

# A REVIEW OF THE UNUSUAL LIVER TUMOURS

Pages with reference to book, From 53 To 56

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Primary carcinoma of the liver is common in Africans and Orientals, accounting for 17 to 23% of all malignancies. In America it represents 1.5 to 2.0% of all malignancies.<sup>1</sup> Liver tumours, though not the first ten commonest tumours in Pakistan, yet account for 3.7% of all malignancies<sup>2</sup>. Unusual tumours of the liver have variable origin and are even rarer<sup>3</sup> yet it is important to recognize them because they grow slowly, metastasize late and therefore are amenable to surgical excision: hence they have better prognosis than hepatocellular and cholangiocarcinoma.<sup>1,3-15</sup> The purpose of this study was to review the various liver tumours in the present series and to assess their frequencies.

## MATERIAL, METHODS AND RESULTS

Six thousand three hundred and twenty one liver biopsies (needle and open biopsies) were received in the Department of Pathology, Basic Medical Sciences Institute, Jinnah Postgraduate Medical Centre, Karachi, during 27 years (1960—1987). The biopsies were received from various departments of the hospital and institutes outside the hospital. Of the total, only 5,308 biopsies (85%) were adequate enough for histopathological reporting. The tissue originally fixed in 10% formalin was processed and stained for light microscopic examination. Besides routine haematoxyline and eosin stain, special stains like Masson's trichrome, Periodic Acid Schiff (PAS) and Gomori's Silver Stain were used where necessary. Immunofluorescence, electron microscopy, immunoperoxidase and marker studies could not be done due to non availability of these facilities. Twenty seven biopsies (43%) of unusual primary liver tumours were received, of which 7 (1.1%) were benign and 20 (3.2%) were malignant. There were 18 males and 9 females and their ages ranged from 9 months to 75 years with 10 children under the age of 14 years. Various types of benign and malignant unusual liver tumours are shown in Table-I.

TABLE - Unusual, Benign & Malignant Tumours of the Liver.

Tumours	Sex	Age (years)	Biopsy	Salient and Unusual Features	% Out of 626 Primary liver tumours	% Out of 27 Unusual liver tumours
1	2	3	4	5	6	7
Bile duct adenoma	M	26	Needle	Collection of bile ducts separated by fibrous stroma	$1.5 \times 10^{-1}$	3.7
Haemangioma	F	65	"	Cavernous type	$1.5 \times 10^{-1}$	3.7
Haemangio endothelioma	M	4	"	Infantile type 1	$3 \times 10^{-4}$	7.4
	M	2	"	"		
Neurilemmoma	M	35	"	Interlacing fibres with palisading nuclei		
Liver cell adenoma	M	6	Resection	Scanty bile ducts, no canaliculi		
Haemangiopericytoma	F	1	Resection	Vascular channels surrounded by pericytes which appeared spreading out from the vessel walls.	$1.5 \times 10^{-1}$	3.7
Hepatoblastoma	M	4	"	All Epithelial type basically		
	F	13	"	Four foetal type		
	M	2½	"	One embryonal type		
	M	12	"	One mixed type	$9.6 \times 10^{-1}$	22.2
	F	9 mo	"	None undifferentiated type		
	M	1	"	In the mixed type, embryonal foetal & mesenchymal elements were seen.		
Neuroblastoma	M	6	Needle		$3 \times 10^{-4}$	7.4
	M	11 mo	"			
Fibrosarcoma	M	45	"			
	F	75	"	Had mastectomy for carcinoma breast and Cholecystectomy. Died three months after the diagnosis.	$3 \times 10^{-4}$	7.4
Leiomyosarcoma	F	65	Resection			
	M	45	"			
	F	22	"	Youngest recorded case in literature, was alive 6 months after the operation.	$4.8 \times 10^{-4}$	11.1
Melanoma	M	50	Needle	Primary could not be located anywhere else.		
	F	75	"			
	F	21	"	She had family history and was suffering from Xeroderma Pigmentosum.	$4.8 \times 10^{-1}$	11.1
Squamous cell Carcinoma	M	50	Needle	Well differentiated with epithelial pearl formation. Primary was not located else where.	$1.5 \times 10^{-1}$	3.7
Carcinoid with Cirrhosis	M	27	"	Primary could not be located else where	$1.5 \times 10^{-1}$	3.7
Neurofibrosarcoma	F	60	Needle	Interlacing bundles of fibres with tapering nuclei were arranged in palisades at places. Variations in the shape and size of the nuclei and loose areas were seen. Primary could not be located elsewhere.	$1.5 \times 10^{-1}$	3.7
Adenoid Cystic Carcinoma	M	45	Resection	Cystically dilated glands lined by cuboid cells were closely arranged. Primary could not be located elsewhere.	$1.5 \times 10^{-1}$	3.7

## DISCUSSION

The human liver as a consequence of its anatomic location, dual blood supply and size, is a favourable site for neoplastic lesions which are greater in number and diversity than in any other organ. The

advent of needle biopsy and other techniques has made diagnosis easier. In literature, unusual types of liver tumours are mostly reported as case reports<sup>1,4-16</sup> In the present series 27 cases of unusual liver tumours are reported their frequency among 625 primary liver tumours is 4.3% and among 5,308 total liver biopsies is 0.5% A frequency of 5.8% among 137 primary liver tumours was reported by El-Demori<sup>17</sup> compared to 43% reported by us. On the other hand a high frequency of benign liver tumours was reported by Edmondson<sup>18,19</sup>, hence in a series of 50,000 autopsies there were 285 (0.6%) hepatic tumours and out of these 200 (70%) were benign and 95 (30%) were malignant. Of the latter there was only one case of unusual type of malignant tumour.

### **BENIGN TUMOURS**

Benign tumours of the liver are rare comprising approximately 5% of all primary hepatic neoplasms<sup>20,21</sup>. In the present series the frequency is 1.1% of all the primary hepatic tumours. The commoner variants of benign neoplasms of the liver are (a) liver cell adenoma, (b) bile duct adenoma and (c) haemangioma. The less common ones are neurofibroma, neurilemmoma and haemangioendothelioma<sup>18-23</sup>. Liver cell adenoma resembles normal liver tissue but the cells are arranged in two to three cell wide trabeculae. Bile canaliculi are absent though bile ducts are present. It may or may not be capsulated. Wide variation of age is encountered. Those seen in females are often related to the long term use of the contraceptive pills<sup>23</sup>. In the present series none was of the latter type. Bile duct adenoma is also very rare and is usually discovered incidentally because of its small size (< 1cm)<sup>23</sup>. Wide variation of age is seen. Haemangioma, usually cavernous is the commonest of the benign tumours. Infantile haemangioendothelioma type—I & 2 are not related to polyvinyl chloride. Type—I shows endothelial lined spaces. The cells are single layered and cuboid. Type-2 shows multilayering and mitotic figures. The latter is often confused with angiosarcomas<sup>23</sup>.

### **MALIGNANT TUMOURS**

#### **Hepatoblastoma:**

Hepatoblastoma is the primary liver cell tumour of the childhood. Ishak<sup>23</sup> has described three variants viz. Epithelial (foetal and embryoma), Epithelial and mesenchymal and undifferentiated in which cartilage bone etc may be seen. Prognosis is best in epithelial (foetal) type. In an autopsy based study Clatworthy<sup>20</sup> reported 12 hepatoblastomas (0.7%) among 1,728 malignancies in children.

#### **Leiomyosarcoma:**

Primary leiomyosarcoma<sup>4,5</sup> is a rare lesion and only ten cases have been reported so far. Their ages ranged from 32 to 63 years and male to female ratio was 2:1. In the present series one of the patients was 21 year old (the youngest case reported so far) and male to female ratio was 1:2.

#### **Fibrosarcoma:**

Like primary leiomyosarcoma of the liver fibrosarcoma too, is an extremely rare neoplasm. Totzke,<sup>7</sup> Shallow<sup>8</sup> and Jaffe<sup>9</sup> have reported one case each and Simpson has reported three cases of primary fibrosarcoma of the liver. Amongst the reported cases, the ages of the patients ranged from 55 to 65 years and male to female ratio was approximately 1:1 Steiner<sup>24</sup> reported 0.1% frequency among 860 primary hepatic malignancies. In the present series the frequency is 0.3% among 619 primary hepatic malignancies.

#### **Melanosarcoma:**

Primary melanosarcoma of the liver is mentioned in the classification of liver tumours by Edmondson<sup>18</sup> and Spellberg,<sup>22</sup> however no case report was available in the literature. In the present series three cases are reported on the presumption that primary was not detected elsewhere.

#### **Squamous Cell Carcinoma:**

Diagnosis of this case was also based upon exclusion viz, primary was not located anywhere else. Though Edmondson has quoted primary squamous cell carcinoma in his classification on liver tumours but any published report on this subject could not be found.

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