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MALIGNANT PERITONEAL MESOTHELIOMA -AN UNFORGOTTEN ABDOMINAL MALIGNANCY

General Surgery

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ABSTRACT

Malignant peritoneal mesothelioma is a rare lethal malignancy of the serosal membranes of the peritoneum. The pathogenesis and association is strongly related with industrial pollutants asbestos, but less than pleural mesothelioma. Symptoms are nonspecific and related to the tumor spread within the abdominal cavity. CT scan is the investigation of choice and mostly disease is discovered incidentally on routine imaging. Diagnosis is confirmed on histopathology as well as immunohistochemical analysis of markers. The mainstay of treatment is cytoreductive surgery with Hyperthermic intraperitoneal chemotherapy. Here we present a very unusual case of malignant peritoneal mesothelioma diagnosed on routine evaluation of a 62 year old male admitted in emergency for obstructed inguinal hernia.

Introduction: Malignant peritoneal Mesothelioma MPM is a very rare malignancy of the abdominal cavity. Mesotheliomas usually originate from the serosal membrane of different body cavities. Pleura is most commonly affected by mesothelioma followed by peritoneum and also other cavities pericardium and tunica vaginalis testis.10 to 30% of all mesothelioma affects peritoneum. Malignant peritoneal mesothelioma is a highly lethal malignant tumor of peritoneum and its pathogenesis is strongly related with industrial pollutant asbestos exposure. Diagnosis is difficult in most of the cases because of its nonspecific presentation and detected on routine abdominal imaging or Surgery.

KEYWORDS

Malignant mesothelioma, Inguinal hernia, Immunohistochemistry

CASE REPORT

62 year old male patient presented to the emergency department of our hospital with the chief complain of pain abdomen, vomiting and swelling on the right inguinoscotal region for 1 day. On examination, abdomen was distended with tenderness all over the abdomen with obstructed right side inguinal hernia. The patient was resuscitated and was prepared for emergency surgery. But during this period the hernia was reduced with relieve of intestinal obstruction. Further evaluation with ultrasound shows right sided inguinal hernia with omentum, mild to moderate ascites and echogenic debris over the diaphragm and omentum. Contrast enhanced CT scan confirmed multiple omental, peritoneal and liver surface deposits suggesting possibility of metastases [Fig1]. On the basis of above findings, diagnostic laparoscopy was done and revealed metastatic peritoneal deposits all over the peritoneal cavity with gross ascites [Fig.2].Biopsy histopathological analysis was Malignant peritoneal mesothelioma. Immunohistochemistry showed CK5/6 strong cytoplasmic activity in more than 90% tumor cell [Fig.3] and CEA negative in tumor cell, D2-40 strong membranous positivity in tumor cell [Fig 4]. There was no history of any chronic asbestos exposure. Patient was planned for cytoreductive surgery with Chemotherapy, but got discharged on request and lost to follow up.

DISCUSSION

First described by Miller and Wynn in 1908, Malignant peritoneal mesothelioma is a very rare and uncommon malignancy of peritoneum [1].Most of this serosal malignancy occurs in the pleural cavity and strongly related to the occupational exposure of asbestos. Epidemiologically peritoneal mesothelioma differs from the common pleural mesothelioma. This MPM is a disease of adults with median age of presentation around 51-59 yrs, younger than pleural mesothelioma. MPM affects equally in both males and females , whereas it predominately affects male in pleural mesothelioma [2]. There is a strong relationship between asbestos exposure and the development of mesothelioma, although the risk is less in peritoneal compared to pleural mesothelioma (33-50% vs. 80%). Asbestos is the best defined risk factor [3] and the latency period between exposure and development is around 20 to 40 yrs [4]. Other identifiable risk factors are direct peritoneal external beam radiation, Simian virus 40, papovavirus, chronic peritonitis and also in genetic inherited susceptibility of BRCA associated protein1 (BAP1) mutation. BAP1 is the most promising marker not only in making diagnosis, but also differentiating benign from malignancy, and germline mutations in younger patients and with a relevant family history.

Symptoms are nonspecific and related to the extent of tumor in the abdominal cavity. The most frequent presentations are abdominal distension and pain in 30-50% of patients due to ascites. Other symptoms are anorexia, weight loss, abdominal mass and new onset abdominal wall hernia [5], like seen in our case in acute presentation. Nonspecific symptoms are confusing leading to late diagnosis and also made as a surprise in routine imaging, laparoscopy or laparotomy. The average time between the onset of symptoms and diagnosis is approximately five months [6]. Rarely patient present with paraneoplastic syndrome like fever, thrombocytosis, hypoglycemia combs' positive hemolytic anemia and baseline thrombocytosis is one of the independent factor related with shortened survival in diffuse MPM[7].The clinical presentation of MPM also depends upon the pattern of spread, whether typical diffuse or less common localized disease with better prognosis.

Contrast CT Scan is the imaging of choice and most of the diagnosis often made after CT scan. Soft heterogeneous tissue mass with diffuse distribution should arouse suspicion. Ascitic fluid is more common findings apart from thickening, caking of omentum and mesentery [8]. Our case also has similar findings on CT scan with suspicion and was confirmed after diagnostic laparoscopy and biopsy. Before making diagnosis, clinical information is utmost important to consider the possibility of MPM. Histologically 3 main subtypes are epithelioid, sarcomatoid and biphagic mixed type. In the immunohistochemical analysis, Tandon et al. found that the most sensitive markers calretinin (100%), WT1 (94%) and CK5/6 (89%) , D2-40 was positive in 80% of cases[9]

The Treatment modality is focused on targeting the peritoneum with Cytoreductive surgery and intraperitoneal chemotherapy. Surgical aim is complete macroscopic removal of affected peritoneum along with visceral resection. Intraperitoneal chemotherapy delivered during surgery is Hyperthermia intraperitoneal chemotherapy (HIPEC) and normothermic Early postoperative chemothetherapy (EPIC).The role of immunotherapy in chemotherapy-refractory peritoneal mesothelioma is evolving and will offer a possible new treatment.

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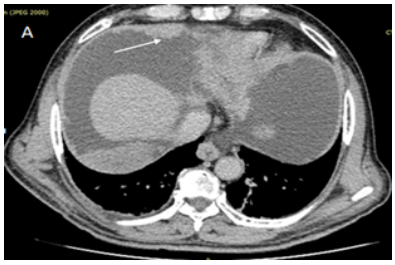

Fig.1-CT Scan showing ascites with multiple deposits

Fig.2 Peritoneal deposits and ascites on Dx. lap.

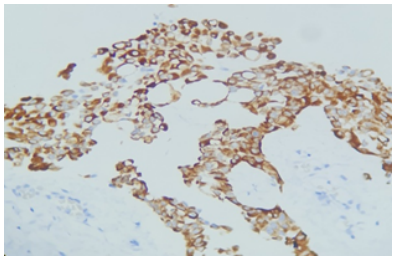

Fig.3 CK 5/6, strongly positive in tumor cell

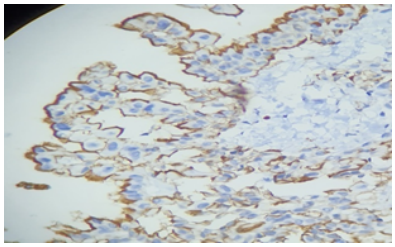

Fig.3 D 2-40-Strongly positive in tumor cells

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