MESOCOLIC MYXOID HAMARTOMA SHOWING NEURAL DIFFERENTIATION: An Ultrastructural and Immunohistochemical Study

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A myxoid mesocolic tumor in a 4-month-old female infant is described. The tumor weighed 190 g and measured 8 cm in diameter and presented as a single mass composed of stellate and round cells with vesicular nuclei and prominent nucleoli in a myxoid, well-vascularized stroma. Electron microscopy showed mesenchymal cells with dilated rough endoplasmic reticulum, cell projections with primitive and desmosome-like cell junctions, and basal membrane-like material. Immunohistochemical studies demonstrated immunoreactivity with antibodies against vimentin and S-100 protein. The infant is well and without evidence of disease 3 years after surgery. This is an example of a peripheral myxoid hamartoma with neural differentiation, and the tumor is expected to have a benign clinical course.

KEY WORDS: myxoid hamartoma, Schwannoma, mesocolon.

INTRODUCTION

Primary mesenteric tumors in children are rare. Most frequently, cystic lesions such as lymphangiomas are found. Less frequently, fibromatosis and undifferentiated sarcomas are observed.1-3 Recently, Gonzalez-Crussi et al.1 described a very unusual primary multifocal omental neoplasm, apparently benign, that they called omental myxoid hamartoma of infancy. Its histogenesis has not been elucidated.

A similar but localized tumor with ultrastructural and immunohistochemical features suggesting neural differentiation is presented. Its differential diagnosis, therapy, and prognosis are discussed.

CLINICAL HISTORY

A 4-month-old female infant presented with multiple congenital malformations. She was the product of a normal, full-term delivery and
weighed 1,950 kg at birth. She had a colostomy for imperforate anus. A persistent ductus arteriosus was also found. On clinical examination an abdominal tumor mass was discovered. At surgery, a solid, well-circumscribed, spherical tumor attached to the middle portion of the transverse mesocolon was identified. No other tumors were found. There was no family history of von Recklinghausen’s neurofibromatosis, and the patient did not have café-au-lait spots or cutaneous pedunculated lesions. Postoperatively, the course was uneventful, and the patient is alive and well without evidence of tumor 3 years after surgery. In the interval cutaneous lesions have not developed.

**MATERIAL AND METHODS**

Samples of the tumor were fixed in 10% neutral formalin, embedded in paraffin, and sections stained with hematoxylin and eosin, PAS with and without diastase digestion, Masson’s trichrome, Alcian blue, toluidine blue, and silver reticulin stains. Oil red 0 on frozen sections and Bodian stain were also performed. For electron microscopy, 1 mm³ pieces were recovered from formalin, postfixed in 1% OsO₄, and embedded in Epon 812. Ultrathin sections were stained with uranyl acetate and lead citrate and observed under a Zeiss EM-109 electron microscope.

Immunohistochemical studies were performed on sections of formalin-fixed and paraffin-embedded tumor tissue using the avidin-biotin complex technique. Primary antibodies included rabbit anti-S100 protein (Dakopatts, Denmark) at 1:100 dilution, rabbit anti-keratin (Dakopatts, Denmark) at 1:400 dilution, and mouse monoclonal antibody against vimentin (clone 13.2, Bio-Yeda, Rehovot, Israel).

**PATHOLOGIC FINDINGS**

The tumor was a solid, spherical mass about 8 cm in diameter and weighed 190 g. The cut surface was whitish yellow, fasciculated, and myxoid (Fig. 1). The consistency was firm, partly soft. No necrosis, cavitation, or hemorrhages were identified.

Microscopic examination disclosed a well-vascularized myxoid tumor with scant mesenchymal cells (Fig. 2a). Cellularity varied in different tumor fields. The tumor cells had round to oval nuclei with prominent nucleoli and moderate amounts of cytoplasm, partly vacuolated. Scattered binucleated and multinucleated cells and slightly irregular and indented nuclei were also observed (Fig. 2b). Mitosis were not found. The stroma gave a positive reaction with Alcian blue stain at pH 2. PAS reac-
Bisected tumor reveals a solid, partly fasciculated and myxoid appearance.

Section and toluidine blue at pH 4, as well as Oil red 0 and Bodian stains, were negative. Sparse collagen fibers and a pericellular pattern of reticulin fibers were found.

Electron microscopy showed tumor cell nuclei with irregular contours and dense nucleoli. The cytoplasm contained scant organelles with prominent dilated rough endoplasmic reticulum (Fig. 3). Basal membrane-like material surrounded some cells, and intercellular desmosome-like junctions were present in relation to closely apposed cytoplasmic projections (Figs. 4, 5). Lipid vacuoles were not identified. No Luse bodies or long-spacing collagen fibers were found.

Immunohistochemically, the tumor cells gave a mild to moderate reaction with antibodies against S-100 and vimentin, but failed to stain with anticytokeratin antibodies.

**DISCUSSION**

The electron microscopic and immunohistochemical results suggest that the tumor cells are mesenchymal, with some features of neural cells. Positive staining for vimentin and S-100 protein, together with the presence of basal membranelike material and cytoplasmic projections with
primitive cell junctions, all support the notion that this lesion may be a variant of Schwannoma (neurilemoma). Neurilemoma is an encapsulated nerve sheath tumor with cellular (Antoni A) and myxoid (Antoni B) areas. Common locations are the head, neck, and flexor surfaces of the extremities, and, when deeply situated, in the posterior mediastinum and retroperitoneum. This tumor shows a consistent positive reaction for S-100 protein and, by electron microscopy, consists almost exclusively of Schwann cells. The main differential diagnosis is the neurofibromata, which ultrastructurally consists of a mixture of mesenchymal cells, axons, fibroblasts, and Schwann cells.

Gonzalez-Crussi et al. recently described 3 similar cases. Two of them, infants between 4 and 6 months of age, presented with a mesenteric mass, 4–10 cm in diameter, associated with multiple peritoneal nodules consisting of similar tumoral tissue. Two of these tumors were considered variants of liposarcoma and treated with chemotherapy. The tumor of the third infant, diagnosed as myxoma or undifferentiated sar-
coma, was surgically resected. The tumors were made up of mesenchymal cells showing prominent vesicular nuclei and one or two nucleoli, within a myxoid matrix and vascularized stroma with varying degrees of inflammatory infiltrate. Ultrastructurally, these cases showed a cell population composed of immature mesenchymal cells, fibroblastlike cells

![Image of tumor cell with rough endoplasmic reticulum, mitochondria, and lysosomes.](image)

**Figure 3.** Well-developed rough endoplasmic reticulum, scant mitochondria, and lysosomes are seen in the cytoplasm of a tumor cell. Uranyl acetate and lead citrate. $\times 20,000.$
and vacuolated cells, presumably phagocytic (histiocytic?). By electron microscopy lipoblasts were not found. The tumor cells were closely apposed, but intercellular junctions were not identified. In our opinion, Figs. 6 and 7 of their paper apparently show a discontinuous external condensation of lamina externalike material. Follow-up in these cases re-
vealed 1–15 years disease-free periods. The histopathologic and ultrastructural features of the present case are similar to those cases. Although in the cases described by Gonzalez-Crussi et al. immunohistochemistry was not performed, the possibility of multifocal omental Schwannomas cannot be ruled out.

**FIGURE 5.** Cytoplasmic projections have desmosomelike cell junctions. Uranyl acetate and lead citrate. × 40,000.
Current knowledge suggests that the most effective therapy for this tumor is surgical resection, because these lesions apparently represent biologically benign tumors. The present case has not recurred after excision.

REFERENCES


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