Patología *1996; 29: 279-283*

Mesoblastic nephroma of adulthood. Immunohistochemical study of a case with unusual growth pattern.

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SUMMARY

Mesoblastic Nephroma (MN) is an uncommon congenital tumor of infancy that rarely occurs in adults. We report a case of congenital mesoblastic nephroma in a 48-year-old man.

The tumor showed an unusual pattern of growth as an intracystic polypoid mass, involving the renal parenchyma without extension into the renal pelvis. Microscopically, it consisted of an uniform spindle cell proliferation with entrapped dilated renal tubules. Spindle cells showed immunoreactivity for vimentin, muscle-specific actin and desmin, but were nonreactive for neuron specific enolase (NSE), S-100 protein, alpha-1-antitrypsin, alpha-1-antichymotrypsin and Factor VIII-related antigen. Keratin and epithelial membrane antigen (EMA) were only positive in the epithelial component.

A combination of histological and immunohistochemical findings is useful in the differential diagnosis of mesoblastic nephroma from other tumors of the kidney with spindle cells component.

Key words: Kidney tumor. Mesoblastic Nephroma. Immunohistochemistry.

INTRODUCTION

Mesoblastic Nephroma (MN) is a rare congenital tumor of infancy first described by Bolande et al (1) in 1967. Prior to the work by Bolande et al, these neoplams had been identified with different terms, including fibroma, leiomyoma (2), and leiomyomatous hamartoma (3). MN is usually discovered during the first few weeks of life and is often confused with Wilms' tumor, but it is almost invariably benign. It usually appears as an unencapsulated tumor composed of conective tissue cells, smooth muscle cells, renal tubules that are often cystic and lack of anaplasia or mitotic activity. The first case of MN in adults was described by Block et al (4) in 1973, and it was grossly and microscopically similar to congenital mesoblastic nefroma. Since then several other cases in adulthood have been published (5-13).

We report a new case of an unusual form of MN in an adult patient which presented as intracystic polypoid mass. An immunohistochemistry study is included.

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CASE REPORT

A 48-year-old man presented with pain in the right flank. The physical examination and routine laboratory tests were normal. Intravenous urography showed a mass in the lower pole of the right kidney distorting the lower calyces. Echography and computed tomography confirmed the presence of a solid-cystic mass in the lower third of the right kidney with no disruption of the capsule or vein invasion (fig. 1A). The patient underwent right radical nephrectomy, and was alive without recurrence 16 months after surgery.

PATHOLOGIC FINDINGS

The right kidney weighed 240 g, and contained an encapsulated, firm and grayish-white, 4-cm-diameter polypoid intracystic tumor that compressed the surrounding renal cortex. The tumor extended to the renal capsule, bulging it but without invasion. This lesion had no continuity with the renal pelvis (fig. 1B). Microscopic examination revealed a circumscribed tumor separated from the uninvolved kidney by a narrow band of fibrous and smooth-muscle tissue. The tumor

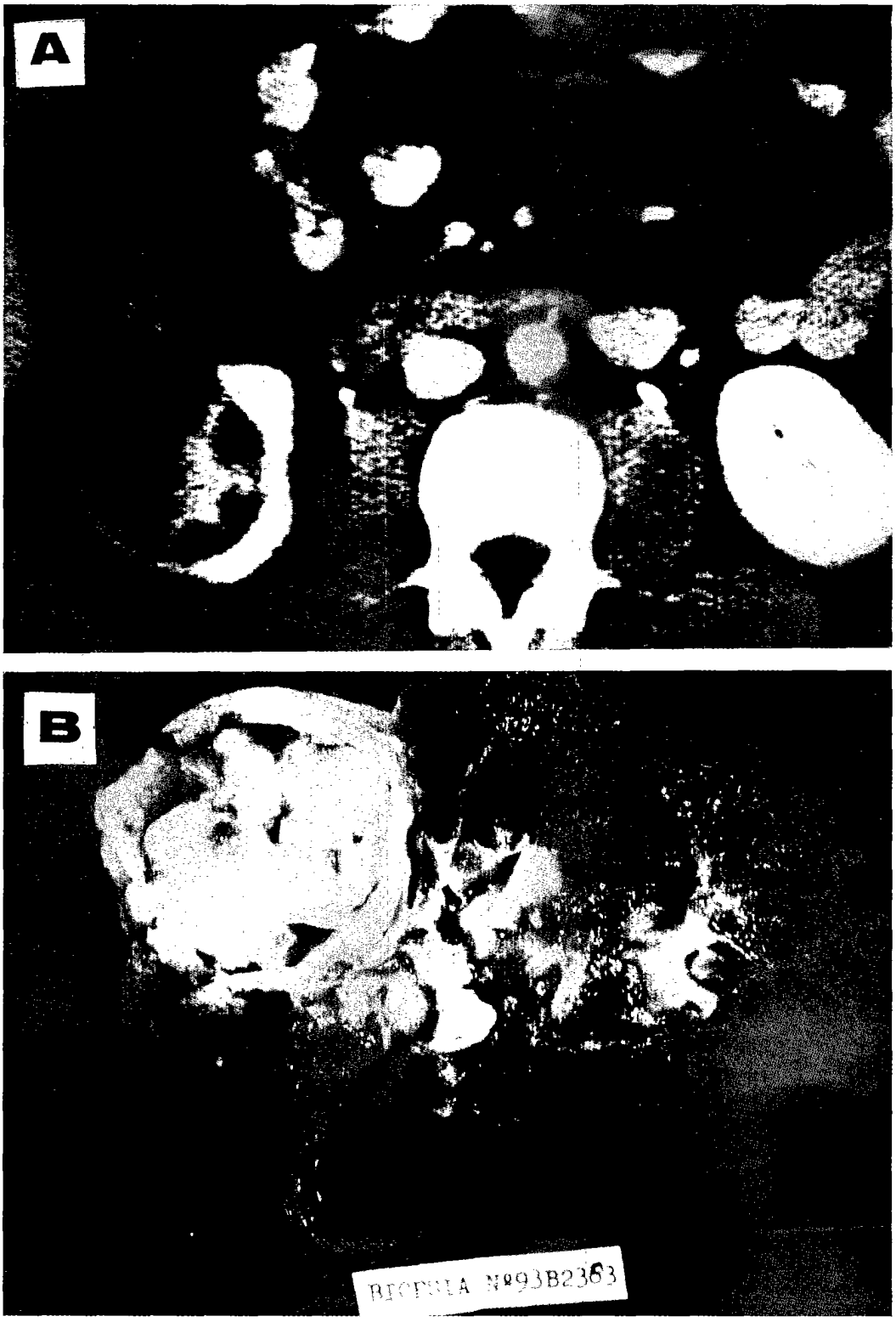

Figure 1.- (A) Computed tomography: solid-cystic mass in the right kidney. (B) Section of the right kidney showing a polypoid intracystic mass without continuity with renal pelvis.

appeared as an intracystic polypoid mass composed in some areas of uniform, tightly packed spindle cells with elongated nuclei, while in others areas the cells were plump (fig. 2A). The polypoid mass was lined by a single layer of cuboidal or flattened epithelium. Among the spindle cell areas and near to the implantation pedicle, a few dilated tubular structures were observed (fig. 2B). Scattered lymphocytes were present. No hemorrage, necrosis areas or mitoses were identified.

Immunohistochemistry was performed on

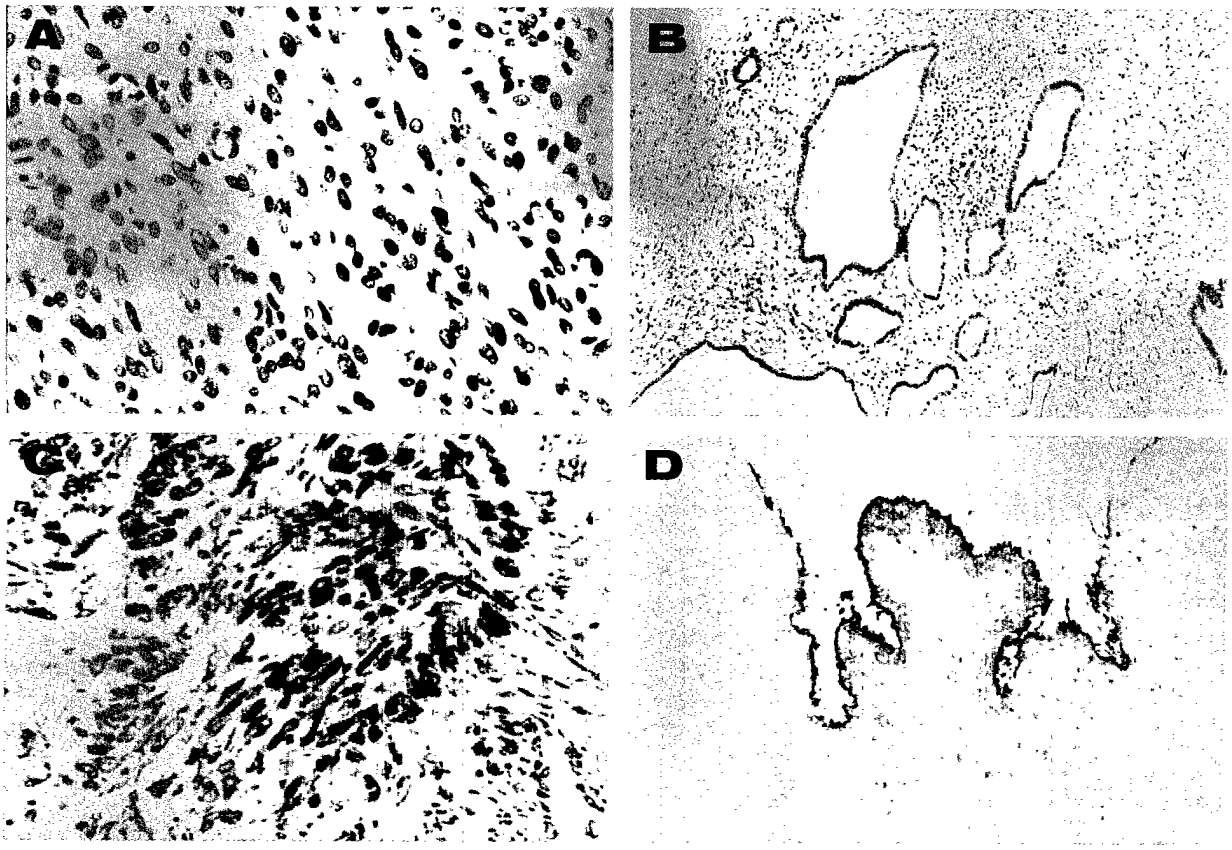

Figure 2.- (A) Histological appearance of mesenchymal stroma composed of spindle-shape cells. (B) Nodular focus of cystically dilated renal tubules lined by cuboidal epithelium. (C) Fascicles of spindle-shape cells showing immunoreactivity for Desmin. (D) Polypoid mass surface lined by cuboidal epithelium positive for EMA.

formalin-fixed and paraffin-embedded tissue using the conventional avidin-biotin alkaline phosphatase method (14). The results of immunohistochemical staining revealed an intense cytoplasmic positivity in the majority of spindle cells for vimentin; many of them also showed immunoreactivity for muscle differentiation markers like desmin (fig. 2C), and muscle-specific actin. The epithelium lining the cysts and tubules stained strongly with wide-spectrum keratin, and epithelial membrane antigen (EMA) (fig. 2D). S-100 protein, neuron specific enolase (NSE), alpha-1-antitrypsin and alpha-1-antichymotrypsin were negative. Factor VIII-related was only positive in in vascular structures.

DISCUSSION

Mesoblastic nephroma rarely occurs in adults, we have found no more than eighteen cases previously reported in the literature (4-13). Interestingly, most of these cases, showed certain differences from MN ocurring in infancy. The cases of adult MN tend to be often encapsulated or well circumscribed, more collagenized and they usually have a more prominent tubular component. Our case displayed an unusual growth pattern as a polypoid intracystic mass without any continuity with the renal pelvis. A similar case of polypoid growth has been previously reported by Durham et al (12), but the authors described it as a tumor with an intrapelvic extension rather than an intracystic growth.

Immunocreactivity for vimentin, desmin and muscle actin, in the spindle cells component, and keratin and EMA in the tubular epithelial component were expressed in our case. Similar results have been bound in other congenital and adult mesoblastic nephromas (15-16).

A combination of histological and immunohistochemical findings is useful to distinguish mesoblasic nephroma from other tumors of the kidney with spindle cells. Wilms' tumor, is rare in adults and can be usually separated from MN by the presence of blastema or embryonal tissue (17). Sarcomatoid renal cell carcinomas show evident cellular atypia and mitosis. Immunohistochemistry and electron microscopy reveal epithelial differentiation in the spindle cell component. Angiomyolipoma is composed of smooth muscle, thick-walled vessels and adipose tissue. When the smooth muscle predominate, may be also mistaken for adult MN and many areas of the tumor must be studied. Multicystic nephroma is encapsulated, unilateral and does not communicate with the pelvis. It is mainly composed of

multiloculated cysts with spindle fibroblastic or scatered muscle cells in the septa of the cystic cavities. Nodular solid areas are also absent (18).Nephrogenic adenofibroma has been described as an entity that occurs in young people (mean age 13.3 years) (19) These neoplams are usually solitary and nonencapsulated. They show a marked proliferation of epithelium similar to the hyperplastic nephrogenic rests (nephrobastomatosis). Spindle cells in nephrogenic adenofibroma are positive for vimentin but not for actin and desmin, whereas the embryonal epithelium stains positively for keratin but not for EMA.

Other entities with spindle cells including those of neurogenic, fibrohistiocytic or vascular origin can be distinguished from MN by nonreactivity for S-100, NSE, alpha-1-antitrypsin, alpha-1-antichymotrypsin and factor VIII.

The clinical behaviour of adult MN appears to be benign. Only one patient monitored for 21 years had local recurrence (5). Inadequate resection of the renal tumor may lead to recurrence, especially for those lesions occurring in infants over 3 months of age and children, showing histologic atypia and zones of necrosis (Atypical Variant of MN) (20). Recent karyotypic reports have suggested that extra copies of chromosome 11 seems to be the most common cytogenetic abnormality in MN, but this aberration has been found in only MN with cellular or atypical components. However, the genetic aberrations described do not seem to correlate with prognosis (21). Because the clinical signs and symptoms and imaging studies are very similar to other renal tumors, including renal carcinoma, the treatment of choice should be nephrectomy, probably curative in most of cases. Careful-long-term followup should be indicated only in those patients that have atypical or cellular MN but not for adult patients with typical histology MN.

RESUMEN

El Nefroma Mesoblástico es un tumor congénito infantil poco frecuente que ocurre muy raramente en la edad adulta. Presentamos un caso de nefroma mesoblástico congénito en un hombre de 48 años de edad.

El tumor mostró un patrón insólito de crecimiento como una masa polipoide intraquística, afectando al parénquima renal y sin extensión a la porción pélvica. Microscópicamente, el tumor estaba constituido por un componente de células fusiformes englobando túbulos renales dilatados. El componente fusocelular mostró positividad para vimentina, actina muscular específica y desmina, siendo negativo para enolasa neuronal específica (NSE), Pr-S100, alfa-1-antitripsina, alfa-1-antiquimotripsina y factor VIII. Citoqueratina y antígeno epitelial de membrana (EMA) mostraron positividad sólamente en el componente epitelial.

El estudio combinado de los hallazgos histológicos e inmunohistoquímicos es un método muy útil para establecer el diagnóstico diferencial del nefroma mesoblástico con otros tumores renales que presenten también componente fusocelular.

Palabras clave: Tumor renal. Nefroma mesoblástico. Inmunohistoquímica.

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