Solitary Fibrous Tumor of the Umbilical Region in a Pediatric Patient

Shruti Vaswani, MBBS, MD,1 Sudeep Khera, MBBS, MD,1 Arvind Sinha, MBBS, MS (Surgery), MCH (Paediatric Surgery)2

1Department of Pathology and Laboratory Medicine, All India Institute of Medical Sciences, Jodhpur, Rajasthan, India 2Department of Paediatric Surgery, All India Institute of Medical Sciences, Jodhpur, Rajasthan, India

INTRODUCTION

Fibrous tumors are rare tumors of mesenchymal origin that arise in the serosa,1 visceral organs, and soft tissues.2 In adults, most fibrous tumors are benign.3,4 The incidence of solitary fibrous tumors in children is rare.5 Solitary fibrous tumors are well-circumscribed, slow-growing lesions for which the treatment of choice is surgical resection. Approximately 15% to 20% of cases have been reported to recur or metastasize.5 Parameters that contribute to recurrence and/or metastasis are tumor size, cellularity, nuclear pleomorphism, hemorrhage, and necrosis.3 We present a case of fibrous tumor in a pediatric patient presenting with a bulge in the umbilical region, a rare site in this age group.

CASE REPORT

A 2-year-old male presented with swelling over the umbilicus for the prior 8 months. The swelling was initially small but gradually increased in size. The mass did not change size with changing positions, coughing, or straining and was not associated with any discharge. The patient had no vomiting, fever, abdominal distension, difficulty in micturition, diarrhea, constipation, or swelling anywhere else in the body. Because umbilical hernia is the most common umbilical disorder in this age group,7 a clinical diagnosis of umbilical hernia was considered.

Laboratory workup revealed hemoglobin of 10.9 g/dL (reference, 12.5 ± 1.5 g/dL), total leukocyte count of 11.78 K/mm3 (reference, 10 ± 5 K/mm3), and platelet count of 390 K/mm3 (reference range, 200–490 K/mm3).8 No radiologic imaging was done at our center. Imaging was done at an outside imaging center, but the parents of the patient lost the report, so we did not have access to the imaging and differentials offered with the imaging studies.

Intraoperatively, the surgeon did not find a hernial sac. The umbilical mass was excised and sent for histopathologic examination. The skin-covered greyish soft tissue mass measured 6 × 5.5 × 4.5 cm. The cut surface showed a homogenous greyish growth. On microscopic examination, a predominantly well-circumscribed encapsulated tumor was noted, with spindle shaped cells arranged in a haphazard manner and ectatic vascular channels. The cells were immunoreactive for CD34 and signal transducer and activator of transcription 6 (STAT6) and negative for smooth muscle actin, desmin, myogenin, MyoD1, CD99, epithelial membrane antigen, and beta-catenin.

Keywords: Molecular diagnostic techniques, solitary fibrous tumors, umbilicus
Figure 1. Gross examination revealed (A) a skin-covered soft tissue mass and (B) homogenous greyish white glistening areas on the cut section.

around the perivascular region. A sprinkling of inflammatory cells composed of plasma cells, mast cells, and lymphocytes was also noted. Mitosis was <1/10 high-power fields (reference, >4/10 high-power fields in malignant solitary fibrous tumors). No necrosis was seen.

Immunohistochemically, the tumor cells showed strong diffuse positivity for CD34 (Figure 2C) and signal transducer and activator of transcription 6 (STAT6) (Figure 2D). The tumor cells were negative for smooth muscle actin (SMA), desmin, myogenin, MyoD1, CD99, epithelial membrane antigen (EMA), and beta-catenin. The Ki-67 proliferation index was 2% to 3% only. Based on morphologic and immunohistochemical reactions, the mass was reported as a solitary fibrous tumor. At 1 year postoperatively, the patient remained disease-free.

DISCUSSION

Solitary fibrous tumors are tumors of fibroblastic origin with spindle cells arranged without a characteristic pattern within a collagenous stroma along with a framework of staghorn-like vessels (hemangiopericytoma pattern). The word hemangiopericytoma was coined in 1942 by Stout and Murray for tumors located in the buttock, retroperitoneum, and thigh. They postulated that these tumors originated from pericytic-modified smooth muscle cells situated around blood vessels. Hemangiopericytomas and solitary fibrous tumors were merged in the fourth edition of the World Health Organization classification of tumors of soft tissue, and since then, the term hemangiopericytoma has been interchangeably used with the term solitary fibrous tumor. The remaining 15 tumors were reclassified as other types of soft tissue tumors after application of STAT6 immunohistochemistry, highlighting the rarity of solitary fibrous tumors in children and the importance of STAT6 as the diagnostic marker. None of the patients in the Tan et al study presented with the umbilicus as the site of tumor.

Umbilical masses in children commonly include umbilical hernias, umbilical granulomas, remnants of the omphalomesenteric duct and urachus, and benign soft tissue masses, such as epidermoid cysts. Umbilical hernia commonly presents during the first few months of life after umbilical cord separation. A sac lined by peritoneum protrudes through an opening in the deep fascia of the abdomen. The protrusion becomes prominent when the baby cries or strains and is usually painless. Most umbilical hernias spontaneously resolve by the age of 3 years.
Pediatric Solitary Fibrous Tumor at the Umbilical Region

Figure 2. Solitary fibrous tumor shows a proliferation of spindle cells without a characteristic pattern and hemangiopericytoma vascular pattern with (A) hematoxylin and eosin (H&E) stain, magnification ×10, (B) H&E stain, magnification ×40, (C) diffuse membranous immunohistochemical staining of CD34 (magnification ×10), and (D) strong diffuse nuclear immunohistochemical staining of STAT6 (magnification ×10).

In the present case scenario, the umbilical mass did not show any movement on coughing or straining, which is unusual, so the clinician decided to excise the mass to avoid any unforeseen complications.

An umbilical granuloma is a nodule of tissue approximately 1 cm in size that might become apparent following umbilical cord separation and is usually pedunculated. The treatment of choice is application of topical silver nitrate.

The omphalomesenteric duct is an in utero connection between the terminal ileum and umbilicus. In most cases, the duct disappears after birth, but a part of it may persist. The Meckel diverticulum, an out-pouching of ileum, is the most common omphalomesenteric duct remnant. The common clinical presentation in children is obstruction. The presence of ectopic tissue increases the chance of the diverticulum becoming symptomatic. Diverticulectomy may be the treatment of choice in these cases.

The urachus connects the dome of the urinary bladder to the abdominal wall anteriorly at the level of the umbilicus. In utero, the urachus is a patent tube that closes after birth and forms the median umbilical ligament. Failure of the lumen to obliterate may result in conditions similar to omphalomesenteric remnants. A urachal cyst is a residual cyst without any communication to the bladder or umbilicus and is noted in the midline of the abdominal wall inferior to the umbilicus. It usually presents as a tender, swollen mass secondary to infection and should be excised.

Epidermal cysts are benign skin lesions usually involving the scalp, face, neck, and back but might be seen at the umbilicus. They may rupture and extrude keratin into the dermis, inciting a foreign body reaction. These cysts are usually asymptomatic unless infected and may be excised to avoid infection.

In the present case, efforts were made to rule out all other tumors of mesenchymal origin. Solitary fibrous tumors need to be histologically differentiated from similar spindle cell tumors such as malignant peripheral nerve sheath tumor, fibrosarcoma, leiomyoma/leiomyosarcoma, and malignant fibrous histiocytoma although these tumors are more common in adults than in children. In pediatric patients, solitary fibrous tumors should be differentiated from inflammatory myofibroblastic tumors.
Benign solitary fibrous tumors also need to be distinguished from malignant solitary fibrous tumors. Morphologic signs of malignancy include high mitoses (>4/10 high-power fields), moderate to marked nuclear pleomorphism, areas of high cellularity with nuclear crowding and overlapping, and the presence of necrosis or hemorrhage along with areas of stromal and/or vascular invasion. Our patient’s tumor had none of these features.

Various immunohistochemical markers were performed to rule out other spindle cell tumors. SMA and desmin ruled out smooth muscle tumors. MyoD1 and myogenin ruled out skeletal muscle tumors. CD99 and EMA ruled out other spindle cell tumors. Immunohistochemistry for analysis of soft tissue tumors. In: Wassef WA, Rhee R, editors. Enzinger and Weiss's Soft Tissue Tumors. 6th ed. Elsevier; 2017:1-201.

6. Robinson DR, Wu YM, Kalyana-Sundaram S, et al. Identification of recurrent NAB2-STAT6 gene fusions in solitary fibrous tumor by integrative sequencing. Nat Genet. 2013;45(2):180-185. doi: 10.1038/ng.2509

7. Disorders of the umbilicus in infants and children: a consensus statement of the Canadian Association of Paediatric Surgeons. Paediatr Child Health. 2001;6(6):312-313. doi: 10.1093/pch/6.6.312

8. Briggs C, Bain J. Reference ranges and normal values. In: Bain BJ, Bates I, Laffan MA. Dacie and Lewis: Practical Haematology. 12th ed. Elsevier; 2017:13.

9. Tan SY, Szymanski LJ, Galliani C, Parham D, Zambrano E. Solitary fibrous tumors in pediatric patients: a rare and potentially overdiagnosed neoplasm, confirmed by STAT6 immunohistochemistry. Pediatr Dev Pathol. 2018;21(4):389-400. doi: 10.1177/1093526617745431

10. Stout AP, Murray MR. Hemangiopericytoma: a vascular tumor featuring Zimmermann’s pericytes. Ann Surg. 1942;116(1):23-63. doi: 10.1097/00000478-199408000-00004

11. Fletcher CDM, Bridge JA, Lee JC. Extrapleural solitary fibrous tumors. In: Fletcher CDM, Bridge JA, Hogendoorn PCW, Mertens F, eds. WHO Classification of Tumours of Soft Tissue and Bone. 4th ed. International Agency for Research on Cancer; 2013:80-82.

12. Park MS, Araujo DM. New insights into the hemangiopericytoma/solitary fibrous tumor spectrum of tumors. Curr Opin Oncol. 2009;21(4):327-331. doi: 10.1097/CCO.0b013e32832f9352

13. Xue Y, Chai G, Xiao F, et al. Post-operative radiotherapy for the treatment of malignant solitary fibrous tumor of the nasal and paranasal area. Jpn J Clin Oncol. 2014;44(10):926-931. doi: 10.1093/jjco/hyu100

14. Demicco EG, Park MS, Araujo DM, et al. Solitary fibrous tumor: a clinicopathological study of 110 cases and proposed risk assessment model. Mod Pathol. 2012;25(9):1298-1306. doi: 10.1038/modpathol.2012.83

15. O’Donnell KA, Glick PL, Caty MG. Pediatric umbilical problems. Pediatr Clin North Am. 1998;45(4):791-799. doi: 10.1016/s0031-3955(05)70045-6

16. Park JJ, Wolff BG, Tollefson MK, Walsh EE, Larson DR. Meckel diverticulum: the Mayo Clinic experience with 1476 patients (1950-2002). Ann Surg. 2005;241(3):529-533. doi: 10.1097/01.sla.0000154270.14308.5f

17. Suzuki S, Nagasaki A, Urachal remnants. Semin Pediatr Surg. 1996;5(2):107-115.

18. Kang S, Cho HK, Cho MK. Two cases of giant epidermal cyst occurring in the neck. Ann Dermatol. 2011;23 Suppl 1(Suppl 1):S135-S138. doi: 10.5021/ad.2011.23.S1.5135

19. Arakami-Hattori N, Tanaka T, Mukai K, Kishi K. Congenital epidermal cyst of the umbilicus: a case report. Mod Plast Surg. 2013;33(4):128-129. doi: 10.4236/mps.2013.34026

20. Matsumoto K, Okabe H, Ishizawa M, Egawa M. Intermittentarsopalangeal burstis induced by a plantar epidermal cyst. Clin Orthop Relat Res. 2001;385:151-156. doi: 10.1097/00000488-198908000-00003

21. Basterzi Y, Sari A, Ayhan S. Giant epidermoid cyst on the forehead. Dermatol Surg. 2002;28(7):639-640. doi: 10.1046/j.1524-4725.2002.01314.x

22. Immunohistochemistry for analysis of soft tissue tumors. In: Goldblum JR, Weiss SW, Folpe AL. Enzinger and Weiss’s Soft Tissue Tumors. 7th ed. Elsevier; 2020:129-201.

CONCLUSION

Solitary fibrous tumors in pediatric patients are rare, and the umbilicus is a rare site of origin. These tumors commonly originate from pleural surfaces but can occur at any site in the body. Histopathology and immunohistochemistry are the methods of choice for diagnosing solitary fibrous tumors. These tumors show haphazardly arranged spindle cells and vascular channels with hemangiopericytoma-like branching patterns showing immunoreactivity for STAT6 and CD34 markers. Surgical excision is currently the recommended therapy in the pediatric population.

ACKNOWLEDGMENTS

The authors have no financial or proprietary interest in the subject matter of this article.

REFERENCES

1. Torabi A, Lele SM, DiMaio D, et al. Lack of a common or characteristic cytogenetic anomaly in solitary fibrous tumor. Cancer Genet Cytogenet. 2008;181(1):60-64. doi: 10.1016/j.cancergencyto.2007.11.005

2. van de Rijn M, Lombard CM, Rouse RV. Expression of CD34 by solitary fibrous tumors of the pleura, mediastinum, and lung. Am J Surg Pathol. 1994;18(8):814-820. doi: 10.1097/00000478-199408000-00008

3. England DM, Hochholzer L, McCarthy MJ. Localized benign and malignant fibrous tumors of the pleura. A clinicopathologic review of 223 cases [published correction appears in Am J Surg Pathol 1991 Aug;15(8):818]. Am J Surg Pathol. 1989;13(8):640-658. doi: 10.1097/00000478-198908000-00003

4. de Perrot M, Fischer S, Bründler MA, Sekine Y, Keshavjee S. Solitary fibrous tumors of the pleura. Ann Thorac Surg. 2002;74(1):285-293. doi: 10.1016/s0003-4975(01)03374-4

5. Kanamori Y, Hashizume K, Sugiyama M, et al. Intrapulmonary solitary fibrous tumor in an eight-year-old male. Pediatr Pulmonol. 2005;40(3):261-264. doi: 10.1002/ppul.20250
