

Congenital occurrence of solitary infantile myofibromatosis of the spleen

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Abstract

Infantile myofibromatosis (IM) is a rare soft tissue tumor of infancy and childhood. We report the case of a newborn girl with an abdominal tumor discovered at 32 weeks of gestation by fetal ultrasound. She underwent a laparotomy for an unexplained abdominal mass 20 days after birth. The tumor originated from the spleen and was removed by splenectomy.

There were no other abnormal findings on diagnostic modalities. Based on the histological examinations, the tumor was diagnosed as an IM.

Though extremely rare during the neonatal period, solitary type IM should be considered as a differential diagnosis of the splenic tumor.

Key words: infantile myofibromatosis, solitary type, spleen

Introduction

Infantile myofibromatosis (IM) originates from the myofibroblasts and occurs mostly in young children under the age of 2 years. IM and infantile hemangiopericytoma (IHP) represent different stages of maturation of the same entity. [1] [2] Only very rarely does IM or IHP originate solitarily in the spleen. The first documented pediatric patient with IHP of the spleen was a 10-year old boy reported by Ciftci et al. [3] in 1999. To the best of our knowledge there have been no earlier reports on IM of the spleen during the neonatal period. We herein report a neonatal patient with solitary type IM of the spleen. Based on our experience with this case, we recommend that IM be considered in the differential diagnosis of splenic masses in neonates.

Case report

The girl was born by a normal delivery after 37 weeks of gestation. An abdominal tumor, 5.7 x 3.9 cm in size, had been discovered five weeks

earlier, at 32 weeks of gestation, by fetal ultrasound. A follow-up fetal ultrasound at 34 weeks of gestation revealed the tumor had grown to 6.0 x 5.5 cm in size. Figure 1 shows magnetic resonance imaging at 36 weeks of gestation. Her weight at birth was 3,170 g, with Apgar scores of 9 and 9 recorded at 1 and 5 minutes, respectively. She was admitted to our hospital on the 11th day after birth for further examination of the undefined abdominal tumor.

Examination on admission revealed a palpable hen-egg sized tumor on her left upper abdomen and cutaneous hemangiomas on her left lateral chest and right elbow. There were no other abnormal lesions on diagnostic modalities. Laboratory test findings were within normal ranges. The five tumor markers tested, alpha-fetoprotein, vanillylmandelic acid, homovanillic acid, neuron specific enolase, and human chorionic gonadotropin, were also within normal ranges. An abdominal computed tomography with contrast enhancement showed a 9.5 cm tumor enhanced heterogeneously in her left lateral abdomen (Figure 2). The spleen was pressed toward the ventral direction by the tumor.

The patient underwent a laparotomy on the 20th day after birth.

Upon completing a transversal incision of the upper abdominal wall, we discovered that the tumor originated in the spleen and occupied almost all of the normal spleen area. Based on these findings, we decided to perform a total splenectomy. The excised tumor weighed 250 g and measured 9.5 cm in diameter at the widest point. The tumor was solid and the cut surface was yellowish white in color (Figure 3). On the basis of the intraoperative histological findings of the frozen sections, the tumor was initially suspected as a hemangiopericytoma.

Microscopically, the mass consisted of a proliferation of spindle- to oval-shaped cells accompanied by primitive hemangiopericytomatous vascular channels. Bundles of myofibroblastic cells with eosinophilic cytoplasm were observed in some areas (Figure 4). We examined five immunohistochemical stains: alpha-smooth muscle actin (reacts with tumors arising from smooth muscles and myoepithelial cells), vimentin (the major subunit protein of the intermediate filaments of mesenchymal cells: immunohistochemical staining for vimentin is characteristic of sarcomas), HHF35 (an antibody recognizes muscle specific alpha and gamma actin isomers), CD34 (an antibody reacts with haematopoietic precursor cells

and endothelial cells) and S-100 protein (immunoreactive S-100 protein localizes in the cytoplasm and nuclei of astrocytes, Schwann's cells, ependymomas and astroglomas). The tumor cells were focally positive for alpha-smooth muscle actin, vimentin, and HHF35, but negative for CD34 and S-100 protein. These features were compatible with solitary type IM.

A prophylactic anti-microbial treatment was administered after the surgery to prevent an overwhelming post-splenectomy infection. The patient recovered fully and has developed without adverse events over the past 3 years since the operation. There has been no evidence of tumor recurrence and the cutaneous hemangiomas are regressing spontaneously.

Discussion

Splenic tumors are classified into four categories: lymphoid tumors, non-lymphoid tumors, metastatic tumors, and tumor-like lesions such as cysts and hamartomas. IM is classified as an unusual non-lymphoid tumor. [3] The concept of IM, as described by Chung and Enzinger, [4] is

that of a tumor originating from the myofibroblasts. IM occurs mostly in young children, usually before the age of 2 years. Histologically, the lesions are made up of plump spindle-shaped tumor cells arranged in short bundles of a type frequently associated with reticulin and/or collagen. The lesions are positively stained with phosphotungstic acid-hematoxylin and immunoreactive with both smooth muscle actin and vimentin. When exposed to desmin, they only respond with variable reactivity. Depending on the area, IM can resemble nodular fasciitis, fibrous histiocytoma, neurofibroma, or infantile fibromatosis. It is not difficult to identify IM, however, when the clinical features and immunohistochemical findings are considered. [5]

There are two types of IM, solitary and multicentric. The solitary type is more common than multicentric and mainly affects the soft tissues of the head, neck, and trunk. Solitary IM in the primary visceral organs is extremely rare, and thus quite difficult prognosticate. On the other hand, multicentric IM of the primary visceral organs has been reported in a substantial number of cases, sometimes with fatal outcome. [6] [7] [8]

Solitary IM recurs in less than 10% of cases after excision, and in some

cases unresected tumors have been reported to spontaneously regress. [9]

The poor prognosis of multicentric IM with visceral organs makes it all the more important to diagnose IM prenatally. Future evaluations of consecutive cases will be required.

Infantile hemangiopericytoma (IHP) is characterized as a rare neoplasm originating from the pericytes, usually within the first year of life. In view of similarities with respect to age, location, histology, and clinical behavior, IM and IHP are considered to represent the same entity at different stages of maturation. [1] [2] In fact, neoplasms that were previously considered IHP are now classified as IM by many authors. [5]

IM and IHP originating from the spleen are quite rare. Since the first report of such an origin, published in 1989, six patients in adulthood and a ten year-old boy have been described in the literature. [3] [10] [11] [12] [13] [14] [15] All seven of these patients were diagnosed as hemangiopericytoma based on the histological findings. Their physical examinations and diagnostic modalities produced no specific presumptive findings that could distinguish hemangiopericytoma from other splenic masses.

To the best of our knowledge, our patient is the first of solitary IM of the spleen identified by prenatal ultrasound and diagnosed in the neonatal period. Splenic tumors are rare in neonates and extremely difficult to definitively diagnose. Thus, we recommend that IM be considered as a differential diagnosis for all unexplained tumors found in the neonate spleen. In this patient we decided to perform the splenectomy based on the intraoperative histological findings of the frozen section. In view of the difficulty of making a definite diagnosis with the frozen section, we consider splenectomy to be the first choice of intervention for this type of case. Tumor biopsy is another option, if the surgeon has some idea about the prognosis of the solitary IM. In treating such patients, it is also crucial to prevent overwhelming post-splenectomy infection and to watch carefully for possible recurrence in other organs.

Figure legends



Figure 1
Fetal MRI at 36 weeks of gestation showing left upper abdominal tumor (arrows).

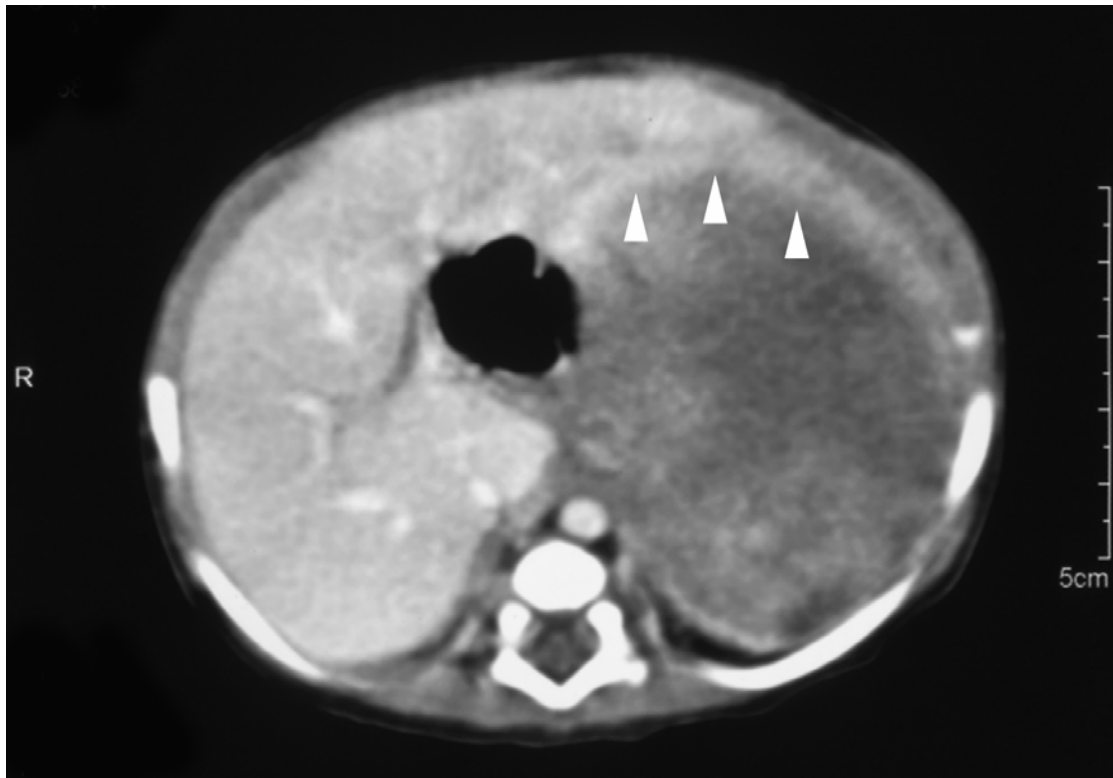


Figure 2

An abdominal CT with contrast enhancement showing a mass lesion in the spleen. The arrowheads indicate the splenic parenchyma forced against the ventral side.

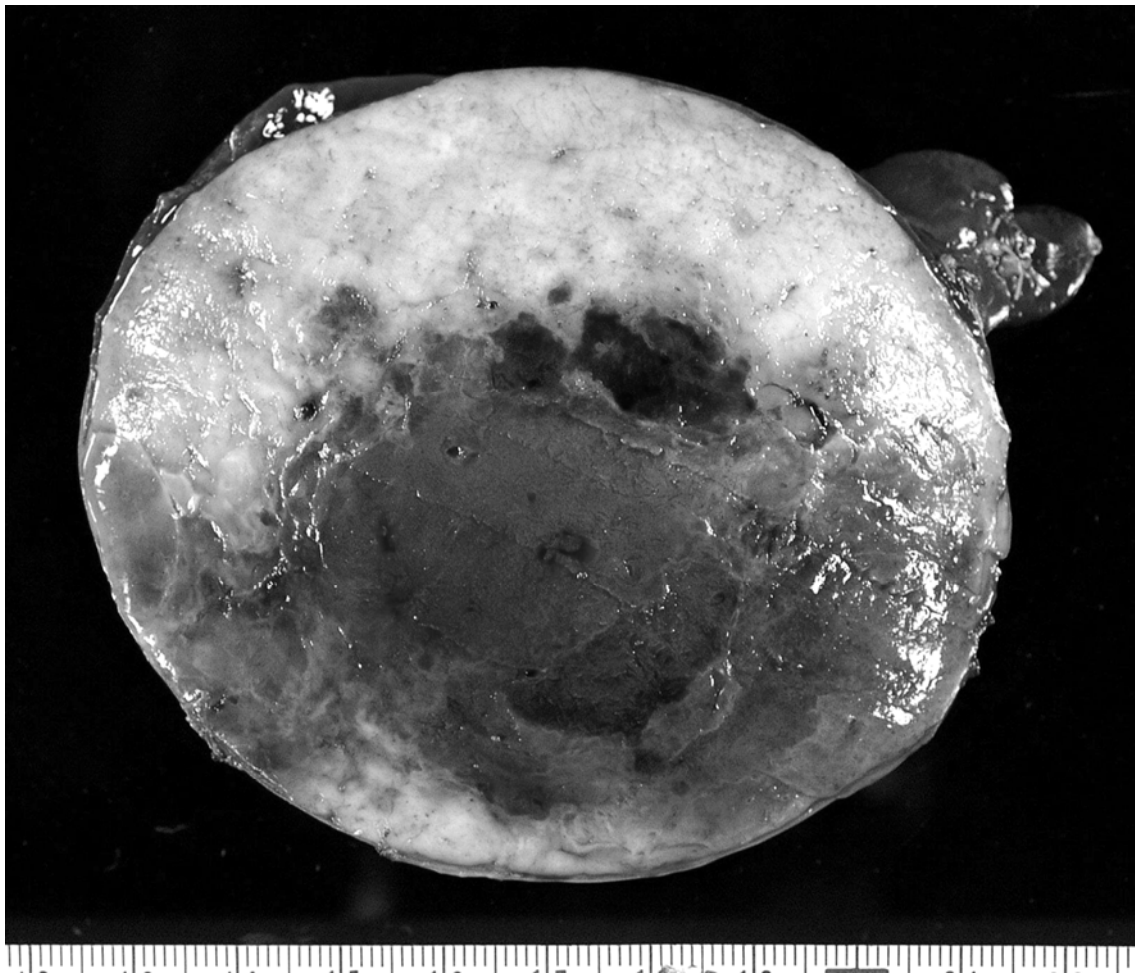


Figure 3
Cut surface of the tumor. The parenchyma is yellowish-white in color with some areas of hemorrhage.

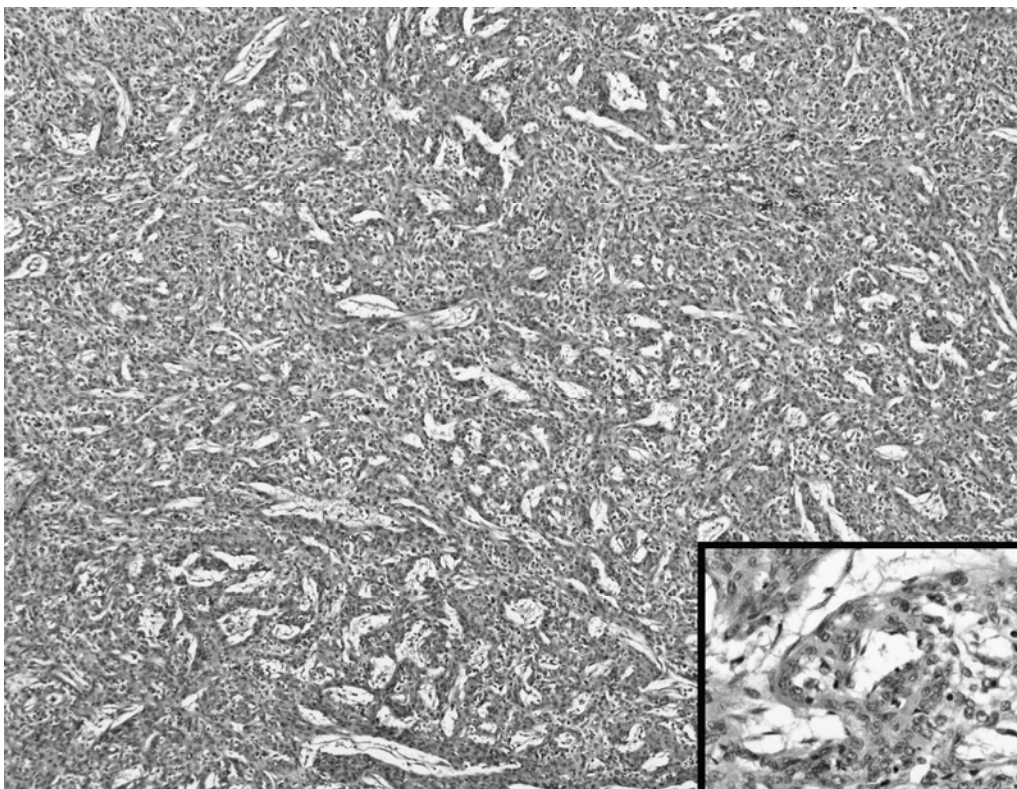


Figure 4

Light microscopy of the tumor. The tumor consists of spindle- to oval-shaped cells surrounded by primitive hemangiopericytomatous vascular spaces.

Hematoxylin & Eosin. Original magnification x10, (inset x40).

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