Case Report

Lower Abdominal Solitary Fibrous Tumor Suspected As A Urachal Lesion : A Case Report with Cytohistological Correlation

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Keywords: *Solitary Fibrous Tumor, Uncommon, Urachus, Intraabdominal*

ABSTRACT

Urachus is the embryological remnant of urogenital sinus and allantois. Benign urachal neoplasms including adenomas, fibromas, fibroadenomas, fibromyomas, and hamartomas have been reported. However, no cases of solitary fibrous tumour of the urachus have been reported.

An 18 year old male presented with a lower abdominal mass with pain and urinary frequency. Computed tomography showed a mass abutting dome of the urinary bladder and anterior abdominal wall; however, the fat plane was maintained. Fine needle aspiration cytology was suggestive of a mesenchymal lesion. Surgical exploration revealed a mass attached to umbilicus on one end and dome of bladder on the other. Histopathological diagnosis was solitary fibrous tumor which was corroborated with immunohistochemistry findings. Patient has not reported back with recurrence.

This case is being presented because of its unusual location and clinical suspicion of urachal lesion with no residual histological evidence of its remnant in the tumour.

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Introduction

Urachus is the embryological remnant of urogenital sinus and allantois. It extends upward from the anterior dome of the bladder towards the umbilicus and normally involutes before birth, remaining as a musculofibrous band in the form of median umbilical ligament with no known function.[1] It lies between the transversalis fascia anteriorly and the peritoneum posteriorly. Persistence of urachal remnant (seen in 50 % cases) or incomplete regression of urachus can give rise to various clinical problems. Benign urachal neoplasms including adenomas, fibromas, fibroadenomas, fibromyomas, and hamartomas have been reported. Malignant urachal neoplasms are also seen representing less than 0.5% of all bladder cancers, and are typically silent because of their extraperitoneal location. [2] However no cases of Solitary fibrous tumour (SFT) urachus have been reported. SFT represent a group of rare tumours of mesenchymal origin usually identified in the pleura. [3] Herein we report a case of intraabdominal SFT at the site of urachus.

Case Report

A 18 year old male presented with a lower abdominal mass of 6 months duration, with complaints of non specific abdominal and urinary frequency. On examination per abdomen a mass arising from the pelvis 2 cm, below the umbilicus was seen. (figure 1A) Computed tomography (CT) showed a, infraumbilical well circumscribed mass displaying peripheral solid component and intralesional hypodense areas of myxoid change, hemorrhage and necrosis, abutting the dome of the urinary bladder and the anterior abdominal wall; however, the fat plane was maintained. (figure 1B) The bowel loops were splayed around the lesion with no obvious evidence of infiltration. Cystoscopy revealed a bulging mass measuring 7.5x7.0x7.0 cm with no significant mucosal changes. Ultrasound guided fine needle aspiration cytology (FNAC) done twice showed hypocellular smears with occasional clusters of plump to ovoid spindle shaped cells, at places entrapped in pink stroma along with numerous acellular stromal fragments in a hemorrhagic background. A possible diagnosis of mesenchymal lesion was given; (Figure 2B) however, in view of the size excision was advised. Surgical exploration revealed that the mass was attached to umbilicus on one end and dome of bladder on the other. (figure1C) The mass was excised as a suspected urachal lesion along with bladder dome to rule out any malignancy. The excised mass was well circumscribed with attached umbilicus on one end with intervening fat and no fibrous band. On the other end of the tumor a part of bladder wall clearly separated from the tumor was identified. Grossly, the tumor was well encapsulated and nodular measuring 7.5 x 7.0 x 7.0cm with attached bladder dome clearly separated from the mass at one end and umbilicus at the other. Intervening fatty area was identified; however, no fibrous band was seen (Figure 2A). Cut section was grey white firm with solid and cystic areas. Microscopically sections showed spindled fibroblasts with minimal pleomorphism and negligible mitosis arranged in a patternless pattern with hyper and hypocellular areas, interlacing and intersecting fascicles with focal myxoid areas along with staghorn vascular proliferation, hemangiopericytoma like areas and perivascular hyalinization. (Figure 2C&D) Immunohistochemical (IHC) staining was performed with 5 micron thick sections on Poly-L-Lysine coated slides, which were stained with monoclonal antibodies against CD 34, CD 99 vimentin, Bcl2, Desmin, CD117, Smooth Muscle Actin (SMA), Epithelial membrane antigen (EMA), S-100 and cytokeratin (CK)(AE1/AE3). Also to evaluate the proliferative activity of neoplastic cells Ki 67 marker was used. Tumour cells were positive for Vimentin, CD34 and CD99, however negative for Bcl2, SMA, Desmin, EMA, S100, cytokeratin and CD-117 (Figure 3). The Ki-67 labelling index was <1% . A final diagnosis of benign SFT was given. However extensive histological examination of the tumour did not reveal any urachal remnants. The patient was from a far area village, so the follow up was hampered although has not reported back with any recurrence.

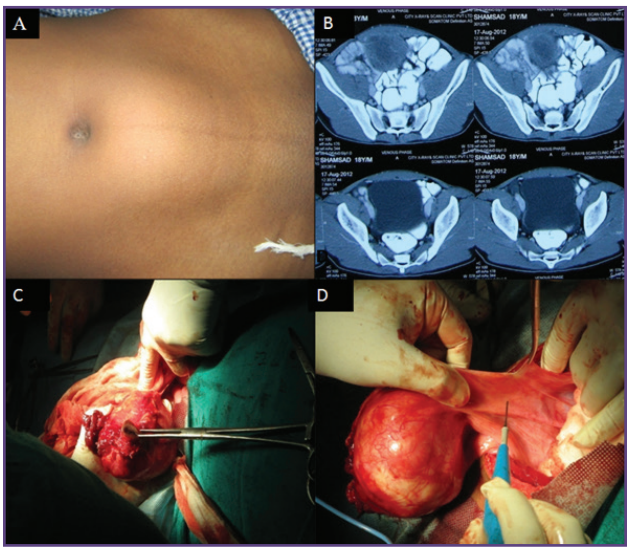

Fig. 1: (A) Patient presenting with a lower abdominal mass arising from the pelvis 2 cm below the umbilicus. (B) Computed tomography showed a well circumscribed mass with peripheral solid component and internal septations with areas of myxoid change , hemorrhage and necrosis abutting the dome of the urinary bladder and the anterior abdominal wall, however the fat plane was maintained. (C) & (D) Tumor removed en bloc together with the umbilicus and bladder dome.

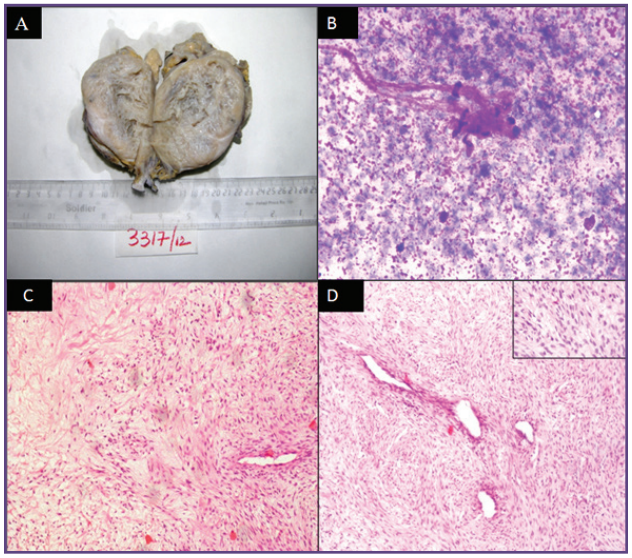

Fig. 2: (A) Cut section shows well encapsulated, nodular grey white tumor mass with solid and cystic areas. Arrow shows intervening fat and no fibrous band. (B) Cytosmear showing a stromal fragment with ovoid to spindle shaped cells and mild inflammatory infiltrate in the background. (Giemsa 40X). (C) Sections showing hyper and hypocellular areas with interlacing and intersecting fascicles with focal myxoid areas (H&E, 10X). (D) Sections showing staghorn vascular proliferation hemangiopericytoma like areas with focal perivascular hyalinization. (H&E, 10X) Inset shows fibroblasts with minimal pleomorphism and negligible mitosis. (H&E, 40X)

Discussion

SFT is a rare benign neoplasm arising from the submesothelial mesenchymal layer first described by Klemperer and Rabin in 1931. It most commonly arises in the pleura, though also recognized in extrapleural locations such as lung, mediastinum, pericardium, mesentery, peritoneum, extraperitoneal spaces, nose and paranasal sinuses. [4] Extrapleural SFTs represent 0.6% of all soft tissue tumors. SFTs of the abdominal wall are extremely rare, with only 16 cases reported in the literature till date. [3] Wherein no urachal origin was suggested.

SFTs usually present as an asymptomatic mass which can become painful as it grows , compressing the surrounding structures. [3] In our case the patient presented with non specific abdominal pain and urinary frequency , due to its pressure on bladder dome. SFTs have a same sex incidence being more frequent in 20-70 years age group, however female predominance has been observed in case of the abdominal wall tumors unlike in our case who is a 18 year old male with intra abdominal mass.

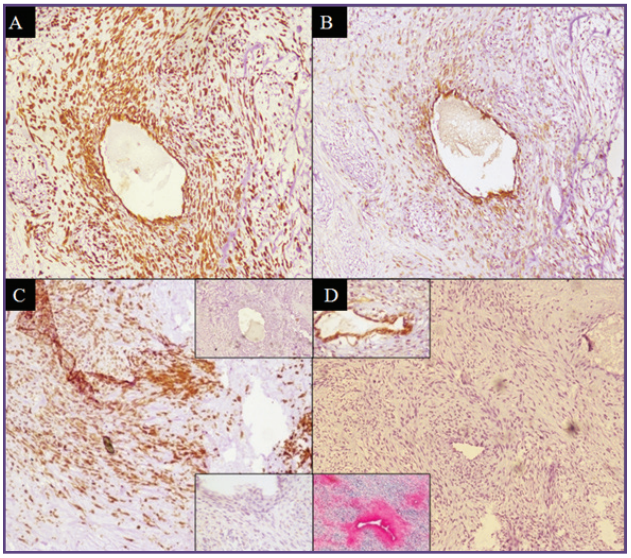

Fig. 3: (A) Strong cytoplasmic Vimentin (V9, IgG/1kappa, RTU, mouse monoclonal, Biocare) positivity (10 X). (B) Diffuse cytoplasmic immunoreactivity of CD 34 (QBEND/10, IgG1/kappa, RTU, mouse monoclonal, Dako) (10 X). (C) Strong CD 99 (H036-1.1, IgM, RTU, mouse monoclonal, Biocare), positivity (10 X). Inset shows CD 117 (Y145, IgG, RTU, Rabbit monoclonal, Biocare) and S-100 (15E2E2+4C4.9, IgG2ak+IgG2a, RTU, mouse monoclonal, Biocare) negative (10X). (D) Desmin (D33, IgG1/kappa, RTU,mouse monoclonal, Biocare) negativity (10 X). Inset shows SMA(IA4, IgG2a/kappa, RTU, mouse monoclonal, Biocare) negative (40X) and dual staining pattern with trichrome.

Histologically, (Table 1) SFTs are well circumscribed, encapsulated, solid, hypervascular tumor masses showing collagenous matrix with arrays of bland spindle cells arranged in a patternless pattern as hyper and hypocellular areas along with numerous branching staghorn vessels , also called as hemangiopericytoma like pattern. [3,4]

As per latest WHO classification 2013 the term "haemangiopericytoma" is abandoned . It is now used only to describe a morphological pattern that is shared by different entities. Currently, solitary fibrous tumour, haemangiopericytoma, lipomatous haemangiopericytoma and giant cell angiofibroma are all lumped under the category of "extrapleural SFT " .[3,4,5,6]

Because SFTs lack distinctive histological features, immunologic staining has frequently been employed to exclude other neoplasms in the differential diagnosis. Usually the ubiquitous and indolent nature of SFTs obscures and delays the diagnosis.

Immunohistochemically, (Table 1) SFTs commonly show strong and diffuse staining for vimentin in 100% of the

cases, CD34 in 85 %, bcl2 in 65 % and CD 99 in 40 %. 20-35% variable positivity for EMA and SMA is seen. S100 proteins, cytokeratin and/or desmin usually show negative staining in majority, however focal and limited reactivity has also occasionally been reported.[3,4,6,7,8,9] But because of its very rare positivity none of the studies mentions the exact percentage positivity of these markers.

The diagnosis of SFT in our case was not made easily. At the first glance by H&E staining, many differentials (Table 2) with one or some features similar to solitary fibrous tumour were considered.[3] Compared with these tumours, the distinct features of solitary fibrous tumour were observed on H & E. IHC showed weak and patchy CD 34 positivity, diffuse CD 99 and vimentin positivity . Bcl2, desmin, CD117, SMA, EMA S-100 and CK were negative. Cytokeratin was used with the aim to rule out a epithelial lesion and also to detect any urachal epithelial remnants. However no remnant of urachal tissue was identified microscopically. Histologically normal urachus consist of an epithelial canal lined by transitional epithelium (in 70%) and columnar epithelium (30%) surrounded by connective tissue and musculature.[2] While completely obliterated urachus appears as a fibrous band.

About 80% of SFTs are benign, malignancy is seen in 10–30% of the patients with prolonged follow up. [3] Such behavior has been related to certain histological features of these tumors. Criteria for malignancy, though still controversial, include tumor size (>5 cm), infiltrative growth, high cellularity, nuclear pleomorphism, necrosis and mitosis > 4/10Hpf , corresponding to a level of Ki 67 staining greater than 10%. [3,4,6,7] In a study of all the potential predictors of malignancy, only mitotic index was associated with malignant behavior.[7] However occasional studies consider tumor size as an independent prognostic factor.[4]

A weak/negative expression of CD 34 has been associated with more aggressive behaviour and an increased incidence of malignancy. [3,8] The staining is nearly uniform in histologically benign cases and patchy in malignant cases. [8] In the present case the tumor size was more than 5 cm and CD 34 staining was weak and patchy; although, the tumor cells showed minimal pleomorphism , negligible mitosis (<1/hpf) and no areas of necrosis. Keeping this in mind the patient was kept on follow up to look for relapse. Also the histological features of the cases negative for Bcl2 as in our case did not differ significantly from those of the positive cases. [8]

TABLE 1: HISTOPATHOLOGY AND IHC OF SFT [3,4,6,7,8]

GROSS	HISTOPATHOLOGY	IMMUNOHISTOCHEMISTRY
Well defined	Patternless pattern	CD 34 (85%), Bcl2 (65%), CD99 (40%)
Solid	Hypo and hypercellular areas	EMA and SMA occasional positive
Hypervascular	Thick collagen bundles, Branching vessels	S-100, Desmin & cytokeratin rare positive

TABLE 2: DIFFERENTIAL DIAGNOSIS OF SOLITARY FIBROUS TUMORS [3]

DIFFERENTIAL TUMOUR	SIMILARITIES WITH SFT	DIFFERENCES FROM SFT
Haemangiopericytoma	- Mesenchymal origin - Staghorn vascular spaces - Spindle-shaped tumor cells	- No combination of varying growth patterns - No collagenous stroma - Reticulin staining pattern - Weakly positive for CD34; negative for CD99
Deep fibrous histiocytoma	- Storiform pattern - Occasional presence of bizarre mesenchymal giant cells	- More circumscribed - More uniform growth pattern - Negative for CD34 - Positive for factor XIIIa
Smooth muscle tumor	- Fascicular growth of spindle cells	- Positive for Smooth muscle actin and desmin - Negative for CD34
Gastrointestinal stromal tumor	- Staghorn vascular spaces - Positive for CD34	- No collagenous stroma - CD 117 positive - Negative for bcl-2
Nerve sheath tumors	- Focal CD 34 positive	- No collagenous background or vascular pattern - Frequent palisading - S-100 positive
Low grade Fibromyxoid sarcoma	- Alternating fibrous and myxoid patterns - EMA and SMA weak occasionally	- No prominent vascular proliferation - Lacks thick collagen bundles - Collagenous rosettes, CD 34 negative

Surgical excision is the treatment of choice and it exhibits a favorable clinical behaviour, with an overall 10 year survival rate in 54-89% after primary surgical resection.[3]

Similar dilemma in the presentation was noted by Mizusawa et al [10] in their study.

We report a case of lower abdominal SFT mimicking as a primary lesion of urachus radiologically and surgically presumptively, due to its location. Although the preoperative diagnosis was a urachal tumor, the pathological diagnosis was a SFT.

Conclusion

This case is being presented because of its unusual location suspected clinically as a urachal lesion with no residual histological evidence of its remnance in the tumor. Care should be taken not to be too invasive, considering the possibility of a benign tumor.

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