Adult rhabdomyosarcoma of the gall bladder: case report and review of published works

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Abstract

A 90 year old woman with alveolar rhabdomyosarcoma of the gall bladder is described. The patient died with tumour metastasis four months after diagnosis despite complete excision of the tumour. The histological, immunohistochemical, and ultrastructural features of the tumour are summarised and published works are reviewed.

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The hepatobiliary system is an extremely rare site for the primary lesion of rhabdomyosarcoma. To our knowledge, rhabdomyosarcoma has been described in the gall bladder of three adult patients,¹⁻³ as reported in English language publications. Two of the cases were of the embryonal type,^{1,3} and one mixed embryonal and alveolar type.²

In this report we present a fourth case of rhabdomyosarcoma of the gall bladder in an adult and the first case of the pure alveolar type. The histological features, immunohistochemical findings, and ultrastructure of the tumour are also described.

Case report

A 90 year old woman was admitted to Princess Basma hospital in June 1991 with a two year history of right upper quadrant pain, which became continuous over the last month and associated with loss of weight and appetite. There was no history of jaundice. On examination she was found to have low grade fever, right upper quadrant tenderness, and a mass. Laboratory investigations showed: packed cell volume 29%, white blood cell count 12 800, and slightly increased bilirubin and liver enzyme activities. Ultrasound showed a gall bladder mass and multiple gall stones.

At laparotomy, a gall bladder mass 10×15 cm in diameter was completely excised. The common bile duct was found dilated and was explored, multiple common bile duct stones were removed, and a T tube was inserted. The liver seemed normal. A T tube cholangiogram showed retained common bile duct stones including some in the left hepatic duct (Fig 1). By the time endoscopic retrograde cholangiopancreatography was done, only one common bile duct stone remained and was extracted at the same time as sphincterotomy. Clinical investigations showed no evidence of a primary tumour in the genital system or in other sites in which rhabdomyosarcoma is known to occur as a primary tumour.

Two months later, the patient started to have recurrence of the right upper quadrant pain and progressive deterioration of her general health. One month later she presented with an enlarged left supraclavicular lymph node. Excisional biopsy was done. There was no further intervention upon the patient's request apart from analgesics until her death four months after her initial presentation. A necropsy was not performed.

Methods

Microscopic sections of formalin fixed tissue were prepared and stained with haematoxylin and eosin. For immunoperoxidase stains, 4 µm sections from paraffin wax embedded tissues were prepared using the avidin-biotin complex technique.⁴ The antibodies used included: cytokeratin (AE 1,3; Hybritech, Indianapolis, IN), carcinoembryonic antigen, epithelial membrane antigen, desmin and vimentin (Dako, Santa Barbara, CA), myosin and myoglobin (Biogenex, Dublin, CA), and muscle specific actin (Enzo, New York, NY). The chromogen used in this study was diaminobenzidine (Sigma, St Louis, MO). Representative sections of formalin fixed tissue were taken for electron microscopic examination.

Figure 1: T tube cholangiogram showing retained stones in the left hepatic and the common bile ducts.

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Review of published reports

Case report	Author/year	Patient		Histology	Survival
		Age	Sex		
1	Edmonson ¹ 1967	80	F	Embryonal	Unknown
2	Yasuma and Yanaka ² 1971	67	F	Embryonal and alveolar	45 Days
3	Willen and Willen ³ 1982	68	F	Embryonal	2 Months
4	Present case 1992	90	F	Alveolar	4 Months

Pathological findings

Macroscopic findings – a tumour mass 10×9×7.5 cm at greatest dimension was seen arising from the posterior wall of the gall bladder. Extensive necrosis and haemorrhage were present in the central regions of the tumour. The gall bladder was filled with stones.

Microscopic and immunohistochemical findings – as Figure 2 shows, the gall bladder epithelium seems intact over the tumour. Tumour cells were round polygonal cells with a high nuclear to cytoplasmic ratio and prominent nucleoli. The cytoplasm was eosinophilic and granular, however, and cross striations were not seen. The tumour cells formed nests surrounded by fibromuscular septa reminiscent of the solid variant of alveolar rhabdomyosarcoma. The results of immunohistological tests showed positive staining of tumour cells for vimentin and desmin; the other muscle markers – actin, myosin, and myoglobin – were negative. Cytokeratin, epithelial membrane antigen, and carcinoembryonic antigen were negative.

Ultrastructural examination showed tumour cells that lacked epithelial characteristics such as cell junctions, desmosomes, villi, and gland formation. No thin filaments, thick filaments or Z bands were seen.

Discussion

Carcinoma is the commonest malignant tumour of the gall bladder, while primary sarcoma of the gall bladder is a rare

occurrence.^{2 3} In adults, rhabdomyosarcoma of the hepatobiliary system is extremely rare.^{2 3} A review of English language published works showed three cases of adult gall bladder rhabdomyosarcoma,^{1–3} two of them of the embryonal type^{1 3} and one of the mixed embryonal and alveolar type (Table).² These three cases and this case report have been reported in women over 65 years. Jaundice has been reported in two of the cases,^{2 3} but jaundice was not found in our patient despite the presence of multiple common bile duct stones. One of the previous cases (patient number 3) had a right upper quadrant mass similar to our patient³ while another (patient number 2) was similar to our patient in having a left supraclavicular lymph node metastasis.² The longest survival was four months after the onset of symptoms (as reported in our patient) despite what seemed to be complete excision. All four patients had gall stones.^{1–3} Our patient, however, was the only one having common bile duct and hepatic duct stones. Gall stones have been reported to occur in about 80% of all sarcomas of the gall bladder.^{2 3} A similar case of gall bladder alveolar rhabdomyosarcoma have been reported in French published works,⁵ and one case of alveolar rhabdomyosarcoma have been reported in the common bile duct.⁶

In the paediatric age group, rhabdomyosarcoma of the biliary tree should be considered in the differential diagnosis of obstructive jaundice after choledochal cyst.⁷ It is usually of the embryonal (sarcoma botryoides) variety.^{7–13} Since the first reported case in 1875 by Wilks and Moxon,¹⁴ 52 cases of hepatobiliary rhabdomyosarcoma in the paediatric age group have been reported in the world publications by 1986.^{6 15} Forty five of these cases were in the biliary tree¹⁵ and one in the ampulla of Vater.¹⁶ A longer survival period has been reported recently, even in the presence of extensive tumour, in those patients given aggressive surgical treatment followed by radiation and chemotherapy.⁹

Although the histology of this tumour is that of a poorly differentiated malignancy, the positive vimentin staining in association with negative expression of epithelial markers (keratin, epithelial membrane antigen, and carcinoembryonic antigen) support the diagnosis of a mesenchymal tumour. The histological picture and positive desmin staining (Fig 2) confirm the diagnosis of alveolar rhabdomyosarcoma. In our case myosin and myoglobin were not expressed but less differentiated rhabdomyosarcoma can lack expression of these two antigens. It should be emphasised that desmin, expressed in our case, is the most sensitive marker for rhabdomyosarcoma.¹⁷

Rhabdomyoblastic differentiation may also represent part of the sarcomatous component of carcinosarcoma or malignant mixed tumour of the gall bladder.¹⁸ Twenty four cases of carcinosarcoma of the gall bladder had been reported in the world publications by 1990, but only in three of them has

Figure 2: Rhabdomyosarcoma cells seen beneath an intact gall bladder mucosa (upper left; haematoxylin and eosin ×100). Tumour cells form nests surrounded by fibrovascular septa, solid alveolar pattern (upper right; haematoxylin and eosin ×400). Positive desmin and vimentin staining of tumour cells in lower left and lower right pictures respectively (avidin-biotin complex technique ×400).

rhabdomyosarcomatous differentiation of a mesenchymal component been reported.¹⁸⁻²⁰

We do not believe that our case belongs to this group of tumours, as the ultrastructural examination failed to show any epithelial differentiation and the immunoperoxidase stains showed negative expression of all epithelial markers used.

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