MALIGNANT CARCINOID TUMOR PRESENTING WITH CHYLOUS ASCITES AND LEFT SUPRACLAVICULAR LYMPHADENOPATHY

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ABSTRACT

Gastrointestinal(GI) Neuroendocrine tumors are derived from the diffuse Neuroendocrine system of GI tract. Carcinoids synthesize peptides and bioactive amines that can give rise to clinical syndromes. These secretions are metabolized in the liver. GI carcinoids account for 74% and respiratory carcinoids account for 25% of all carcinoids. The rest accounting for 1%.

The presenting complaints in a carcinoid tumor are varied. As a consequence it presents diagnostic difficulties to both clinicians and pathologists. We present a case of a 60 year old man who presented with ascites and on subsequent work up, was diagnosed to have a malignant ileocecal carcinoid with chylous ascites and left supraclavicular lymph node metastases.

This case is being presented to highlight the unusual presentation of carcinoid, like chylous ascites and left supraclavicular lymph node metastasis. It also brings to the fore the role of FNAC in diagnosis, especially of lesions which cannot be easily biopsied.

Key words: *Malignant carcinoid, Ascites, chylous, Supraclavicular Lymphadenopathy.*

AIM

1. To highlight the unusual presentation of carcinoid like chylous ascites and left supraclavicular lymph node metastasis.
2. To highlight the role of FNAC in the diagnosis, especially of lesions which cannot be easily biopsied.

CASE REPORT

A 60 year old man presented to the medical OPD with abdominal distention and generalized weakness since 1 month. There was no history of fever, loss of appetite or loss of weight.

On examination, patient had mild pallor. There was no lymphadenopathy. He was hemodynamically stable. Pulse was 80/min, regular. Blood pressure was 130/80. There was no raised JVP.

Abdominal examination revealed abdominal distention with ascites. There was no mass or organomegaly .

Examination of the other systems was normal.

On investigating, patient was found to have ascites. The fluid was apirated and sent for analysis.

No abdominal mass was detected on ultrasound at that time.

CT scan of the abdomen showed ascites with normal architecture of the liver.

The ascitic fluid tapped was milky white in colour, turbidity disappeared on adding ether

Hence the possibility of chylous ascites was considered. Fluid analysis revealed 250 cells/cumm, lymphocytes and mesothelial cells with elevated Triglyceride (140mg/dl) levels, both of which were in favour of chylous ascites. Fluid cytology did not reveal any abnormal cells. In our country, since tuberculosis is the commonest cause of chylous ascites, patient was empirically put on antitubercular drugs and advised to come for regular follow up.

On follow up after 2 months, general physical examination showed that the patient had a left supraclavicular lymph node enlargement measuring 2x2 cms, firm in consistency. Repeat ultrasound of the abdomen revealed a mass measuring 4x3 cms in ileocaecal region. CT scan of the abdomen could not be repeated due to financial constraints.

An ultrasound guided FNAC of the ileocaecal mass was done along with a biopsy of left supraclavicular lymph node. This way diagnosis was obtained from both lesions. Since the supraclavicular node was more amenable to biopsy, a biopsy was performed rather than a FNAC. FNAC smears from the mass showed moderate cellular yield with uniform round cells in clusters and sheets. The cells had scanty cytoplasm with salt and peppery chromatin in

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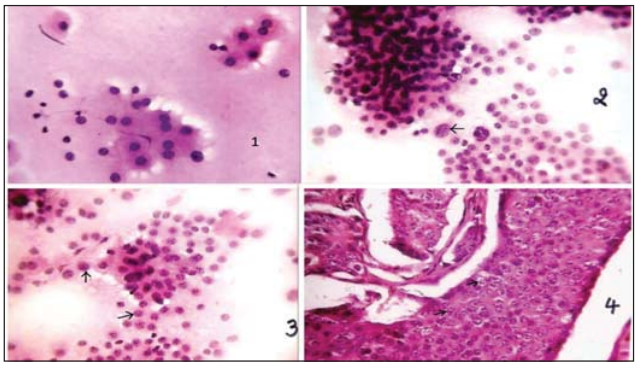
a haemorrhagic background (Fig 1 & 2). A diagnosis of carcinoid was thought of. Absence of signet ring cells and mucin helped in ruling out an Adenocarcinoma.

Histopathology of the lymph node revealed tumor cells in nests, sheets and trabeculae replacing most of the lymph node. The tumors cells were round to oval with stippled chromatin admixed with polymorphous population of lymphocytes (Fig 3 & 4). Ocassional mitotic figures were seen. No signet ring cells were seen. Hence a diagnosis of malignant carcinoid of ileocaecal region metastasing to the left supraclavicular lymph node with ascites was made. Patient refused further treatment and was lost for follow-up.



Figures:

1. Ascitic fluid 45x Lymphocytes and mesothelial cells
2. FNAC abdominal mass 45x : cells having stippled chromatin(carcinoid cells)(Single bold arrow) and:Acinar cells in groups(double bodied thin arrow)
3. FNAC abdominal mass 45x : Acinar cells (double bodied thin arrow) Carcinoid cells(Single bold arrow)
4. Lymph node Histopathology 45x Cell having stippled chromatin (carcinoid cells)replacing lymphocytes in lymph node(Single bold arrow) A few mitoses can be appreciated(double bodied thin arrow)

DISCUSSION

GI carcinoids are ill-understood, enigmatic malignancies, which, although slow growing compared with adenocarcinoma, can behave aggressively. Carcinoids are classified based on organ, site and cell of origin and occurs more frequently in the GI tract (67%) where they are more common in small intestine (25%), appendix (12%), rectum (14%) and caecum (5%)[1,2].

The small bowel is the most common site for carcinoid tumors, accounting for almost 25% of all carcinoids. The ileum is much more commonly affected than the rest of the small bowel. Carcinoid tumors pose a diagnostic challenge because they are often asymptomatic. Because many carcinoid tumors are indolent, their true prevalence maybe higher than estimated[3].

All carcinoids are potentially malignant. Appendiceal and rectal carcinoids are seldom malignant while on the other hand, ileal, gastric and colonic carcinoids are frequently malignant. A high proportion of these have already metastasized by the time they are detected. More than 66% of carcinoids greater than 2cm show metastasis when first detected, whereas less than 5% of those under 1cm do so. However, tumor size is an unreliable predictor of metastatic disease, and metastases have been reported even from tumors measuring less than 0.5 cm in diameter[2,4].

FNAC can be a useful tool in the diagnosis of metastatic carcinoid tumors avoiding the need for more invasive procedures like biopsy. Despite the infrequent occurrence of these tumors their interesting clinical and pathological characteristics have been extensively reported. Chylous ascites associated with malignancy is rarely encountered in clinical practice. 28 cases were reported from Massachusetts General Hospital in 20yrs. Chylous ascites is most frequently associated with malignant conditions; particularly in lymphomas and disseminated Carcinoma from primaries in pancreas, breast, colon, ovary and kidney. Inflammatory disorders, such as tuberculosis, can infrequently be associated with chylous ascites. In one series, of the 275 cases of carcinoids reported, only 3 patients had chylous ascites[5,6,7]. Chylous ascites may be caused by either of 2 mechanisms: interruption of the normal lymphatic drainage at the lymph node level due to neoplastic invasion or fibrosis, or obstruction of the lymphatic ducts caused by fibrosis of the surrounding tissue8leading to extravasation of milky chyle into peritoneal cavity.

Carcinoids metastasizing to supra clavicular nodes is very uncommon. Distant metastasis occurs more commonly to liver, bone and lung. Small bowel carcinoid tumors usually metastasize to regional lymph nodes and the liver but metastases to the neck are extremely rare. In one study, of the 48cases diagnosed over a period of 10 years, only 2 cases had metastasis to cervical lymph nodes. Only 3 of 22 carcinoids had supraclavicular metastasis in another study[2,9,10].

Treatment decisions for patients with carcinoid tumors are complex and related to the location of the primary tumor and whether or not metastasis has occurred.

Options include surgery, chemotherapy, and radiation with somatostatin analogues such as octreotide or alpha-interferon[3].

CONCLUSIONS

Carcinoid tumors pose a diagnostic challenge because they often are asymptomatic.

Carcinoid tumor with chylous ascites and metastasis to supraclavicular lymph nodes are uncommon occurrences, as has been discussed. This makes our case an interesting one since both these rare manifestations were present in the same patient.

It also emphasizes the role of FNAC in diagnosis.

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