Turner's Syndrome With Hepatocellular Cancer And Ulcerative Colitis: A Case Report

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Citation

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Abstract

Turner's syndrome (45xo) is associated with various autoimmune disorders and malignancies ; in particular colon cancer, germ cell tumour, and cholangiocarcinoma have been reported[4]. We report a case of Turner syndrome that presented with colitis but succumbed due to bleeding inside hepatocellular carcinoma.

CASE REPORT

A 28 year old Turner's syndrome patient was admitted with a three week history of fresh rectal bleeding . This bleeding was accompanied with mucus and a watery stool about 5-6 times per day. Other associated symptoms included colicky lower abdominal pain, occasional vomiting, nausea and decreased appetite. Her previous medication included oestrogen.

On examination, the patient had a tachycardia of 110/min and abdominal examination showed a diffuse lower abdominal tenderness without guarding and rigidity. Rectal examination revealed brown stools. Blood results showed a HB-9.6g/dl, WBC-15.5x109/l, CRP-134, Bilirubin-4umol/l, ALT-14iu/l, ALP-434iu/l(normal valuesofALP-50-270iu/l). A rigid sigmoidoscopy revealed florid proctitis and a biopsy was taken. She was commenced on mesalazine , ferrous sulphate and oral prednisolone.

Over the next seven days she had very little bleeding, but diarrhoea continued about 4-5 times a day. Repeat haemoglobin was 10.5g/dl . On the ninth day following her admission she had a unexpected cardio-respiratory arrest from which she failed to be resuscitated.

Histology of the rectal biopsy showed a chronic proctitis with ulceration; no granulomata were identified and the picture was consistent with ulcerative colitis .

The post-mortem report revealed a florid ulcerative colitis of the left colon. An unexpected finding was of a highly vascular large hepatocellular cancer along with some fatty change (Figure 1 & Figure 2),which was pointed as contributory to Cardio-respiratory arrest.

Figure 1

Figure 1 :A well circumscribed neoplasm in the liver has a thick fibrous capsule . The adjacent liver shows fatty change but no evidence of Hepatitis B infection & no cirrhosis.(H & E x 4 objective)

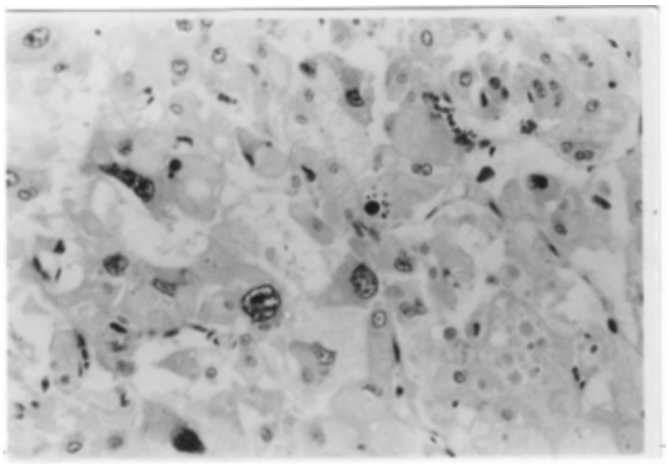
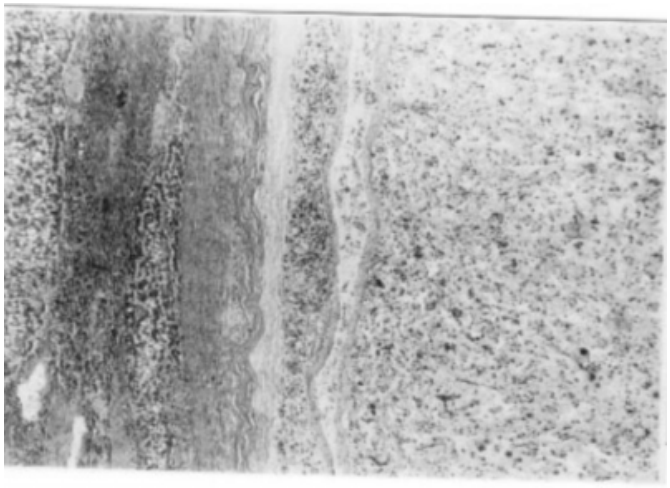


Figure 2

Figure 2 : At high power, the tumor cells have markedly pleomorphic nuclei and copious eosinophilic ; hepatocyte bile cytoplasm. ntervening Sinusoids and foci of bile formation are present. These features are mostly in keeping with a Primary Hepatocellular Cancer. (H & E x 25 objective)



DISCUSSION

Turner's syndrome is a chromosomal disorder (45xo) affecting 50 per 100,000 females. It is characterised by growth retardation, ovarian dysgenesis and infertility and is commonly associated with coronary artery disease and coarctation of aorta. The incidence of inflammatory bowel disease is considerably higher than the reported frequency of new cases of inflammatory bowel disease in the general population ([1]). An apparent association with a karyotype abnormality which includes a structurally abnormal X chromosome is noted. ([1],[4]).

Abnormalities of liver function tests have been reported in adults with Turner's syndrome ([2]). The prevalence of cryptogenic liver disease in patients with Turner's syndrome seems to be relatively high ([2]). A study demonstrating biochemical liver cholestasis has been reported in sixteen women all of whom had previously been treated with estrogen.

The authors however concluded that the aetiology remains uncertain ([3]). In patients with liver abnormalities there is no long-term study to report progression to any serious liver disease. These patients might also demonstrate a particular karyotype abnormality. This rare but significant risk of Turner's syndrome developing liver abnormalities would raise the suggestion of some sort of surveillance (possibly regular liver function tests, ultrasound examination of liver) so that hopefully any abnormality can be picked up at an early stage, where treatment might be possible. Further long-term studies are needed to justify this.

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