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The Muculent Bleb-Mucinous Cystic Neoplasm-Hepatobiliary Region

Bajaj A*

Panjab University, India

***Corresponding author:** Anubha Bajaj, Panjab University, A.B. Diagnostics, A-1, Ring Road, Rajouri Garden, New Delhi-110027, India, Email: anubha.bajaj@gmail.com

Editorial

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Abbreviations: ER: Oestrogen Receptors; PR: Progesterone Receptors; FNAC: Fine Needle Aspiration Cytology; CEA: Arcinoembryonic Antigen.

Editorial

Mucinous cystic neoplasm configures as a cystic neoplasm incriminating the hepatic parenchyma. Tumour is constituted of multi-locular or cystic cavity layered with mucinous or biliary type epithelium encompassed within an ovarian type stroma. The cystic lesion appears non communicative with adjoining bile ducts. Additionally designated as hepatobiliary cystadenoma, biliary cystadenoma or cystadenoma with mesenchymal stroma, neoplasms associated with invasive carcinoma component may be denominated as hepatobiliary cystadenocarcinoma.

Comprehensive surgical eradication of the neoplasm accompanied with extensive tissue sampling for cogent histological assessment is mandated in order to ascertain foci of invasive carcinoma and decimate possible tumour reoccurrence. Tumefaction is preponderantly (~90%) benign although foci of invasive carcinoma may be enunciated. Tumefaction exhibits a female preponderance. Mean age of discernment of non-invasive lesions appears at 45 years wherein lesions concurrent with invasive carcinoma enunciate mean age of detection at 59 years [1,2].

Although mucinous cystic neoplasm commonly incriminates hepatic parenchyma, exceptionally the tumefaction may be confined to extrahepatic bile ducts or gallbladder [1,2]. Of obscure aetiology, mucinous cystic neoplasm of biliary tract exhibits ovarian type stroma which is posited to originate from developing, embryonic gonads demonstrating approximation to hepatic parenchyma during the course of embryologic development [1,2].

Characteristically, neoplasm represents with clinical symptoms as abdominal pain or abdominal tumefaction. Alternatively, symptoms of biliary obstruction as hyperbilirubinemia may be observed [1,2]. Cytological examination exemplifies scanty cellularity with groups of bland, degenerated, columnar or cuboidal epithelial cells. Epithelial cells appear intermingled within an accumulate of inflammatory cells as neutrophils, lymphocytes or macrophages. Circumscribing stromal components or foci of ovarian type stroma appear absent [2,3].

Grossly, a cystic lesion comprised of multitudinous, intercommunicating lobules of varying magnitude pervaded with clear, thin cystic fluid is discerned. Neoplastic lobules are layered by singular coat of smooth and glistening epithelial cells. Extensive tissue sampling of nodules, thickened areas or lesion in entirety is recommended in order to detect foci of high grade dysplasia [2,3].

Upon microscopy, multi-locular cysts appear to be layered by columnar, cuboidal or flattened biliary subtype of epithelium. Alternatively, mucinous epithelial cells appear superimposed upon ovarian type stroma [3,4]. Constituent ovarian type stroma is comprised of dense aggregates of spindle shaped to ovoid cells. Focal luteinisation of stromal cells may occur.

Metamorphosis into high grade epithelial dysplasia is accompanied nuclear pleomorphism and complex architectural alterations as tubulopapillary projections. Focal areas of invasive carcinoma, commonly ductal adenocarcinoma may be observed. Mitotic figures are frequently enunciated (Figure 1) [3,4]. Enlarged lesions exhibit focal epithelial injury with areas of ulceration,

inflammation, xanthogranulomatous reaction, extravasation of cyst fluid into cyst wall or stroma and foci of scarring and calcification (Figure 2) and (Table 1) [3,4].

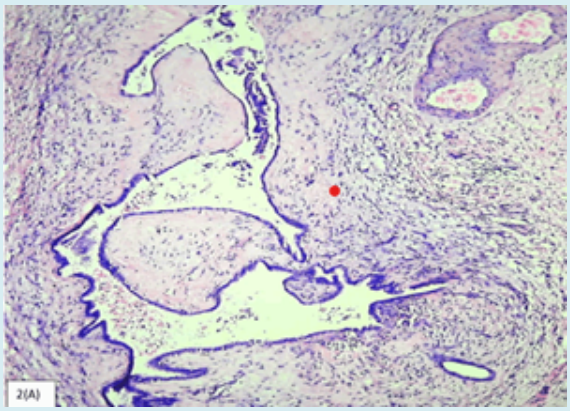

Figure 1: Mucinous cystic neoplasm demonstrating multi-locular cystic spaces layered by cuboidal epithelium surrounded by ovarian type stroma with focal luteinisation [5].

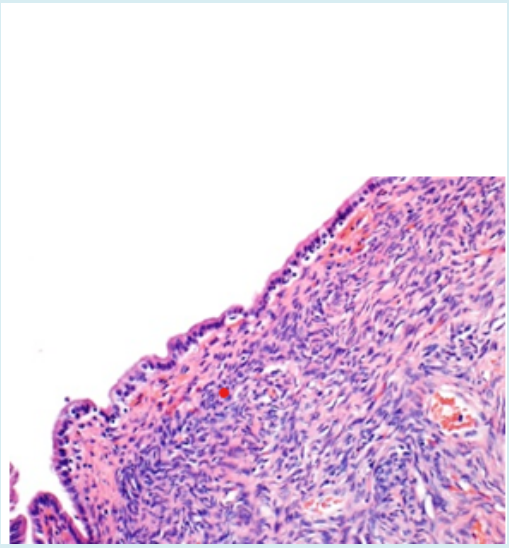

Figure 2: Mucinous cystic neoplasm delineating cystic spaces layered by columnar epithelium encompassed by ovarian type stroma with focal luteinisation and haemorrhage [6].

	Mucinous Cystic Neoplasm Liver	Intraductal Papillary Neoplasm Bile Duct
Imaging features	Multilocular fluid-filled mass with small cysts within cyst wall	Multicystic with grape-like appearance, papillary nodules and peripheral bile duct dilatation
Ductal communication	Commonly absent	Present
Stroma	Ovarian-like stroma, ER+, PR+	Fibrous
Epithelial antigens (immune reactivity)	MUC5AC-, CK7+,CK20-,MUC2-MUC6-	MUC5AC+, variable CK7,CK20, MUC2, MUC6
Malignant potential	Low	High

Table 1: Comparison between Mucinous Cystic Neoplasm Liver and Intraductal Papillary Neoplasm of Bile Duct [1].

Neoplastic cells appear circumscribed by ovarian type stroma which is immune reactive to oestrogen receptors (ER), progesterone receptors (PR) or inhibin A. Epithelial cells layering the cystic lesion appear immune reactive to CK7, CK8, CK18 and CK19. Alternatively, tumour cells within mucinous zones may be highlighted by mucicarmine stains. Epithelial tumour cells appear immune non-reactive to oestrogen receptors (ER) or CK20 [7,8].

Ovarian type stroma appears immune non-reactive to CD10. Mucinous cystic neoplasm of biliary tract requires segregation from neoplasms as endometriosis, intraductal papillary neoplasm, simple cyst incriminating hepatic parenchyma, simple biliary cyst or bile duct cyst [7,8].

Fine needle aspiration cytology (FNAC) and imaging studies appear unsuitable for appropriate distinction of mucinous cystic neoplasms from various categories of hepatic cysts. Serum carcinoembryonic antigen (CEA) and serum CA19-9 levels within constituent cyst fluid may be assayed in order to segregate mucinous neoplasms from non-neoplastic lesions.

Serum CA19-9 values may be elevated, especially within lesions concordant with invasive carcinoma [7,8]. Upon imaging, an enlarged, multilocular cyst traversed with internal septa may be discerned. The lesion may depict focal calcification, mural nodules or foci of papillary projections. Generally, distinction of benign lesions from malignant neoplasms appears challenging upon imaging studies [7,8]. Cogent, definitive tumour discernment necessitates precise histopathological assessment and confirmation of ovarian type stroma which may be obtained with tissue sampling of surgical excision specimens [7,8].

Comprehensive surgical extermination of the neoplasm appears curative. Incomplete surgical eradication of the tumefaction is associated with localized tumour reoccurrence and possible occurrence of malignant metamorphosis [7,8]. Therapeutic manoeuvers as comprehensive surgical eradication of the neoplasm contributes to superior prognostic outcomes [7,8].

Nevertheless, appropriate ascertainment of prognostic significance of invasive tumour component may be challenging as several neoplasms delineating distinct invasive carcinoma component appear devoid of ovarian type stroma. Besides, aforesaid concurrent tumefaction depict a male preponderance and may represent disparate lesions as intraductal papillary neoplasm, in contrast to mucinous cystic neoplasm [7,8].

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