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MALIGNANT MESOTHELIOMA PRESENTING AS MEDIASTINAL MASS - A RARE CASE REPORT

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Abstract : Malignant mesothelioma is a rare neoplasm. It occurs over the serosal surface of the pleura, pericardium, peritoneum and tunica vaginalis of testis. It is commonly seen in adult males, in the age group between 45 to 75 years. Cases occurring in families have also been reported. Mesotheliomas typically present with pain and effusion. We reported one such interesting case of anterior mediastinal mass lesion in a 65 year old male who presented with right sided chest pain, dry cough and facial puffiness for one month duration. The biopsy of the mass was taken and sent for histopathological examination. The microscopic examination and immunohistochemistry confirmed the diagnosis as malignant mesothelioma.

Keyword : Malignant mesothelioma, Mediastinal mass, Immunohistochemistry

INTRODUCTION:

Malignant mesothelioma is an aggressive tumour that arises from mesothelial cells, which makes up the normal lining of the pleural, pericardial, and abdominal cavity. Incidence of malignant mesothelioma is 0.9 per 100,000 persons annually (1).It is more common in men than in women, with a male-to-female ratio of 3:1.

CASE REPORT:

A sixty five year old male, nonsmoker presented with history of right sided chestpain, drycough and facial puffiness for a period of one month. There is no history of occupational exposure to asbestos. Clinical examination showed facial edema ,bilaterally distended neck veins, fullness over right infraclavicular area, dullness on percussion and diminished air entry over the right side of the chest. X-ray chest showed a mass lesion inthe anterior mediastinum. Computed tomography of chest suggested, it as bronchogenic carcinoma with mediastinal invasion. CT-guided biopsy was done and sent for histopathological examination. Histopathological examination showed a cellular neoplasm arranged in papillary pattern with central edematous fibrovascular cores. The papillae were lined by a single layer of bland flattened to cuboidal mesothelial cells with centrally placed nuclei and inconspicuous nucleoli. The mitotic figures were absent or rare. The tumor cells showed evidence of invasion to stroma in a focal area. Based on the histopathological features it was diagnosed as well differentiated papillary malignant mesothelioma. Immunohistochemistry was done to confirm the diagnosis , which showed diffuse positivity for calretinin and WT-1 and negativity for TTF1 and CEA.

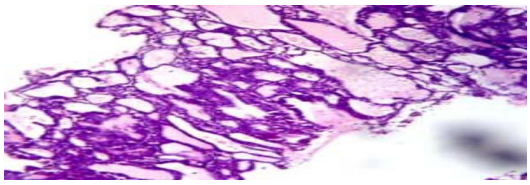

Figure 1: well differentiated papillary malignant mesothelioma (H&E, Low power view)

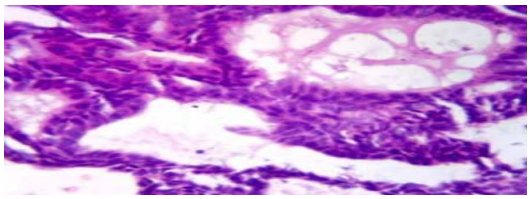

Figure 2: well differentiated papillary malignant mesothelioma (H&E ,High power view)

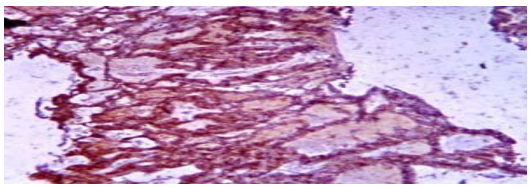

Figure 3:Tumor cells show nuclear and cytoplasmic positivity for calretinin.

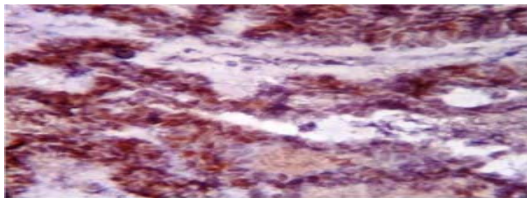

Figure 4:Tumor cells show nuclear positivity for WT-1.

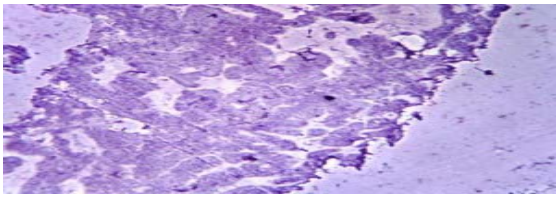

Figure 5:Tumor cells show negative for TTF-1.

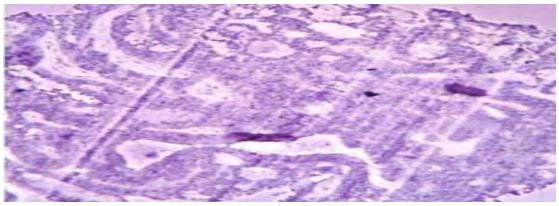

Figure 6: Tumor cells show negativity for Carcino embriogenic antigen.

With the above features it was confirmed as infiltrating well differentiated papillary malignant mesothelioma.

DISCUSSION:

Malignant mesothelioma is an extremely rare neoplasm. It occurs over the serosal surfaces of pleura, pericardium, peritoneum and tunica vaginalis of testis. The tumor mainly occurs in adults in the age group between 45 to 75 years , with male predominance and rarely occurs in children. Pleura is the common site for malignant mesothelioma .Exposure to asbestos increases the risk of mesothelioma(2). This tumor usually presents with chestpain, breathlessness, weight loss, fever and cough. Physical examination reveals decreased breath sounds and pleural effusion on the right side of the chest. Radiologically pleural effusion, atelectasis, diffuse and irregular nodular thickening of pleura can occur(3). Pleural mesothelioma commonly presents with multiple gray white illdefined nodules with diffuse thickening(3,4) and rarely it may present as localized mass6. Histologically they are classified in to four types(3,4)

(1) Diffuse malignant mesothelioma – commonest type and accounts for 85% of the cases .
(2) Well differentiated papillary mesothelioma
(3) Multicystic peritoneal mesothelioma
(4) Adenomatoid tumor.

Diffuse malignant mesothelioma is histologically divided into Epithelial, Fibrous and Mixed types. Among them epithelial type is the most common one. It can be identified by their characteristic focal presentation of tubulopapillary pattern. This pattern consists of papillary structures, branching tubules, and gland-like acinar and cystic spaces lined by either uniform cuboidal or flattened epithelial-like cells with vesicular nuclei, one or two nucleoli, and abundant eosinophilic cytoplasm with distinct cytoplasmic borders(5). Fibrous or sarcomatoid mesothelioma will have well oriented, spindle shaped, fibroblast like cells with nuclear pleomorphism, hyperchromatism with areas of fibrosis, hyalinization, necrosis and with increased mitotic rate. In mixed or biphasic type have mixtures of both epithelioid and fibrous components. Well differentiated papillary mesothelioma is characterized by a proliferation of uniform, cuboidal cells with centrally placed nuclei and inconspicuous nucleoli that line well-formed papillary structures. Mitotic figures are absent or rare. Multicystic peritoneal mesothelioma , shows one or more variously sized, round or irregularly shaped cystic spaces lined by a single layer of flattened or cuboidal mesothelial cells. Adenomatoid tumor shows a variable structural pattern ranging from irregularly arranged, dilated tubular channels and gland-like spaces lined by flattened or cuboidal cells to solid nests and strands of plump cells with abundant eosinophilic cytoplasm.

DIFFERENTIAL DIAGNOSIS:

Differential diagnosis includes,

1. Reactive mesothelial proliferation
2. Primary adenocarcinoma of the lung
3. Metastatic carcinoma of other epithelial neoplasms.

1.Reactive mesothelial proliferation:

Reactive mesothelial proliferations are limited to the serosal surfaces, where they may form small papillary structures, usually with gradual transitions between normal and hyperplastic mesothelium. Malignant mesothelial cells show more nuclear atypia and more prominent nucleoli than in reactive mesothelial cells. The presence of necrosis and invasion in to underlying tissues favors the diagnosis of mesothelioma3.Most epithelial mesotheliomas exhibit extensive, strong linear membrane staining using antibodies to epithelial membrane antigen, in contrast to the weak or undetectable staining seen in reactive mesothelial hyperplasia.

2. Primary adenocarcinoma of the lung:

These tumors show infiltration of the pleura by nests or sheets of cells that focally form glands or tubulopapillary structures. Psammoma bodies can also be found, particularly in the papillary areas. Isolated glands lying within a fibrotic stromaare a common finding. The glandular lumens are oftenfilled with PAS-positive, diastase-resistant ,mucin or intracytoplasmic vacuoles. Immunohistochemical studies show strong positivity for polyclonal CEA and low molecular weight cytokeratins in 100% of cases.(5)

3. Metastatic carcinoma of other epithelial neoplasm:

These includes tumors like metastatic serous ovarian carcinoma and thyroid tumors. Mesotheliomas are positive for calretinin, cytokeratin and WT-1 Metastatic serous ovarian carcinomas are positive for ER,PR and MOC-31. Metastatic thyroid tumors are positive for TTF-1.

CONCLUSION:

Even though, occupation history is negative ,elderly person with mediastinal mass malignant mesothelioma should be considered as a important differential diagnosis. Here immunohistochemistry plays an important role for confirmation.

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