Wilms's tumour and neurofibromatosis

The association between Wilms's tumour and certain congenital anomalies is well documented. An increased incidence of aniridia, hemihypertrophy, and abnormalities of the genitourinary tract has been described.¹ We report the case of a child with Wilms's tumour and neurofibromatosis.

Case report

A 3-year-old Negro girl was admitted on 10 June 1975 with a six-month history of a swelling in the abdomen. Physical examination showed widespread café-au-lait spots over the chest, trunk, and arms (see figure) and a swelling on the wrist thought to be a neurofibroma. A large smooth mass

The patient, showing widespread café-au-lait spots.

was palpable in the left hypochondrium. The patient's father, a paternal aunt, and the paternal grandfather all had neurofibromatosis. An intravenous pyelogram showed distortion of the left kidney with displacement by a soft mass extending across the mid-line. The chest x-ray film and a skeletal survey were normal. Vanillylmandelic (VMA) acid excretion was within normal limits. On 20 June, a large nephroblastoma (15.5 × 10.5 cm), together with the left kidney and 6.5 cm of the ureter, were removed; to the naked eye the tumour had not extended beyond the capsule of the kidney. Between July 1975 and July 1976 she received 11 courses of chemotherapy with Actinomycin D and vincristine. She remains in remission with no evidence of recurrence.

Discussion

Wilms's tumour is the second commonest intra-abdominal neoplasm in childhood: the incidence is roughly one case per 200 000 children per year.² It is more common in patients with certain developmental anomalies, occurring in about a third of patients with congenital aniridia.³ Ipsilateral or contralateral hemihypertrophy is also commonly associated with this tumour. Other urinary tract abnormalities have been described in Wilms's tumour, including hypospadias, cryptorchidism, horseshoe or multicystic renal disease, duplicated ureters, renal hypoplasia, and pseudohermaphroditism.

Neurofibromatosis occurs in 1 per 3000 live births and is characterised by café-au-lait spots and multiple neurofibromas. Inherited as an autosomal dominant characteristic of variable expressivity, it forms one of the group of hamartomatous disorders in which there is

localised excess of growth and occasionally neoplastic change. There is an increased incidence of gliomas, acoustic neuromas, meningiomas, and pheochromocytomas. Familial aggregations of tumours of neurogenic origin have been described but only a few reports cite an association between Wilms's tumour and neurofibromatosis.^{1, 4} Although the incidence of congenital anomalies is much greater in bilateral cases of Wilms's tumour,³ the occurrence of two neoplasms in which a genetic influence is particularly strong is probably more than coincidental. Familial cases of Wilms's tumour have been described without expression in the parent and this has been ascribed to a delayed mutation, possibly viral-induced.⁵

The importance of excluding an underlying Wilms's tumour in any child who presents with one or more of these distinctive congenital anomalies deserves wider recognition in view of the fact that between 80-90% of early cases are now curable.

Requests for reprints should be sent to KDB.

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Body weight correlates with REM sleep

The fact that REM (rapid eye movement) (paradoxical) sleep is a time when dreaming occurs¹ has led to a neglect of somatic functions at this time and to unsuccessful attempts to relate an individual's personality traits to his characteristic amount of REM sleep. To obtain a reliable measure of the latter it is important to allow adaptation to the sleep laboratory, to record a large number of nights, preferably consecutive, and to allow adequate time in bed each night.

Methods and results

Sixteen healthy volunteers aged 52-67 (mean 59 years), six men and 10 women, slept in the laboratory every four weeks during a 16-week period and always did so for six consecutive nights, of which the first was for adaptation. Lights-out was from 2215 to 0700 h. At the start and end of each week of attendance they were weighed in light clothing. None of the volunteers were receiving drugs affecting the nervous system and they were asked to abstain from alcohol throughout. The 320 electrophysiological records were scored in terms of the usual international criteria. The amounts of each sleep stage on each night were calculated and also, for each subject, the means for his or her 20 nights. The means of the eight weight measures were also calculated.

It was found that body weight and the percentage of total sleep that was spent in REM sleep were related. Spearman's rank correlation test was employed and $r_s = 0.774$, $t = 4.57$, $P < 0.001$, 2-tailed. Log body weight was also correlated with mean total number of minutes of REM sleep (with a product-moment correlation, $r = 0.643$, $t = 3.07$, $p < 0.01$, 2-tailed). The figure illustrates the simple correlation between body weight and percentage of REM sleep. No correlations were found between body weight and total sleep, slow wave sleep or other sleep variables.

Discussion

When patients with anorexia nervosa gained weight the most significant change in their sleep lay in an increase of REM sleep,² while in a study of acute starvation REM sleep fell significantly.³

In rats the amount of REM sleep in a 12 h period accurately predicted food intake in the subsequent 12 h.⁴ These observations are consistent with the present finding and since log body weight is highly correlated with the daily metabolic rate⁵ it seems likely that REM sleep is closely linked to general body metabolism and restitution.

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Pseudotumours of the pancreas

A mass felt in the pancreas at laparotomy is normally regarded as a tumour and, indeed, it usually is. Three tumour-like lesions of the pancreas have recently been encountered in this hospital, one of which was treated by a pancreaticoduodenectomy. All three showed the same distinctive histology, which does not appear to have been reported.

Case reports

Case 1—A 46-year-old man was admitted for surgical treatment of a chronic duodenal ulcer and gall stones. At laparotomy the presence of a duodenal ulcer and of gall stones was confirmed, but there was also a discreet hard mass in the head of the pancreas, which was considered to be a carcinoma. The liver was abnormal and a wedge biopsy was taken. A pancreaticoduodenectomy was performed with a cholecystectomy. Recovery from the operation was uneventful. Pathological examination showed a well-defined ovoid mass 1.6 × 0.8 cm on the superior border of the head of the pancreas. Microscopically, the lesion consisted of lobulated connective tissue enclosing irregular, branching pancreatic ducts, acinar tissue, and islet cells in a disorderly arrangement (figure). Multiple blocks taken from other areas of the pancreas showed a few tiny nodules of similar appearance. There was

no evidence of pancreatitis. The liver biopsy taken at operation showed partial nodular transformation.

Case 2—A 35-year-old man gave a five-year history of recurrent attacks of epigastric pain due to chronic pancreatitis. An endoscopic retrograde pancreatogram showed a blocked pancreatic duct. At laparotomy the tail of the pancreas was removed and an antegrade pancreatogram, which showed a normal pancreatic duct, was performed. Histology of the tail of the pancreas showed mild chronic pancreatitis with intra- and interlobular fibrosis and patches of old fat necrosis. There were also multiple discreet nodules of duct proliferation surrounded by fibrosis similar to those seen in case 1.

Case 3—A 58-year-old man died of disseminated malignant lymphoma. There had been no symptoms or signs of gastrointestinal or pancreatic disease. At routine necropsy a 1.0 × 0.5 cm solitary nodule was found in the head of the pancreas. Histological examination of this lesion showed a lobulated mass of loose connective tissue enclosing dilated pancreatic ducts with acinar and islet cells. The lesion was again well circumscribed and it appeared to be associated with stenosis of a major duct.

Pseudotumour of the pancreas (case 1): a well-defined mass made up of lobulated connective tissue enclosing ducts and acinar and islet cells in complete disarray. (Haematoxylin-eosin. × 5, insert × 50.)

Discussion

Histology of the lesion in all three cases showed lobulated connective tissue surrounding acinar cells and pancreatic ducts. In cases 1 and 3 islet cells were present. The general appearances suggested a malformation or hamartoma, and, moreover, in case 1 the liver showed partial nodular transformation—a condition also considered to be hamartomatous.¹ A computer-aided search of published reports has failed to find a report of pancreatic hamartomas other than those which are lymphovascular in origin and are readily detectable as such.^{2,3} Cases 2 and 3 were associated with duct stenosis, and possibly this may lead to pancreatitis which then may be focal or generalised, depending on the level and size of duct affected. Pancreatitis was seen in case 2 but was absent in cases 1 and 3. For the time being, the most appropriate designation of these lesions would be "pseudotumour."

The condition may be commoner than the absence of documentation suggests, and may be an important source of diagnostic error at laparotomy. Indeed, some of the long-term survivors after palliative procedures for carcinoma may have had a pseudotumour. Such lesions should be easily distinguished by a thin needle biopsy, which can be performed safely at the time of operation.⁴

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