THE BERNARDSOULIER SYNDROME – REPORT OF A CASE

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The Bernard-Soulier Syndrome is an uncommon congenital disorder of platelet function. The clinical findings and detailed laboratory investigation of a case is presented together with family studies, review of the literature and a brief review of other congenital platelet function defects.

SERUM IGM-GLOBULIN IN MALAYSIAN NORMAL AND DEFECTIVE INFANTS

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The IgM-globulin levels were determined for 5,167 cord-sera of apparently normal infants and 281 sera of defective infants aged 4 months and younger. The significant level for IgM-globulin in neonates was found to be 20 mgm/dl (2 S.D. above mean of the normal) above which the level was regarded as abnormally raised.

Significant levels of IgM-globulin were found in 0.2% (11/5,167) of normal neonates and in 40.6% (114/281) of defective infants which is more than 200 times the normal value. Combining the normal and abnormal rates, an overall figure of 23 intra-uterine infections per 1,000 live births were obtained for Malaysia.

The advantages and disadvantages of the use of serum IgM-globulin elevations for the diagnosis of intrauterine infections will be discussed.

SEROLOGIC MARKERS AND LIVER-FUNCTION STATUS IN ASYMPTOMATIC HEPATITIS-B SURFACE ANTIGEN CARRIERS

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Hepatitis-B virus infection is characterised by several distinctive serologic and immunologic responses which serve as useful guides for monitoring the course of the condition. Among these markers, the e-antigen and e-antibody are believed to be of value in determining the degree of infectivity and the prognosis of the infection.

In the present study, normal blood donors who are positive for the hepatitis-B surface antigen (HBsAg) are followed up in our outpatient department. The presence of the e-antigen and e-antibody, and the liver function profile are studied in these asymptomatic carriers of the HBsAg.

Preliminary results of the study indicate that about 30% of the subjects are positive for the e-antigen, and about 60% are positive for the e-antibody. The rest of the carriers are negative for both markers. Abnormal liver function results are observed in 25 – 30% of the cases, most of whom show increased levels of serum transaminases. Approximately 40% of the e-antigen positive patients show abnormality of liver-function, in contrast to about 20% of the e-antigen negative subjects.

AN INVESTIGATION OF THE PROTEIN COMPONENTS OF AMYLOID USING THE IMMUNOPEROXIDASE METHOD ON TISSUE SECTIONS

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It is known that the major protein component of the amyloid fibril differs in the various types of amyloidosis. We have investigated for the presence of AA protein, K immunoglobulin light chains and calcitonin in amyloid deposits using the indirect immunoperoxidase and the peroxidase-anti-peroxidase methods. These staining techniques were applied to histological sections from formalin-fixed, paraffin-embedded tissues containing amyloid from patients with generalised primary and secondary amyloidosis and localised amyloidosis associated with a variety of epithelial and endocrine tumours. AA protein was consistently detected in deposits of secondary amyloidosis and calcitonin was demonstrated in amyloid deposits within Medullary carcinoma of the thyroid. Staining for light chains, particularly >-chains, was positive in some deposits of primary amyloidosis and tumour-associated amyloidosis.
REVIEW OF OSTEOSARCOMA WITH RADIOLOGICAL CORRELATION

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Tumour tissue from 32 cases of histologically proven osteosarcoma received by the Department of Pathology, University Hospital over a 12-year period, from the year 1969 to 1981 were reviewed. The lower limb was the most commonly affected site and the femur was the commonest bone involved. The patients were mainly young males between the ages of 15 to 25 years. The youngest patient in this series was a 10-year old male child. The most common histological type was the osteogenic type, but other types encountered included parosteal osteogenic sarcoma.

The histological findings and the value of radiographs in the diagnosis of osteosarcoma will be discussed.

LYMPHOMA OF THE STOMACH

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All gastric biopsy specimens received at Pathology Department, University Hospital, Kuala Lumpur, over a 5-year period, from June 1978 and May 1983, were reviewed. There were 6 cases of lymphoma of the stomach; 3 cases underwent gastrectomy and chemotherapy. One of the cases showed lymphoma associated with non-caseating epitheloid granulomata, in the absence of clinical and radiological evidence of tuberculosis. The pathological features clinical presentation, management and prognosis are discussed in the context of current available literature.

PATTERNS OF EXPOSURE, CLINICAL MANIFESTATIONS AND SEQUENTIAL SEROLOGICAL PROFILE OF HEPATITIS B VIRUS MARKERS IN MEDICAL DOCTORS

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The pattern of exposure, clinical manifestation and sequential serological profile of hepatitis B virus (HBV) in the sera of ten medical doctors at the General Hospital, Kuala Lumpur exposed to the virus were studied. The source of infection of five of the doctors was known. Of the ten doctors exposed, seven (70%) developed acute hepatitis with jaundice while three (30%) had subclinical infection. Of the seven who had acute hepatitis, two (28.5%) developed chronic hepatitis while the rest finally recovered with the formation of antibodies. The reason why some individuals develop chronic hepatitis is discussed.

SQUAMOUS CELL CARCINOMA ARISING IN AN EPIDERMID CYST – A CASE REPORT AND REVIEW OF LITERATURE

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A 59-year old Indian lady was admitted to the University Hospital for a swelling on the left thigh, clinically diagnosed to be an Epidermoid cyst. Local excision was done and histopathological examination showed a squamous cell carcinoma arising from an Epidermoid cyst. Subsequently, the patient underwent wide local excision and inguinal block dissection and the lymph nodes revealed metastatic involvement. From the review of available literature, this appears to be an extremely rare occurrence. The histology and the significance of this finding are briefly discussed.
PROGNOSTIC FEATURES IN RETINOBLASTOMA

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In the 13-year period between January 1968 and December 1982, 30 children with retinoblastoma were admitted to the University Hospital, Kuala Lumpur. Pathological examination of the enucleated eyes were done in 26 cases. All the patients presented with either stage V or VI disease. 20% of them died, 40% are still alive and 40% were lost to follow up. The significance of pathological staging and correlation between staging and histological features are discussed.

CLINICO-PATHOLOGICAL STUDY OF 30 CASES OF PAPILLARY CARCINOMA OF THYROID FROM EAST MALAYSIA

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246 cases of thyroid surgical specimens from East Malaysia were seen at the Department of Pathology, University Hospital from 1981 – 1982. Of these 30 cases (12.2%) of papillary carcinoma of thyroid were diagnosed. The age, sex and racial distribution of these cases are outlined. Clinically these cases presented as solitary thyroid nodules (20 cases), thyroid cysts (4 cases), thyroid nodule with palpable cervical lymph nodes (3 cases) and palpable cervical lymph nodes (3 cases). Histological confirmation of papillary carcinoma of thyroid was done in all cases and the pathological features discussed.