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Mesothelioma of Anterior Abdominal Wall- Biphasic Variant A Rare Entity

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Abstract: Mesothelioma is a malignant tumor arising from mesothelial lining of pleura, peritoneum, pericardium, and tunica vaginalis. Visceral pleura are the most common location for the malignancy, followed by peritoneum. It is a rare malignancy which is diagnosed at advanced stage with very poor prognosis. Here we report a case of biphasic variant of malignant mesothelioma of anterior abdominal wall involving peritoneum, rectus muscle and momentum diagnosed with the help of histopathology and immunohistochemistry.
Keywords: Anterior Abdominal Wall tumour, malignant mesothelioma, Calretinin, WT1

INTRODUCTION

Mesothelioma is a malignant tumour arising from mesothelial lining of pleura, peritoneum, pericardium, and tunica vaginalis. It is a rare malignancy which is diagnosed at advanced stage with very poor prognosis. It is associated with exposure to asbestos for a long time among construction workers. This case is presented for it biphasic variant a rare entity.

CASE REPORT

77 year male patient who was a construction worker by profession came with complaints of painful swelling with burning sensation present over the upper abdomen for 1 month.

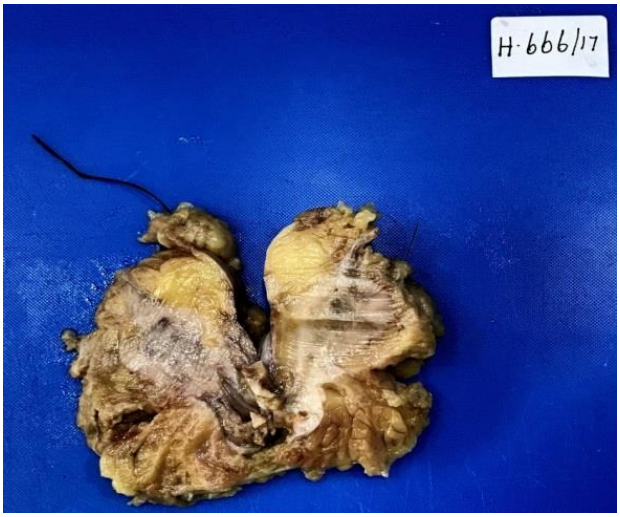
O/E- 6 x 5 cm swelling was seen over the upper abdomen in the epigastric region. The swelling was firm in consistency and immobile, not trans-illuminant and skin over the swelling was pinch able.

Radiological examination: A large soft tissue density lesion with lobulated margins and heterogeneous enhancement measuring 6.0 x 4.3 x 7.6 cm was seen in the anterior abdominal wall in the region of rectus sheath and rectus abdominis muscle in the epigastric region. Part of the lesion was seen extending into the peritoneum and was abutting the transverse colon. Multiple soft tissue density nodules were seen along the anterior wall of the adjacent transverse colon. Omentum showed nodules and possibilities of nodes or peritoneal extension or deposits were considered. Radiologically a diagnosis suspicious of desmoids tumour was given.

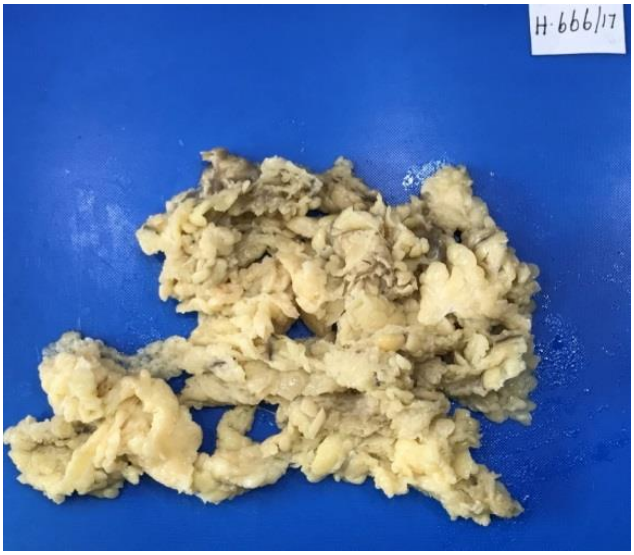
Intra-operative findings: Tumour was seen adherent to posterior of the falciform ligaments and momentum. Wide local excision of the mass with portion of omentectomy in the involved site was done.

Gross Findings

Received soft tissue mass with surrounding fibro-fatty tissue measuring 10.5 x 9.5 x 5.5 cm. The cut section of the mass showed a lesion of 6.5 x 5 x 5 cm. The lesion was solid, gray white and showed irregular, infiltrating margin. Omentum showed multiple gray white nodules about seven in number ranging in size from 0.5 cm to 5 cm.



Fig–1: The cut section of the tumour showing predominantly solid, gray-white areas with irregular margins



Fig–2: The omentum studded with tumour nodules

Microscopic Findings

Multiple sections studied show a cellular neoplasm with spindle cells in interlacing bundles and fascicles with elongated nuclei and prominent nucleoli. There was presence of oval to polygonal cells with eosinophilic cytoplasm and hyperchromatic nuclei. Infiltration into surrounding tissues seen with areas of hyalinization and necrosis. There is dense collagenized tissue separated by atypical cells and also the presence of microcystic spaces. Scattered lymphocytes are seen surrounding the stroma. Immunomarkers Calretinin and WT1 were done which proved to be positive. Hence a diagnosis of mesothelioma was made.

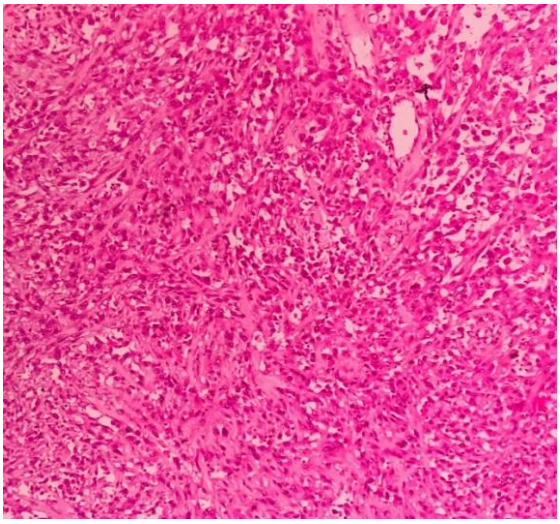


Fig-3: Low Power view (x 10 magnification) H&E stain showing oval to polygonal cells with eosinophilic cytoplasm and hyperchromatic nuclei, along with spindle cells in interlacing bundles and fascicles with elongated nuclei

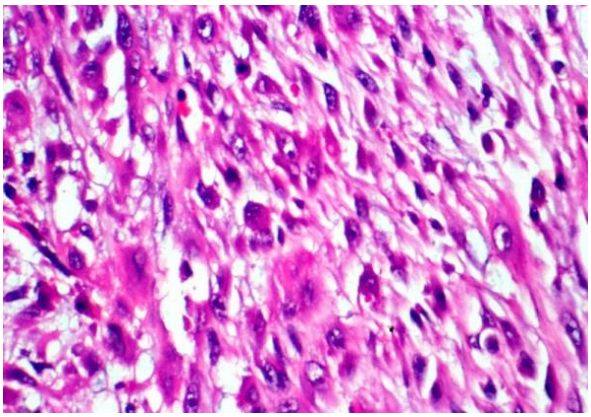


Fig-4: High power view (x 40 magnification) H&E stain showing oval to polygonal cells with eosinophilic cytoplasm and hyperchromatic nuclei along with spindle cells in interlacing bundles and fascicles with elongated nuclei and prominent nucleoli

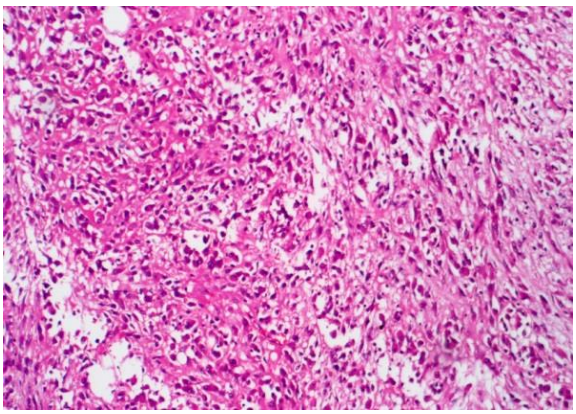


Fig-5: Low Power view (x 10 magnification) H&E stain showing oval to polygonal cells with eosinophilic cytoplasm and hyperchromatic nuclei

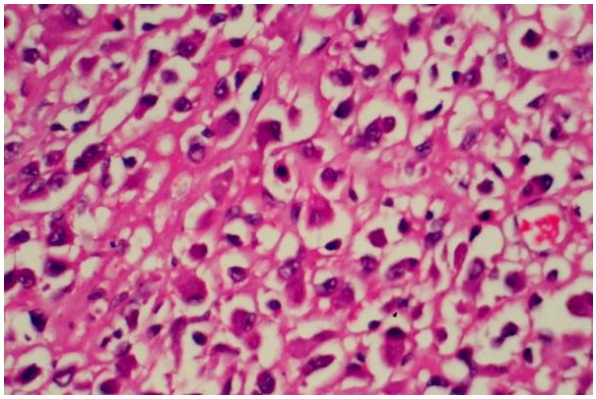


Fig-6: High power view (x 40 magnification) H&E stain showing oval to polygonal cells with eosinophilic cytoplasm and hyperchromatic nuclei

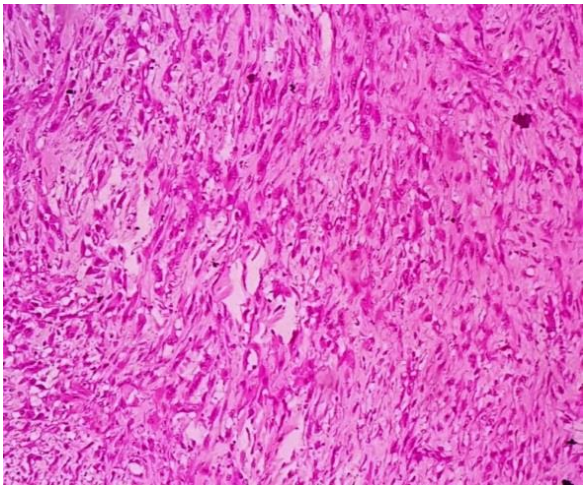


Fig-7: Low Power view (x 10 magnification) H&E stain showing spindle cells in interlacing bundles and fascicles with elongated nuclei

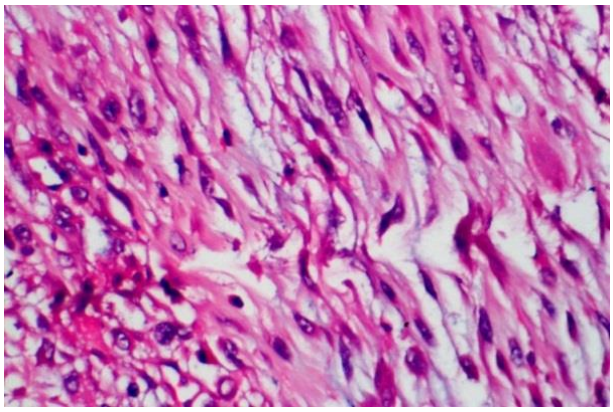


Fig-8: High power view (x 40 magnification) H&E stain showing spindle cells in fascicles with elongated and hyperchromatic nuclei

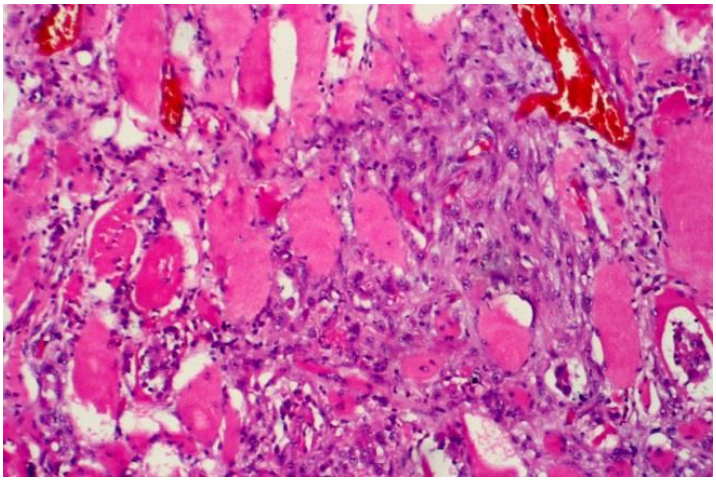


Fig-9: Low power view (x 10 magnification) H&E stain showing infiltration of the tumour cells into the muscle bundles

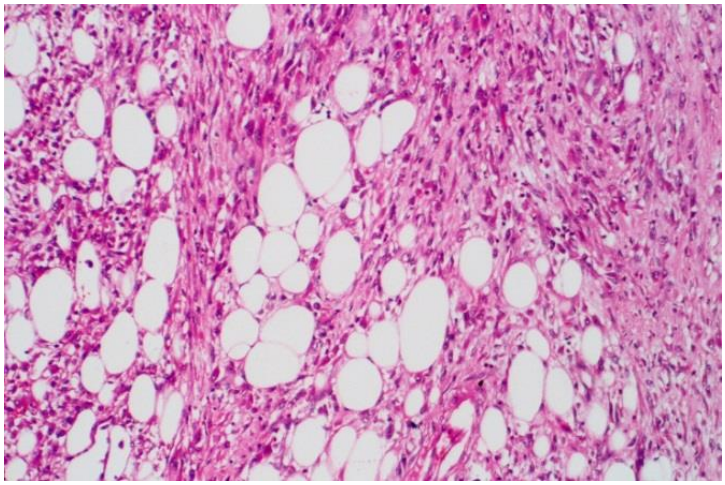


Fig-10: Low power view (x 10 magnification) H&E stain showing infiltration of the tumour cells into the fatty tissue

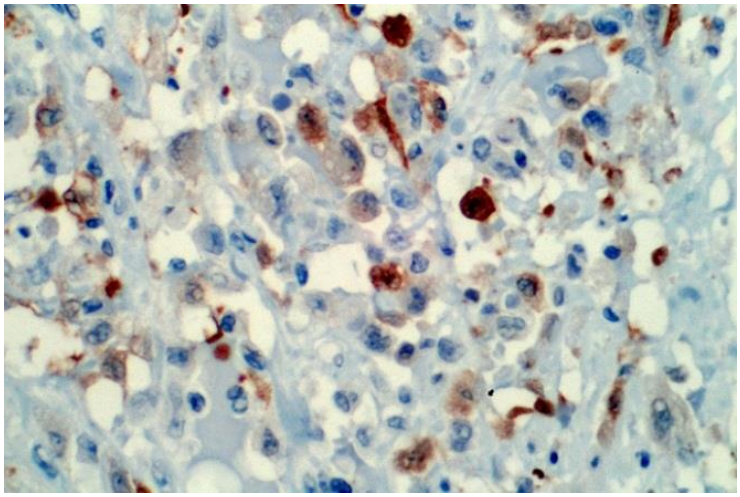
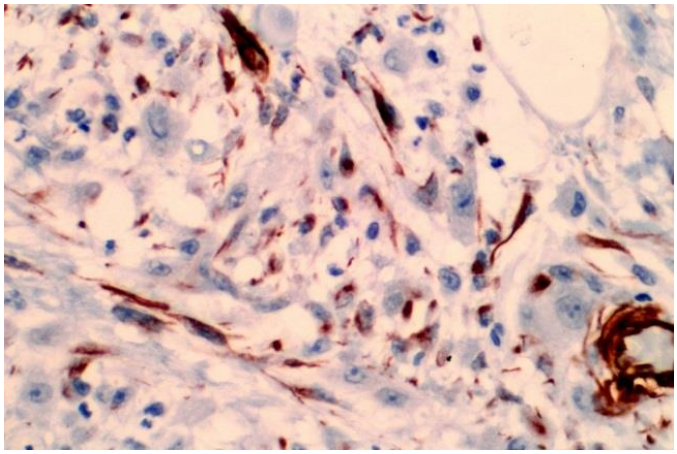
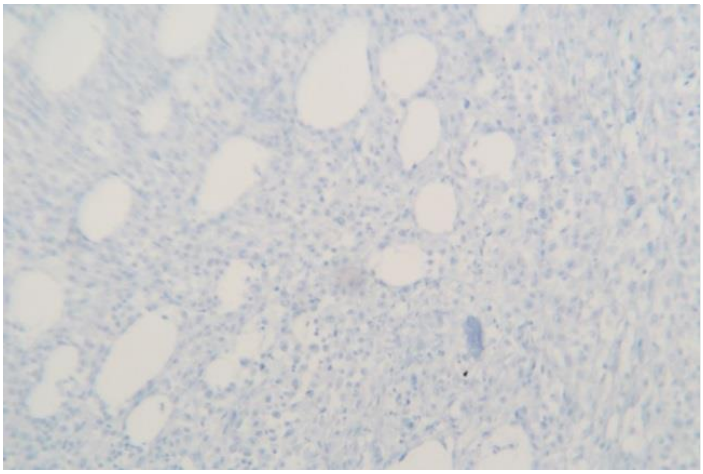


Fig-11: Calretinin immunostaining shows focal cytoplasmic and nuclear immunoreactivity



Fig−12: WT1 immunostaining shows focal cytoplasmic and nuclear immunoreactivity



Fig−13: Tumour cells were negative for Desmin immunomarker

DISCUSSION

Mesotheliomas are rare neoplasms arising from the mesothelial lining of pleura, pericardium, peritoneum, and tunica vaginalis. They occur in adults above 50 years of age. Males are three times more commonly affected than females [1].Environmental pollutants, particularly asbestos exposure is one of the aetiologic factors for the development of mesothelioma. Exposure towards asbestos is often more than 20 years. Other risk factors associated are radiation, minerals like mica and thorium exposure. The population associated with asbestos exposure range from construction workers, electricians and painters [1]. The highest rates are reported in United Kingdom [2]. In India, mesothelioma cases have been reported only in four of twenty five registries with 0.05-0.08 per 100,000 among men and 0.05-0.1 among women [3]. The pathogenesis of mesothelioma includes a hypothesis that asbestos fibers preferentially gather near the mesothelial cell layer and generate reactive oxygen species that causes DNA damage and mutations. Mutation of BAP1 gene which encodes a tumour suppressor involved in DNA repair has a high incidence of mesotheliomas [4].

The diagnosis of malignant mesothelioma is very difficult and it depends on features such as 1. History of asbestos exposure, 2. Clinical signs and symptoms, 3. Radiography, 4. Histopathological examination. Grossly, the tumours may appear as small whitish nodules to large fleshy masses invading the adjacent organs. Multifocal studding is one of the commonest features for malignant mesothelioma.

Microscopically, mesotheliomas have been classified as [5]

- Epithelioid -includes tubulo-papillary and deciduoid type.
- Sarcomatoid- includes desmoplastic, clear cell, and small cell type.
- Biphasic/Mixed.

Epithelioid mesothelioma is characterized by proliferation of round to polygonal tumour cells with abundant densely eosinophilic cytoplasm and bland nuclei that are devoid of mitotic activity. The sarcomatoid variant is composed of fascicular proliferation of spindle cells with oval nuclei, scant amphophilic cytoplasm, occasional prominent nucleoli

with more atypia & foci of necrosis than epithelioid counterpart. The features closely resemble malignant fibrous histiocytoma with elongated fascicles of tumour cells with abundant intercellular collagen. The desmoplastic variant is characterized by deposition of excess collagen.

The biphasic/mixed variant under consideration in the case has a combination of both easily recognizable epithelioid & sarcomatous components. It comprises 20-40 % of all mesothelioma cases. The immunomarkers used for diagnosis of mesothelial tumours are [6]:

Table–2: Sensitivity of markers used to diagnose mesothelioma

Marker	Sensitivity
Calretinin	>90 %
WT1	70 – 95 %
CK 5/6	75 – 100 %
D2-40	90 – 100 %

In our case, the patient who is a construction worker came with complaints of burning pain over the upper abdomen for 1 month. Ultrasound imaging gave a provisional diagnosis of desmoid tumour and wide local excision was done. Histological examination revealed possibilities of biphasic variant of mesothelioma, undifferentiated pleomorphic sarcoma and pleomorphic rhabdomyosarcoma. Undifferentiated pleomorphic sarcoma is a diagnosis of exclusion and is characterized by storiform to pleomorphic areas and is composed of pleomorphic spindle cells, tumour giant cells and histiocytic cells.

Pleomorphic Rhabdomyosarcoma [7] is a tumour of young adults characterized by presence of large round or pleomorphic cells arranged loosely in haphazardly oriented pattern. Cells with cross striations are less in number, but racket or tadpole shaped rhabdomyoblasts can be seen.

In this case, we established the diagnosis of mesothelioma by positive staining for Calretinin, WT1 and negative staining for desmin of tumour cells. Prognosis of the disease depends on the variant of the disease.

Table–1: Comparison of prognostic index

Variant	Time	Percentage survived	Time	Percentage survived
Epithelioid	2 years	50%	3 years	42%
Sarcomatoid	2 years	7.5%	3 years	No reports of people living beyond 25 months
Biphasic/Mixed	The tumour has a mixed prognosis which is poor than epithelioid variant and better than sarcomatoid variant.			

CONCLUSION:

The biphasic variant poses diagnostic dilemma to surgical pathologists and immunomarkers are mandatory for correct diagnosis. The case is presented for its rarity and to highlight the importance of awareness about histological and immunohistochemically features of this rare variant of mesothelioma.

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