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Undifferentiated Spindle Cell Carcinoma of Gallbladder presenting as Gallbladder perforation- A Case Report

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ABSTRACT

Gallbladder carcinoma is an aggressively malignant disease carrying an extremely poor prognosis. Undifferentiated spindle cell carcinoma, the rarest of all gallbladder cancers also has the same prognosis. Patients usually have no specific or vague presenting symptoms, thus, in such scenario presentation with late-stage disease is often common.

We herein have reported a case of 63-year-old male who presented with short history of abdominal pain and fever. Our case was different as his radiologic investigations were suggestive of cholecystitis with cholelithiasis for which cholecystectomy was done but it was later diagnosed to be a case of SpCC.

KEYWORDS: Gallbladder, Carcinoma, Sarcoma, spindle cell, perforation

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INTRODUCTION

Undifferentiated spindle-cell carcinoma (SpCC) of the gallbladder is one of the rarest gallbladder cancer comprising predominantly of sarcomatous elements along with carcinomatous elements [1]. Evidences from various reports suggests that it has epithelial origin with sarcomatous dedifferentiation or stroma induction supported by focal immunopositivity of the mesenchymal component for epithelial markers [2, 3]. Thus, morphological demonstration of biphasic appearances is essential for a diagnosis of spindle-cell carcinoma; however, in some instances, this may be difficult even after multiple sections and immunohistochemistry [4, 5].

SpCC have worse prognosis and poor survival as compared to conventional gallbladder carcinoma following treatment [2, 5].

CASE REPORT

A 63yr old male carpenter by occupation presented with complains of sudden, sharp upper abdominal pain and low-grade fever (on and off) since 4-5 days. Pain was sudden in onset, sharp gripping in nature, progressive, diffuse over whole of the upper abdomen (more on right side), non-radiating, continuous and was relieved only on medication, no aggravating factors. Patient had no complains of dyspepsia, nausea/vomiting, hematemesis/melaena, loss of appetite, diarrhea/constipation

On examination, patient conscious, oriented; well-built and nourished. Pallor, icterus, cyanosis, clubbing, lymphadenopathy, pedal oedema were absent. His pulse was 84/min, blood pressure 136/74mmHg, respiratory rate 18/min, SpO_2:94% at room air, and was afebrile. Abdomen on examination was soft, tenderness in right hypochondrial and epigastric region, no local rise in temperature, umbilicus inverted, no visible swelling/dilated vessel/scar/peristalsis, no palpable organomegaly. Liver dullness present. Bowel sounds were good.

Patient was admitted and routine investigations and abdominal USG was done. The abdominal ultrasonography showed “Cholelithiasis (16 mm) in gall bladder lumen with cholecystitis with thickening? Neoplastic and Fatty liver, grade I.” Chest X-ray and 2D ECHO of patient was normal. Complete blood count was normal except leukocytosis (TLC 14,700/cumm). His total bilirubin was mildly raised (total=1.32mg/dl, direct= 0.88mg/dl, indirect= 0.44mg/dl), Serum alkaline phosphatase was 219 U/L, CEA was 2.49 and CA 19-9 was 15.1. following this to confirm USG findings CT scan and MRCP was also done. Computed tomography scan revealed “Gallbladder perforation with adjacent collection with Cholelithiasis 16 mm in gall bladder lumen

with cholecystitis. Hiatus hernia present." (Figure 2) and MRCP showing thickened gall bladder wall. (Figure 2)

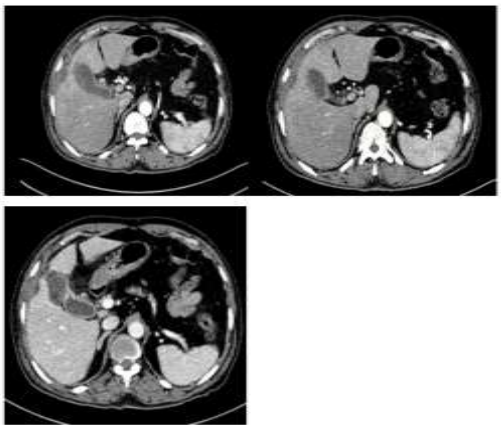

Figure 1. CT scan showing gallbladder perforation with adjacent collection with Cholelithiasis with cholecystitis.

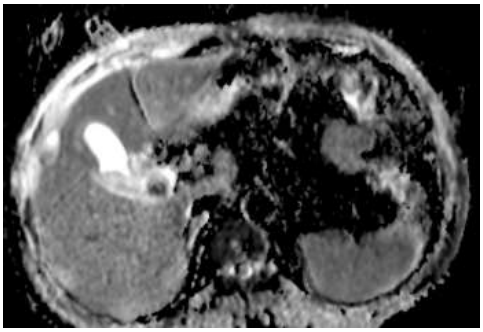

Figure 2. MRCP showing thickened gallbladder wall

Considering it to be a case of perforated gall bladder with cholecystitis patient was admitted and planned for cholecystectomy. Open cholecystectomy was done under GA the following day. Significant intraoperative findings include thickened and edematous GB; Omentum, colon and duodenum were fully covering gall bladder; Gall bladder was perforated at fundus with pus coming out; large stone was impacted at neck with pus filled GB. Following cholecystectomy, the removed GB with stone was sent for histopathology examination.

On histopathology gallbladder specimen was found to be showing spindle cell carcinoma (Figure 3). IHC (immunohistochemistry) was also done suggestive of SSC (Figure 4).

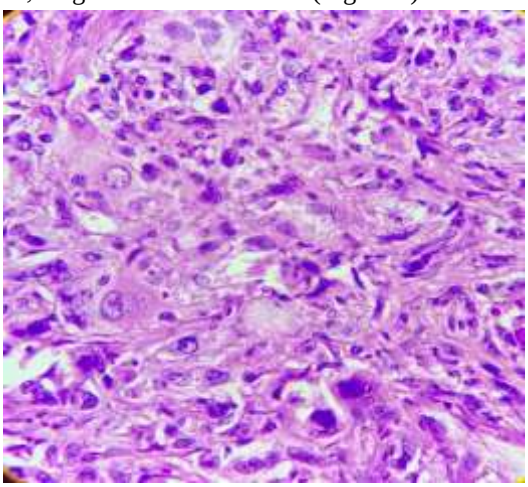

Figure 3. showing pleomorphic spindle cells with few epitheloid cells(H&Ex400)

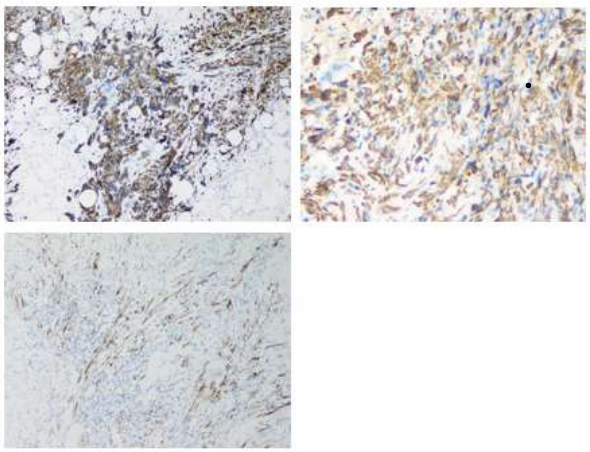

Figure 4. IHC showing CKA, SMA and Desmin reactivity.

In view of histopathology and IHC findings suggestive of SPCC patient was offered palliative chemotherapy as disease was considered M1 in view of gall bladder perforation; but patient refused for the same. Patient was on regular follow since then and is doing well, had no complains except for occasional indigestion.

DISCUSSION

The majority of gallbladder cancers are conventional adenocarcinoma, whereas spindle-cell carcinoma (SpCC) is very rare [6, 7]. One of the recognized histologic forms of spindle cell carcinoma of the gallbladder is that termed anaplastic or undifferentiated, however detailed morphological descriptions of these GB neoplasms is still rarely reported [8,9,10,11]. Women above 60 years of age having symptomatic history gallstones since several years are considered more likely to develop gallbladder carcinoma.

However, specific predisposition factor for SpCC is still not well known, neither its presentation. Most of these diagnoses are made in post-operative period based on histopathology and IHC studies. In view of these rare tumor type no consensus guidelines are available as of now. So, most of the clinicians follow available adenocarcinoma guidelines for further treatment.

From a prognostic point of view, there is only little difference in survival of patients with pleomorphic adenocarcinoma, so called sarcoma [12,13,14]. Life expectancy after diagnosis is measured just in months, rather than in years. It is often found that most patients die from or with their neoplasms within one year after the onset of symptoms.

The involvement of serosa of GB and/or of other organs as well as advanced stage are two identified factors that leads to poor postsurgical outcomes in these patients. This points to the fact that curative surgery is nearly impossible in most patients with gallbladder carcinoma due to the advanced stage at presentation. Thus, patients and their relatives need to be thoroughly briefed about this tumor and its prognosis before and following surgery. Newer chemo-radiotherapeutic agents and the molecular targeted therapy would be essential to improve recurrence free survival following surgery [15]. Often undifferentiated spindle cell carcinoma in its initial stages is considered to be a sarcoma, and addition of glandular element leads towards the diagnosis of carcinosarcoma.

However, early diagnosis and surgical intervention with better understanding of this carcinoma biology may help improve prognosis and survival in this rare cancer.

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CONFLICT OF INTEREST

None

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