cells showed C) nuclear and cytoplasmic positivity for calretinin (Calretinin ×400), D) patchy positivity for cytokeratin 5/6(CK 5/6 ×400), E)diffuse cytoplasmic positivity for Pan CK(AE1/AE3 ×100) and F) nuclear positivity for P53 (p53 ×200).

Histologically deciduoid mesothelioma is composed of large oval to polygonal cells with abundant glassy eosinophilic cytoplasm, distinct cell borders and round vesicular nuclei with prominent nucleoli (4, 10). These cells are arranged in sheets, nests, trabeculae and papillaroid structures. Nuclear atypia and mitotic activity are variable (7). The cells may be binucleated or multinucleated and contain pseudoinclusions. This case showed most of these typical morphological features.

The diagnosis of deciduoid mesothelioma is difficult and a panel of immunohistochemical markers is essential for an accurate diagnosis. The choice of markers varies according to the location and the morphology of the tumour. On electron microscopy the cells of deciduoid mesothelioma show characteristic microvilli and many cytoplasmic intermediate filaments which result in the abundant glassy eosinophilic cytoplasm on light microscopy (4).

The differential diagnosis of deciduoid mesthelioma includes, reactive mesothelial proliferation, decidualised tissue, and other tumours with epithelioid morphology including epithelioid haemangioendothelioma, rhabdomyosarcoma, sex cord-stromal tumours and metastatic deposits of hepatocellular carcinoma, melanoma, squamous cell carcinoma, adenocarcinoma of lung, breast and ovary, chromophobe renal cell carcinoma, adrenal cortical carcinoma and trophoblastic tumours (1,4).

Differentiation of deciduoid mesothelioma from reactive mesothelial proliferation is challenging, especially in the early stages. The presence of invasion of the underlying tissue is the most reliable criteria of malignancy(1,10,11). Other features supporting malignancy include full thickness involvement, expansile nodules, random variation in cellularity, architectural complexity and irregular distribution of vessels(1). Detection of deletions of p16/CDKN2A by fluorescence in situ hybridisation (FISH) or polymerase chain reaction (PCR) techniques and loss of BAP 1 by immunohistochemistry are helpful in diagnosing mesothelioma(1,11). Immunohistochemical markers for epithelial membrane antigen (EMA), p53, GLUT1 and IMP3 show limited value as they are less sensitive and specific. Mesothelial markers are also expressed in other malignancies. For example WT1 can be expressed in breast and ovarian serous carcinoma and CK5/6 is positive in squamous cell carcinoma. Therefore, distinguishing mesothelioma from other carcinomas warrants the use of a minimum of two mesothelial markers and two carcinoma markers in addition to pancytokeratin (1,4). The selection of carcinoma marker depends on the differentials based on the morphological features and site of the tumour. Commonly used mesothelial markers are calretinin, CK5/6, D2-40, and WT1. Mesotheliomas are positive for thrombomodulin, HBME1, CK 7 and pancytokeratins. The common carcinoma markers used to differentiate carcinoma from mesothelioma are claudin-4, BerEP4, B72.3, MOC31, CEA, CD15 and BG8 (1) which are negative in mesothelioma. Mesothelioma is also negative for CK 20, ER, PR and HepPar 1.

Deposits of renal carcinoma can be differentiated from mesothelioma using PAX 8 which stains positive in renal cell carcinoma and negative in mesothelioma (2,7). Carcinomas of lung primary do not express WT1 or calretinin and mesotheliomas are negative for TTF1.

Melanomas can be excluded by negative staining for melanoma markers like HMB45 and Melan A which are negative in mesothelioma.

Decidualised tissue is an important differential diagnosis particularly in females in the reproductive age group. Decidualised tissue shows positivity for PR, ER and inhibin which are negative in deciduoid mesothelioma (4). Another condition occurring in women, confused with deciduoid mesothelioma is trophoblastic neoplasms. Usually, they occur after pregnancy and express HCG. Immunohistochemistry for HCG can be used to differentiate the two.

The diagnosis in this case was based on the distinct cytomorphological features, evidence of invasion by the presence of fat infiltration and expression of mesothelial cell markers on immunohistochemistry.

Table 1: Immunohistochemical markers useful in the differential diagnosis of deciduoid mesothelioma

	Deciduoid mesothelioma	Reactive mesothelial proliferation	Adenocarcinoma	Melanoma	Rhabdomyosarcoma	Present case
WT1	+	+	-	-	-	+
Calretinin	+	+	-	-	-	+
CK 5/6	+	+	-	-	-	+
D2-40	+	+	-	-	-	NA*
BerEP4	-	-	+	-	-	NA*
Claudin 4	-	-	+	-	-	NA*
MOC31	-	-	+		-	NA*
B72.3	-	-	+	-	-	NA*
CK 7	+	+	+/-	-	-	+
PanCK	+	+	+	-	-	+
EMA	+	+	+	-	-	+
TTF1	-	-	+/-	-	-	Not done
CK20	-	-	+/-	-	-	-
HepPar1	-	-	-/+	-	-	-
Melan A	-	-	-	+	-	-
HMB45	-	-	-	+	-	Not done
P53	+/-	-			-	+
Desmin	-/+	+/-	-	-	+	-
BAP 1	Loss	Expressed				NA*

**NA – Not available*

The morphological and immunohistochemical profile helped to differentiate to exclude the differential diagnoses of other malignancies and reactive mesothelial hyperplasia and establish the diagnosis of mesothelioma (Table 1). The differentials considered in our case based on the morphological and clinical findings were reactive mesothelial proliferation and deposits from hepatocellular carcinoma, gastrointestinal tract carcinoma and melanoma. Our case showed positive staining for pancytokeratin (AE1/AE3), mesothelial markers (CK5/6, calretinin) CK7, EMA and P53. It was negative for CK 20, melan A, desmin and HepPar 1 which helped to exclude colorectal carcinoma, melanoma and hepatocellular carcinoma. The positivity for two mesothelial markers (calretinin/CK5/6) favoured a mesothelial lesion over a carcinoma and presence of fat infiltration accompanied by positivity for EMA and p53 and negativity for desmin favoured the diagnosis of mesothelioma over a reactive mesothelial proliferation. Immunohistochemical stains like BerEP4 or MOC31 which help to distinguish mesothelial lesions from an adenocarcinoma, and molecular tests and BAP-1 immunohistochemistry which are useful in distinguishing mesothelioma from a mesothelial proliferation were not available in our centre.

The overall long term survival rate of mesothelioma patients is poor with median survival times of 19, 13 and eight months in patients with epithelioid, biphasic and sarcomatoid tumours respectively (1). The epithelioid subtype shows relatively longer survival duration compared to the other subtypes (1). However, the aggressiveness of the clinical course varies in some patterns of epithelioid mesothelioma. Studies have shown a mean survival time of 19 months in the deciduoid type of epithelioid mesothelioma (2). However, whether the prognosis is better or worse compared to the overall prognosis of mesothelioma or the epithelioid subtype is not clear. The presence of high nuclear grade (nuclear atypia and increased mitotic count) is an independent poor prognostic factor for mesothelioma irrespective of the morphological subtype/pattern (1). The variation in prognosis reported in deciduoid mesothelioma could be due to the existence of a high-grade subgroup that presents highly

aggressive clinical behaviour (2). Therefore, when a high-grade deciduoid mesothelioma is present, it should be reported as it can significantly affect prognosis and treatment.

Conclusion

Although deciduoid mesothelioma is rare, pathologists should be aware of this entity as it can be mimicked by many other common malignant and benign conditions. Given its rarity, other potential differential diagnoses must be thoroughly ruled out. Accurate diagnosis relies heavily on the careful selection of an appropriate immunohistochemical panel. Molecular tests are also available to aid in diagnosis.

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