

the diagnosis of anterior sacral meningocele.

The 'Scimitar sign' seen in approximately half the cases of anterior sacral meningocele is produced by a unilateral sacral defect with a smooth margin simulating the shape of the old Arabic sabre(5). Around 20% of patients, however, have a midline sacral defect with the lateral wings of the sacrum draping either side of the defect(6). Ultrasonography, CT scan and magnetic resonance imaging (MRI) may be used to demonstrate the extent of the meningocele, characteristics of the sac and the width of the stalk(1,7).

The surgical approaches to presacral meningocele include the posterior trans-sacral by laminectomy, the perineal and the anterior transabdominal approach. The posterior approach allows identification and preservation of the nerve roots but water tight closure of the pedicle may not be possible(7). The anterior trans-abdominal approach is more straightforward and offers better access to the neck of the sac although there is more potential risk of damage to nerve roots. In our patient exposure as well as excision of the meningocele was entirely satisfactory using the trans-abdominal approach.

In conclusion, although anterior sacral meningocele is an uncommon cause of constipation in early childhood it should be kept in the differential diagnosis whenever a child presents with constipation. The simple but often omitted per-rectal examination and a plain skiagram showing the characteristic sacral bony deformity are sufficient to diagnose this unusual congenital condition.

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Congenital Hemihypertrophy and Wilms' Tumor

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Congenital idiopathic hemihypertrophy has been reported to be associated with a

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number of intra-abdominal malignancies, including Wilms' tumor, hepatoma and adrenal cortical neoplasia(1). The present report describes a child with congenital hemihypertrophy in whom a Wilms' tumor was diagnosed on follow up and stresses the necessity of a suitable screening regimen for patients with this anomaly.

Case Report

A male child, born at term with a birth weight of 3.5 kg, was apparently normal till the age of 2 months when a prominent hypertrophy of the left upper and lower limbs (Fig. 1) was noted on a routine follow up examination. The left femur and humerus were longer than the right on roent-

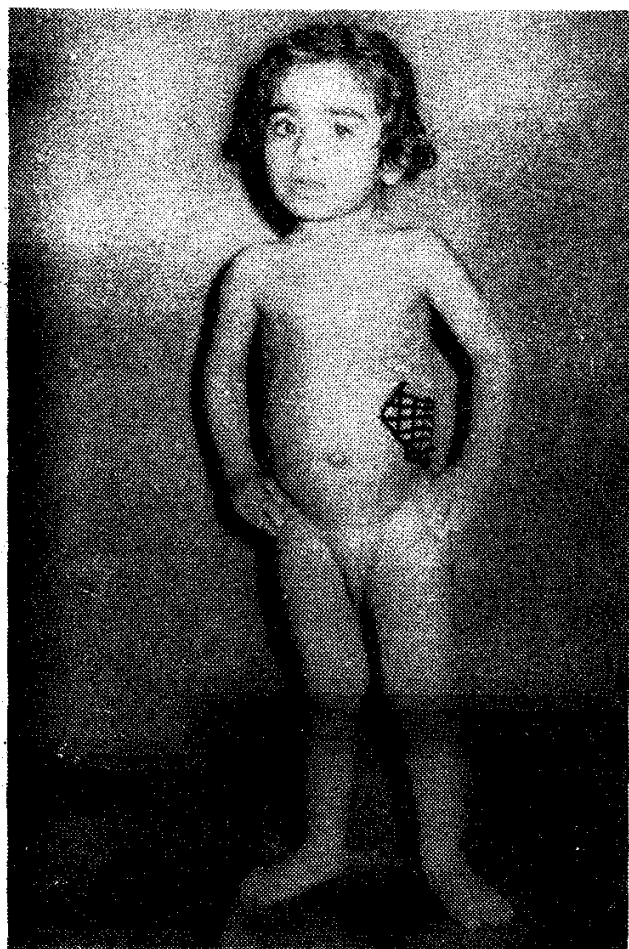


Fig. 1. Hypertrophy of left upper and lower limbs.

genographic examination. With a diagnosis of hemihypertrophy, the patient was called every 6 months for a detailed physical examination and ultrasound study of the abdomen, keeping in mind the known association of intra-abdominal malignancies. The child did well during the first 26 months of regular follow up except for occasional minor illnesses. At the age of 33 months, he was brought to the hospital with colicky abdominal pain and was found to have severe hypertension (BP 180/130 mm of Hg) and a large intra-abdominal mass (10×11 cm) in the left lumbar region (Fig. 1) which was bi-manually palpable and ballotable. An abdominal ultrasound examination revealed that the middle and lower pole of the left kidney were replaced by a mass of mixed echotexture, extending upto the midline. The right kidney and liver were normal. The inferior vena cava and left renal vein were patent. Renal function tests and urine routine examination were within normal limits. A diagnosis of left sided Wilms' tumor was made and after controlling the hypertension with α -methyl dopa, captopril and nifedipine, a left radical nephroureterectomy was performed. Fig. 2 shows the gross appearance of the resected specimen. A diagnosis of Wilms' tumor with lymph node metastasis was confirmed histopathologically and the tumor categorized as Stage III. He received external left flank radiotherapy (2000 cGy delivered over a period of 4 weeks) followed by chemotherapy with vincristine, Actinomycin-D and Adriamycin according to DD-NWTS-III regimen for a total of 60 weeks. He remains disease free for over 24 months.

Discussion

Congenital idiopathic hemihypertrophy

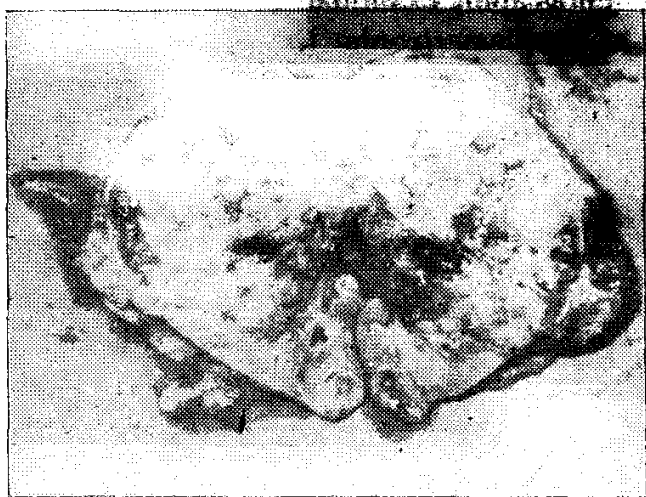


Fig. 2. Gross appearance of resected specimen.

is an uncommon condition but is associated with a high incidence of malignant and nonmalignant abdominal masses. In 1965, Ringrose *et al.* reviewed 129 cases of hemihypertrophy and found associated serious pathological conditions like Wilms' tumor and adrenal tumor in as many as 5% of the patients(2). If this incidence is valid, then it would be prudent to screen all patients with hemihypertrophy with a combination of clinical, radiographic (IVP) and sonographic techniques at regular intervals. In addition, other biochemical markers like α -fetoprotein, β -subunit of human chorionic gonadotrophin (HCG), androgen dihydroepiandrosterone and plasma cortisol may require monitoring. Abdominal ultrasound with special attention to the kidneys, liver and adrenal glands has been found to be a useful non-invasive modality for detection of intra-abdominal masses and Wilms' tumor in particular, even before it becomes clinically apparent. Sonographic techniques have proved to be superior to routine urinalysis, biochemical studies and radiographic techniques like IVP in this regard in patients with congenital hemihypertrophy(3). Moreover, it distinguishes benign nephromegaly, also seen

in hemihypertrophy, from Wilms' tumor by demonstration of altered pelvicalyceal pattern. Thus, routine physical examination and ultrasound study of the abdomen at regular intervals, as was done in our patient, is suggested for early detection of intra-abdominal masses in all such patients. Follow up however, should be more frequent, preferably at 3 monthly intervals as is quite obvious from our patient in whom development of Wilms' tumor could be detected only when it was already in an advanced stage (Stage III) inspite of 6 monthly follow up, which is the follow up programme suggested by Tolchin *et al.*(3). Although, the likelihood of developing tumor decreases with age, patients with hemihypertrophy can develop more than one intra-abdominal neoplasm(4). Therefore, the present patient, who has already had a Wilms' tumor will continue to undergo surveillance for other tumors.

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