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Case report

Renal dysplasia of the sacral region: Metanephric dysplastic hamartoma of the sacral region

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Summary. We believe this to be the third reported case of an unusual congenital sacral tumor-like lesion characterized histologically by the presence of dysplastic and immature renal-like tissue with a predominant glomerular and tubular structure.

This lesion appears important in the differential diagnosis of extrarenal nephroblastoma, and should be regarded as a dysplastic overgrowth.

An origin from mesonephric or metanephric remnants is suggested.

Key words: Sacral region – Retroperitoneum – Kidney, congenital abnormalities – Kidney, dysplasia – Kidney, neoplasms

Heterotopic renal tissue, a finding distinctly different from kidney ectopia, is not a new occurrence. Gruenwald in 1942 described a group of glomeruli and tubules in the presacral region of a 20 mm human embryo. His observation was doubted by Willis (1958).

Renal tissue was reported in the gubernaculum of the testis by Chevassu. Milliser et al. described the incidental histological finding of heterotopic renal tissue in the adrenal gland and heart (1969, 1972). Immature renal structures are present in sacrococcygeal teratomas (Gonzales-Crussi et al. 1978; Valdisserri and Yunis 1981; Ward and Dehner 1974).

In 1980 Cozzutto and Lazzaroni-Fossati described an unusual and huge sacral-perineal mass in a male newborn characterized by histological findings compatible with renal dysplasia. In 1976 Drake et al. contributed a very brief report of a 3-month-old female infant with a small lipomatous mass of the sacral region, which histologically revealed immature renal tissue.

We have had recently the opportunity to study an additional case very similar to that reported by Drake et al. (1976).

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Case report

A 3-year-old female child was referred in 1981 to the local hospital for a congenital subcutaneous well demarcated nut-sized mass in the sacral region. The small tumor was soft, covered by normal skin, attached to the sacrum, and asymptomatic. The remaining clinical examination of the patient was entirely negative with anatomically normal external genitalia and absence of palpable abdominal masses. Specifically, urograms demonstrated normally shaped kidneys, ureters and bladder, with a normal excretory activity. Abdominal tomography was noncontributory. Radiographic survey of the vertebral column and thorax was negative. The mass did not increase in size until it was removed at 3 years of age. Clinical examination of the child 2 years after the excision was negative.

The round fibrolipomatous mass measuring 3 cm in diameter was easily excised.

Histology

The mass consisted of abundant fibrous dense tissue and cellular adipose tissue. In both tissue types numerous glomerular and tubular structures were either scattered or, more often, clustered (Fig. 1). The glomeruli had a primitive appearance characterized by segmentation into scanty lobules and by plump cuboidal epithelium with dark nuclei (Fig. 2). Many glomeruli were large and irregularly shaped, with a coarse mesangial stalk and scanty capillaries. Some of them were markedly dilated and had multiple distinctly separated capillary tufts, abutting on the cavity (Fig. 2).

Others had a remarkably bizarre appearance, characterized by capillary loops appearing as large frond-like structures which contained another smaller glomerulus (Fig. 3).

The glomeruli were clustered in intimately packed groups from 3 to more than 15, some of them merging into a fused polylobated Bowman's capsule (Figs. 2 and 3). Several glomeruli were hyalinized. Many tubules were mature and covered with a low cuboidal or cylindrical

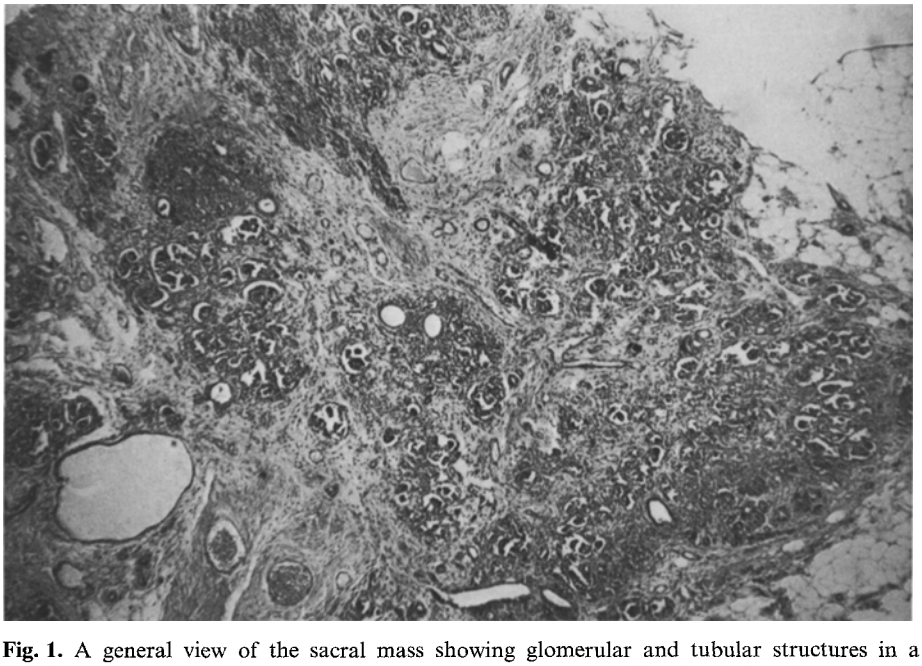

Fig. 1. A general view of the sacral mass showing glomerular and tubular structures in a lobular arrangement, thick fibrous septa, adipose tissue, and dilated tubules. Haematoxylin and eosin, $\times 40$

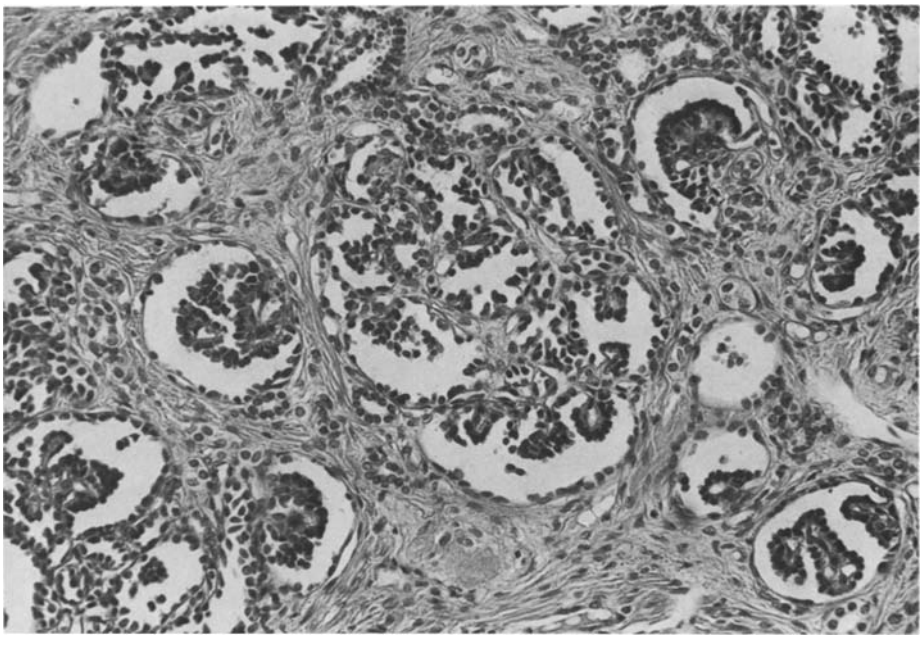

Fig. 2. Several confluent glomeruli. Note the immature glomerular appearance, the cuboid epithelial lining and the scanty capillaries. Haematoxylin and eosin, × 160

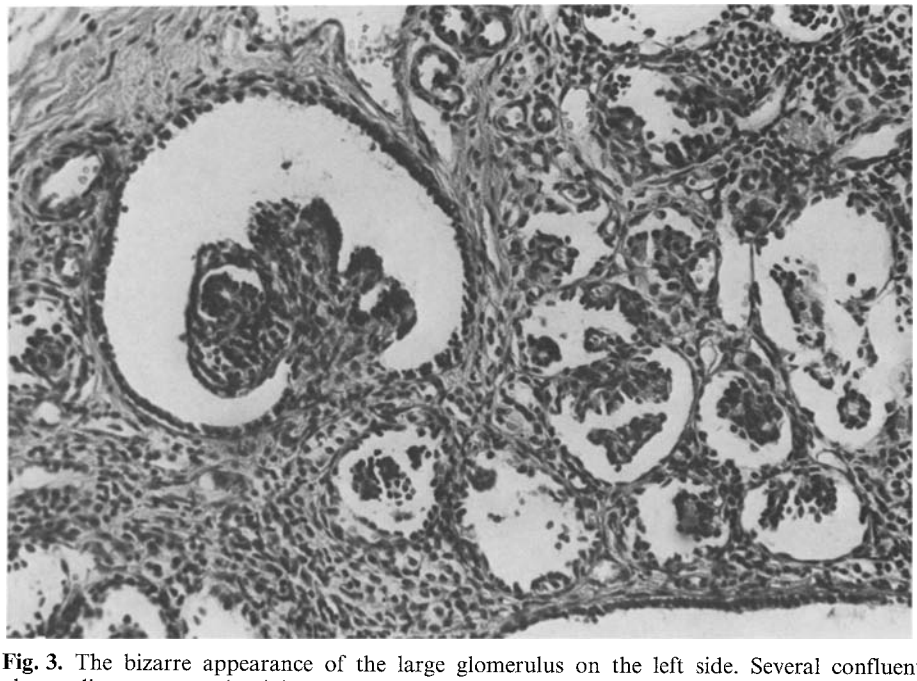

Fig. 3. The bizarre appearance of the large glomerulus on the left side. Several confluent glomeruli are seen on the right. Haematoxylin and eosin, × 160

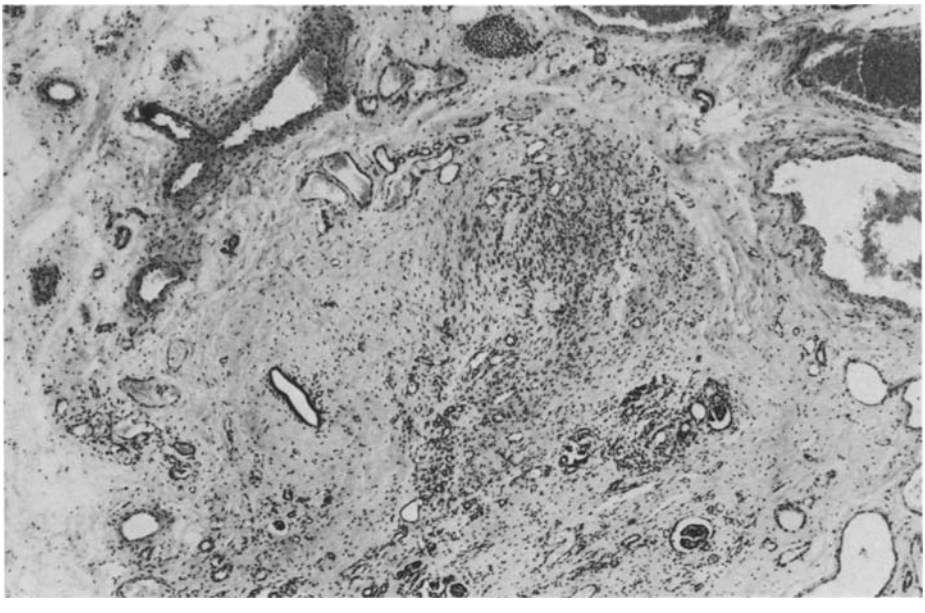

Fig. 4. A large area with dysplastic features. Immature glomeruli and tubules and dilated tubules surrounded by fibrous tissue can be seen. A small cartilage focus is seen at the top center of the picture. Haematoxylin and eosin, ×40

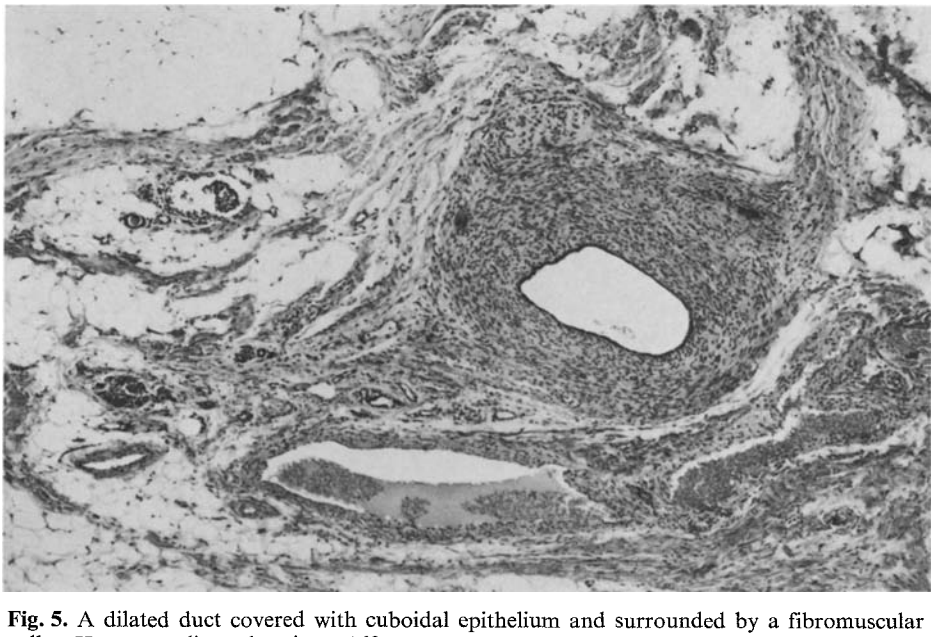

Fig. 5. A dilated duct covered with cuboidal epithelium and surrounded by a fibromuscular collar. Haematoxylin and eosin, ×160

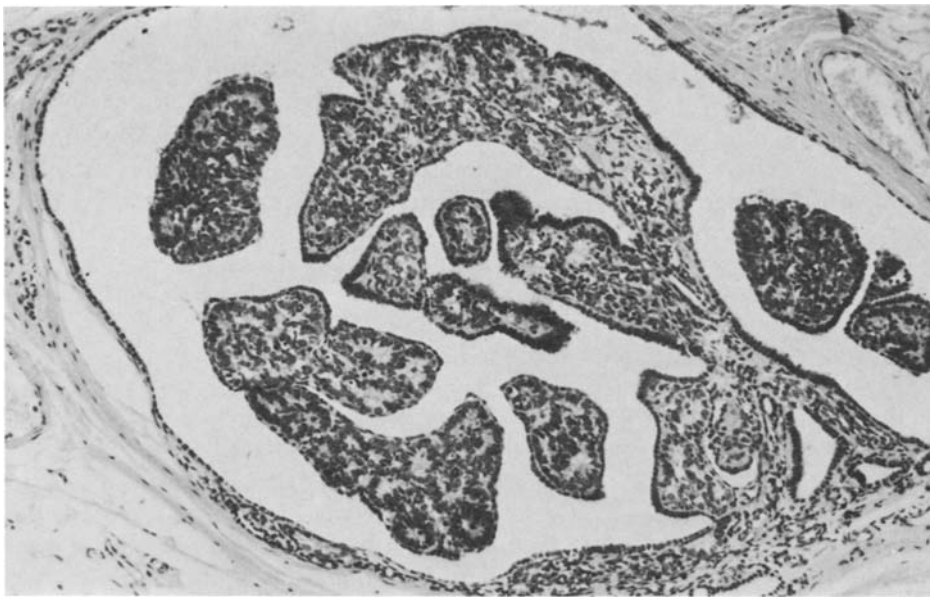

Fig. 6. Papillary folds resembling mesonephric structures. Haematoxylin and eosin, ×160

monolayered epithelium. Some were dilated and contained hyaline casts inside the lumen. Many other tubules were small, immature, and inconspicuous, showing an apparent lumen.

An interesting finding was the presence of intertubular stroma containing cords and rows of small cells which in some places imitated the small tubular structures which indicated the early stage of tubular formation.

Dilated ducts encircled by a thick mesenchymal collar and scattered nests of hyaline cartilage, two findings which are characteristic of renal dysplasia, were also present (Figs. 4 and 5).

Smooth muscle fibres, nerve bundles, foci of mucin and calcospherites were noted inside the stroma, as well as small scattered cysts covered by low cuboidal epithelium.

A final observation was a small focus of primitive looking pseudotubular structures of embryonal appearance allocated inside papillary folds (Fig. 6). These structures were quite similar in appearance to the human embryonal mesonephros. Very rare small immature tubules of embryonal appearance similar to those found in Wilms' tumor were seen intermingled with glomeruli and more mature tubules.

Discussion

The overall features of this case strongly support the concept of renal dysplasia. This diagnosis is specifically sustained by the following findings: 1) the presence of primitive dilated ducts surrounded by concentric mantles of smooth muscle; 2) numerous primitive tubules and glomeruli; 3) nests of hyaline catilage (Ericsson and Ivemark 1958a and b; Vellios and Garrett 1961; Bernstein 1974; Kissane 1974 and 1975; Boccon-Gibod and Petit 1975). The dilated primitive ducts encircled by a fibromuscular collar and the cysts covered by cuboidal epithelium represent altered derivatives of

the mesonephric duct from which they originate through the ureteric bud. The immature glomeruli and tubules derive from the metanephric blastema. Therefore, derivatives from both the metanephric blastema and mesonephric duct coexist in the present case (McCrory 1972; Kissane 1974 and 1975).

The case of a huge congenital infiltrating mass of the sacral and perineal region, reported in 1980 by Cozzutto and Lazzaroni-Fossati, was characterized histologically by a more abundant fibrous stroma with less abundant tubules and rare glomeruli. In contrast, the present case reveals a striking number of glomeruli which interestingly demonstrate several bizarre morphological appearances. Nevertheless, both cases represent a dysplastic overgrowth of renal tissue in the sacral region.

Mostofi, commenting on the case described by Cozzutto and Lazzaroni-Fossati, expressed the view that the observed lesion type was more than a dysplastic overgrowth of mesonephric or metanephric derivation and could represent a tumor-like overgrowth of renal-like tissue.

In 1976 Drake et al. briefly described a case of ectopic renal tissue in the sacral region, but data regarding morphological features were not detailed, thus confirming only the clinico-pathological relevance of this lesion.

All the three cases reported to date have been congenital sacral masses (Drake et al. 1976; Cozzutto and Lazzaroni-Fossati 1980). It may be suggested that this type of lesion is the result of detachment and consequent dysplastic growth of an embryonal focus originating from the metanephros. This contention is sustained by the finding of an embryonal focus resembling mesonephric structures. It might also be argued that this process represents the development of a focal or segmental renal dysplasia in the sacral region. The presence of very rare immature tubules of embryonal appearance in our case confirms the existence of a relationship between Wilms tumor and renal dysplasia, as recently reviewed by Marsden and Lawler (1982).

The case reported here could be defined as a dysplastic metanephric hamartoma of the sacral region. The differential diagnosis should include extrarenal Wilms tumor (Malik et al. 1967; Thompson et al. 1973; Dehner 1975; Akhart et al. 1977; Bittencourt et al. 1981). The absence of undifferentiated cellular mesenchyme and the prevalence of well differentiated and dysplastic renal structures are the basic distinguishing features. Bittencourt et al. recently reported an extrarenal Wilms' tumor of the uterus and pouch of Douglas which was a very well differentiated nephroblastoma with preponderant glomerular and tubular structures, thereby assuming an appearance similar to our case.

Metanephrogenic tissue has been described in ovarian teratomas (Nicholson 1934; Nogales et al. 1980).

Another consideration is the supernumerary kidney which usually lies within the renal fascia and possesses a drainage to the normal ipsilateral ureter. Even when it is distantly displaced, it can be recognized by its renal configuration (Burges 1969).

Nephrogenic adenoma of the bladder is a rare lesion of the trigone with a cystoscopic papillomatous appearance whose histology reveals tubular and cystic structures covered by cuboidal or columnar epithelium resem-

bling distal convoluted tubules, loops of Henle and collecting tubules (Goldman 1972; Allan 1975; Sussman et al. 1974; O'Shea et al. 1981). However, glomeruli and Bowman's capsules are absent in this lesion type, for which a metaplastic pathogenesis has been advocated elicited by chronic inflammation instead of dysontogenetic origin. Clinically this lesion affects adults and manifests with haematuria, dysuria, nocturia and backache.

Similarly, mesonephric tumors of the female genital tract and urethra can be easily distinguished from our lesion type by means of histological and clinical criteria (Wade-Evans and Laugley 1961; Novak and Woodruff 1974; Schnoy and Leistenschneider 1982).

Renal angiomyolipoma with tubular epithelial structures reported by Greville Williams (1963) and retroperitoneal lipoma with haematopoiesis and urogenital tissues reported by Tokumitsu et al. (1981) bear a partial but vague histological affinity to our case.

The present case is reported to provide additional information and confirm the intimate dysplastic nature of what appears to be an extremely unusual expression of renal maldevelopment.

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