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CASE REPORT

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A Giant Fibrosarcoma of Gall bladder - A Rare Presentation- A Case Report

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ABSTRACT

Introduction: Primary gall bladder sarcoma (PGBS) is a rare histopathological diagnosis with uncertainties regarding management. Evidence is available only in the form of anecdotal reports.

Case Description: A 57-year lady presented to our centre with upper abdominal pain and a palpable mass in the right hypochondrium. CT scan revealed a distended gallbladder with a mass in the gallbladder. Repeated Gall bladder biopsy was inconclusive of malignancy, and workup revealed no other evidence of distant metastasis. In view of the massively enlarged gallbladder with corroborating clinical symptoms, cholecystectomy with liver wedge excision and periportal lymph nodal sampling was performed. The histopathological and immunohistochemical evaluation confirmed the diagnosis of a primary gall bladder sarcoma. The patient recovered well and has received adjuvant chemotherapy.

Conclusion: A rare histological diagnosis masquerading gallbladder cancer may present as a clinical surprise. These rare histologies ought to be reported to help in the timely management of curable diagnostic dilemmas.

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Case Management

A 57-year lady with no contributing past medical or surgical history presented to our centre with complaints of upper abdominal pain for two months. On examination, 8x8 cm hard mass was palpable in the right hypochondrium extending to the right lumbar region. Serum CA 19-9 was 20 U/ml. Contrast-enhanced Computed Tomography scan of thorax, abdomen and pelvis (CECT TAP) showed a grossly distended gallbladder, with ill-defined solid enhancing endophytic areas measuring 2.8 x 2.7 cm in the gall bladder neck and 8.5x8.2 cm mass in the fundus with compression on the biliary tree leading to mild intrahepatic biliary radical dilatation. Multiple enlarged suspicious periportal lymph nodes were seen (Figure 1).

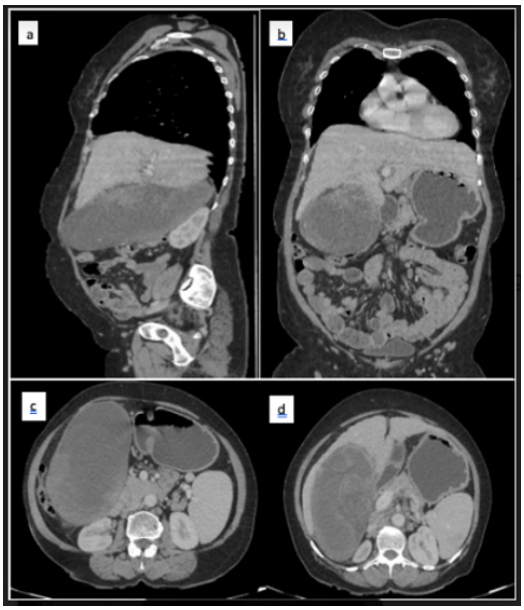

Figure 1: CECT images showing distended GB with mass lesion

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Percutaneous biopsy from gallbladder mass on two separate attempts showed only necrotic tissue. The patient was planned for cholecystectomy because of the grossly enlarged gallbladder with related symptoms of abdominal pain

Intraoperatively, a huge distended gall bladder infiltrating the liver with multiple soft, enlarged periportal lymph nodes were seen. A fundus first cholecystectomy with liver wedge excision along with peri-portal lymphadenectomy was performed (Figure 2).

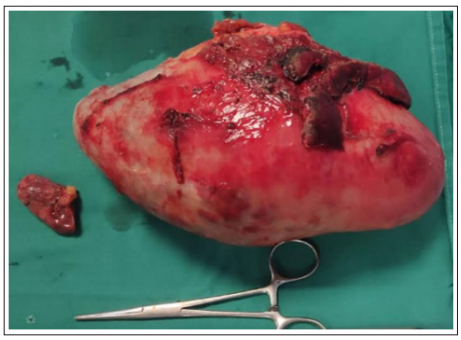

Figure 2: Cholecystectomy with liver wedge specimen (with periportal node)

The postoperative recovery was uneventful, and the patient was discharged on postoperative day 4. Final histopathology revealed a high-grade sarcoma favouring fibrosarcoma limited to the gallbladder, with all resected margins free of tumour and no lymph node metastases (Figure 3).

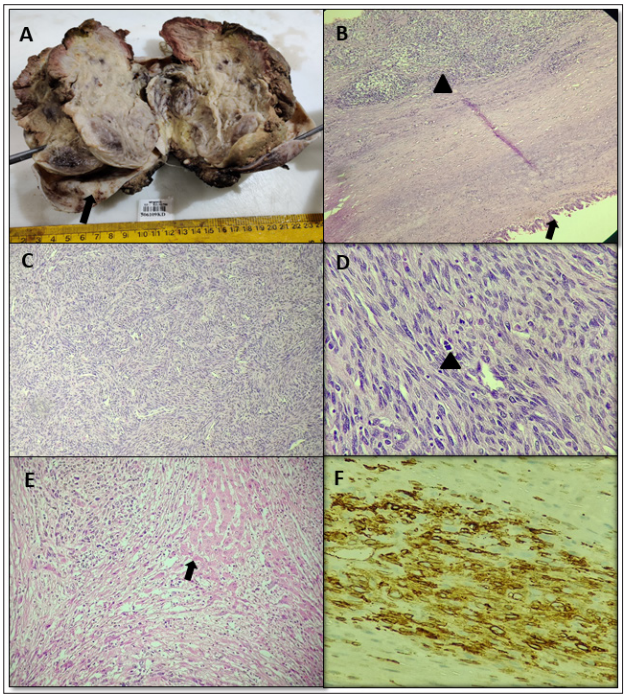

Figure 3: A) Gross image shows distended gall bladder lumen (arrow) with a solid grey-white lesion of 14x 7.5x 9.9 cm involving the body and neck, B) H & E (10X) Shows unremarkable gall bladder mucosa on the right side lower corner (arrow) and tumor on the upper end (arrow head), C) H & E (20X) Highlights the typical "Herringbone" pattern of arrangement of the tumor cells, D) H & E (40X) Shows the spindle cells having bipolar cytoplasm, mild to moderate nuclear pleomorphism and inconspicuous nucleoli. Numerous atypical mitotic figures are seen (arrow head), E) Tumor cells infiltrating the normal hepatic parenchyma (arrow), F) Strong positivity of the tumor cells by CD34 immunostain.

The patient received six cycles of Ifosfamide and Adriamycin based adjuvant chemotherapy and is currently disease-free for six months.

Discussion

Malignant mesenchymal tumours of the gallbladder are extremely rare [1]. Various tumour types have been reported, such as Leiomyosarcoma, rhabdomyosarcoma, angiosarcoma, Kaposi's sarcoma, malignant fibrous histiocytoma (MFH), and synovial sarcoma [1-3]. Leiomyosarcoma is the most common type of primary gallbladder sarcoma, while MFH is more common in some series [3].

Primary sarcoma of the gall bladder is more common in the sixth and seventh decade of life and rarely occur in children [3, 4]. Women are affected more commonly than men, like gall bladder carcinoma. The adult sarcomas are heterogeneous histologically and have poorer outcomes. But, pediatric tumours, in some series like botryoid embryonal rhabdomyosarcoma, are associated with favourable outcomes following adjuvant therapy [3].

Most primary gall bladder sarcoma patients present with abdominal pain, fever, jaundice, and weight loss [3,5] In some cases, acute or chronic cholecystitis accompanied by cholelithiasis led to cholecystectomy and sarcoma was an incidental histopathological diagnosis [3,4,6]. Preoperative evaluation can raise a suspicion of malignancy which can be confirmed only after histopathological examination [6].

The differential diagnosis of gallbladder sarcoma includes primary or metastatic undifferentiated carcinoma,

carcinosarcoma, and metastatic melanoma. These are excluded by histological examination and immunohistochemical staining with pankeratin, CK18, S100 protein, and HMB45 [3].

Etiopathogenesis of gall bladder fibrosarcoma is not well understood. Gall stones and chronic inflammation are likely predisposing factors [4, 5]. Genetic alterations in regulatory genes p53 and RB are also suggested as a cause for PGBS [4].

Because of the limited experience with PGBS, there are no clearly defined guidelines. Suspicion for a malignant gallbladder tumour is an indication for laparotomy [3]. The current practice is surgical treatment with a grossly negative margin, which usually includes cholecystectomy with wedge resection of liver and hepatoduodenal lymphadenectomy [6-8]. Frozen section is used to guide the extent of intraoperative excision. The most important prognostic factor is postoperative margin status [9]. Postoperative radiation therapy is considered in patients with high-grade disease following a negative margin resection (R0 resection) or for positive microscopic margins (R1 resection). The role of revision surgery after an incidental diagnosis has not been established and would mandate a tumour board discussion. The extent of liver resection is debatable, with most experts recommending a margin negative liver wedge excision.

For patients with unresectable disease, preoperative chemotherapy should be considered. There is not sufficient evidence in the literature regarding the effectiveness of adjuvant chemotherapy or radiation therapy. However, some authors report that chemotherapy with doxorubicin, mitomycin C may improve long term survival following surgery [10,11]. Combination regimens used are AD (doxorubicin, dacarbazine), AIM (doxorubicin, ifosfamide, mesna), and MAID [12,13]. The single agents used

include dacarbazine, doxorubicin, epirubicin, gemcitabine, ifosfamide, liposomal doxorubicin and temozolomide [14, 15].

The overall prognosis of gall bladder sarcoma is dismal. The five-year survival rate is less than 5%. This is attributed to most cases being advanced at the time of presentation or surgery. Almost 75% of cases involve the liver [13,14]. Aggressive multimodality treatment should be considered for young and healthy patients and will likely improve the survival outcome [7].

Conclusion

Surgery and chemotherapy are the mainstays of treatment of gall bladder sarcoma. Such cases, though rare, need aggressive management for the best outcome. Cholecystectomy with liver wedge excision with lymph nodal dissection should be performed in the setting of high clinical-radiological suspicion [16,17].

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